Volume 23 • Supplement 1 • Pages S1–S144
doi:10.1017/S104795111300036X
Cardiology in the Young

47th Annual Meeting of the Association for European Paediatric Cardiology, AEPC with joint sessions with the Japanese Society of Pediatric Cardiology and Cardiac Surgery and Asia-Pacific Pediatric Cardiac Society

O1-1

Postoperative systemic inflammatory response is an important determinant for adverse two-year neurodevelopmental outcomes after the Norwood procedure

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Objectives: Neonatal cardiopulmonary surgery, particularly the Norwood procedure, carries a significant risk factor for adverse neurodevelopmental outcomes, which may be related to early postoperative systemic inflammatory response. We examined the relationship between postoperative C-reactive protein level (CRP), a marker of systemic inflammatory response, and two-year neurodevelopmental outcomes among the survivors after the Norwood procedure.
Methods: Charts of 53 neonates undergoing the Norwood procedure from 2003–2009 were reviewed. CRP was measured in 43 neonates twice weekly within postoperative day 20. Peak CRP levels were recorded, with peak total and differential white blood cell counts (lowest level of lymphocytes), glucose and lactate. Demographic data included age at surgery; gender; durations of CPB, aortic cross clamp, deep hypothermic circulatory arrest, and ICU stay; and socioeconomic status of the families. Two-year neurodevelopment of cognition, motor and language were prospectively assessed with The Bayley Scales of Infant and Toddler Development-III in 26 patients (9 deaths, 2 lost, 6 assessed with Bayley Scales of Infant Development-II).
Results: Mean ± SD scores were: coginitve, 91 ± 13; language, 86 ± 13; and motor, 85 ± 17. The peak CRP was 79 ± 37 mg/L. Univariate regression showed that cognitive scores significantly and negatively correlated with peak CRP ($p = 0.004$), and trended to a negative correlation with age ($p = 0.097$). Language scores significantly and negatively correlated with peak CRP ($p < 0.0001$) and age ($p = 0.005$). Motor scores trended to a negative correlation with age ($p = 0.08$). Multivariate regression showed that both cognitive and language scores significantly and negatively correlated only with peak CRP ($p < 0.01$ for both), but not with other clinical variables.
Conclusions: The magnitude of systemic inflammatory response, among the perioperative risk factors examined, is the most important determinant for adverse two-year neurodevelopmental outcomes of cognition and language after the Norwood procedure.

O1-2

Liver Fibrosis in Patients with Fontan Circulation Leads to Alteration in Portal Venous Hemodynamics

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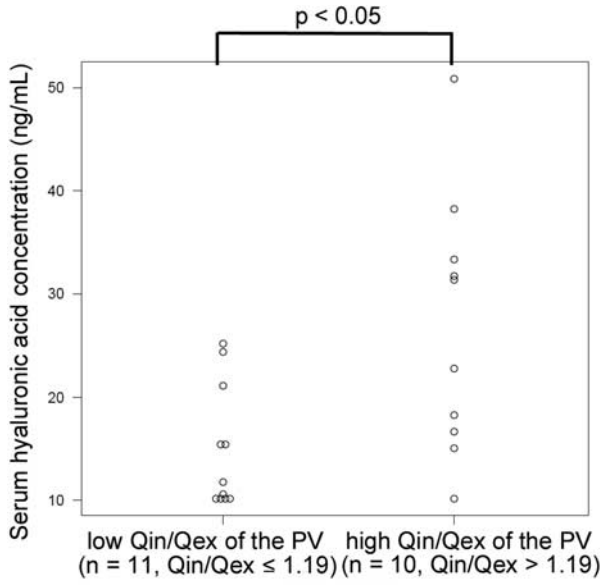
Introduction: Inspiratory-to-expiratory flow rate ratio (Qin/Qex) of the portal vein (PV) increases in patients with functionally poorer Fontan circulation. This alteration in PV hemodynamics may lead to long-term gastrointestinal complications of Fontan circulation, including liver dysfunction and protein-losing enteropathy. Moreover, it is not uncommon for Fontan patients to develop liver fibrosis in the late postoperative period. Although it is hypothesized that liver fibrosis may further exacerbate PV hemodynamics by impeding splanchnic blood flow, the influence of liver fibrosis on PV hemodynamics has not been investigated. We aimed to evaluate the correlation between Qin/Qex of the PV and liver fibrosis.
Methods: We studied 21 consecutive patients with Fontan circulation who underwent postoperative cardiac catheterization for various indications. Serum hyaluronic acid (HA) concentration was used as an indicator of liver fibrosis. Pulsed-wave Doppler recordings at the main portal trunk in the supine position was used to calculate the Qin/Qex; the median value was used to divide the patients into 2 groups.
Results: The median age at examination was 5.3 years (range, 2.2–25.8 years), and the median time interval between Fontan procedure and examination was 2.0 years (range, 3 months to 19.7 years). Three patients were classified as NYHA functional class II, and the others as NYHA functional class I. The median Qin/Qex of the PV was 1.19 (range, 0.96–1.48). Patients in the high Qin/Qex group had significantly higher serum HA concentration than those in the low Qin/Qex group (median 27.1 [10–50.7] ng/mL vs. median 11.9 [10–25.2] ng/mL, respectively; $p < 0.05$). No significant difference was noted in laboratory data including total bilirubin, liver enzyme, brain

natriuretic peptide levels, and platelet counts between both groups. Inferior vena cava pressure, superior vena cava pressure, systemic end-diastolic ventricular pressure, and Nakata index were similar for both groups.
Conclusions: Fontan patients with higher Qin/Qex of the PV had significantly higher serum HA concentration. Liver fibrosis in Fontan patients leads to worsening of PV hemodynamics, which may exert further negative effects on Fontan circulation.



O1-3
Outcome of dilated cardiomyopathy in very young children
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Introduction: Dilated cardiomyopathy (DCM) is a heterogeneous group of diseases linked by a common phenotype of cardiac chamber dilation. The prognosis is varying according to the underlying aetiology; however DCM is a serious disorder and according to literature up to 50% of children die or receive a heart transplant in the first five years after diagnosis
Design and setting: Our institutional database was retrospectively screened for patients below the age of ≤ 3 years diagnosed between January 01, 2006 and December 31, 2011 with DCM ($n = 50$). An extensive initial diagnoses program identified a cause for DCM in 72% of the patients (28% idiopathic). The patients were treated based on the actual guidelines. However, keeping in mind the poor outcome of these patients, we also used individual drug therapy and compassionate therapy approaches like bone marrow derived progenitor cell therapy or pulmonary artery banding in selected cases.
Results: The median age at presentation was 4.9 months. The median follow up was 19.7 months. Kaplan-Meier analysis of survival after DCM diagnosis revealed a 1-year survival of 97% and a 5-year survival of 86%. The rate of freedom from death or transplantation was 77% at 1 year and 67% at 5 years. The patients that survived and were not heart transplanted (median follow up 34.3 months) showed a significant increase in clinical condition (NYHA classification class 3.25 ± 0.85 to 1.42 ± 0.72), BNP (3436 ± 4132 pg/ml to 453 ± 965), FS (14.18 ± 6.25 to 24.61 ± 10.64) and LVEDD (3.49 ± 3.32 to 0.55 ± 2.68).
Conclusion: With the present data we can show that the rate of death or transplantation can be reduced distinctly in very young children with dilated cardiomyopathy. Our therapy was based on the actual guidelines combined with personal compassionate therapeutical approaches.

O1-4
Long-term outcome of 117 patients with univentricular heart and common atrioventricular valve
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Introduction: Few studies investigated the long-term outcome of patients with univentricular heart and common atrioventricular valve.
Method: We retrospectively analysed the medical files of all patients univentricular heart with common atrioventricular valve in the setting of heterotaxy or with unbalanced atrioventricular septal defect preventing biventricular repair.
Results: 117 patients were identified during the study period. 89/117 had a postnatal diagnosis. 28/117 patients never underwent surgery, 25/117 underwent one palliation surgery (Blalock-Taussig-shunt (BTS)/pulmonary banding), and finally, 61/117 patients entered a sequential cavopulmonary connection program: 37/61 had partial cavopulmonary connection at the time of data analysis while 24/61 had total cavopulmonary connection (TCPC). The average age at TCPC was 7.6 years ±4 years [1.7–16 years]. Three patients were eventually transplanted.
The overall mortality was 59% (69/117): 65% and 30% in heterotaxy and in patients with unbalanced atrioventricular septal defect respectively. Mortality was 85% (24/28) in the subgroup of patients who never underwent surgery, 93% in the subgroup of patients who had a BTS, and 89% after pulmonary banding. In the subgroup planed to have TCPC, 49% died after partial cavopulmonary connection and survival rate was 71.6% [50.7–100] in patients who had TCPC.
Conclusion: The long-term outcome of univentricular hearts with common atrioventricular valve treated in a tertiary referral center showed a high mortality rate. Patients with this kind of complex congenital heart disease should undergo the sequential univentricular program without delay in order to obtain better long-term survival.

O1-5
Exercise capacity in children after total cavopulmonary connection; lateral tunnel versus extracardiac conduit technique
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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Introduction: In patients with univentricular heart disease the total cavopulmonary connection (TCPC) has been the preferred technique for the last 20 years. Since the introduction, mortality and morbidity of the Fontan operation have improved vastly. Limited exercise tolerance remains a problem. It is not clear if exercise tolerance differs between both available techniques (i.e. intra-atrial lateral tunnel (ILT) or extracadiac conduit (ECC)) in the modern, 2-stage approach of the Fontan circulation. Purpose of this study was to compare exercise testing results between these two groups.
Methods: 82 Fontan patients (50 male) with a TCPC, age 12.4 ± 2.6 years, age at TCPC completion 3.2 ± 1.4 years, successfully underwent cardiopulmonary exercise testing (CPET) (peak respiratory exchange rate (RERpeak) >1.00) on a bicycle ergometer. Peak workload (Wpeak), peak heart rate (HRpeak), peak VO2 (VO2peak) and VE/VCO2-slope were determined. Predicted values were derived from a group of healthy controls. A distinction was made between ILT (n = 33) and ECC (n = 49) modifications of the TCPC-technique.
Results: For the entire group mean RERpeak was 1.08 ± 0.05, mean Wpeak was 70 ± 16%, mean VO2peak was 73 ± 15%, median VE/VCO2 slope was 107% (interquartile range (IQR) 16%) and mean HRpeak (170 ± 18/min) was 91 ± 10% of the predicted value.
There was no difference in age at time of the test between the 2 groups (13.0 ± 3.0 (ILT) vs. 11.9 ± 2.2 (ECC) years, p = 0.77). Outcomes for the ILT and ECC group were comparable for percentage of predicted values of Wpeak (66 ± 17% vs. 71 ± 15%, p = 0.134), HRpeak (90 ± 6% vs. 92 ± 11%, p = 0.358) and VE/VCO2-slope (105% (IQR 24%) vs. 109% (IQR 14%), p = 0.98). The reached percentage of predicted VO2peak was lower for the ILT group compared to the ECC group (69 ± 14% vs. 76 ± 16%, p = 0.046).
Conclusion: CPET parameters Wpeak, VO2peak and HRpeak and VE/VCO2-slope are impaired in contemporary Fontan patients. The results are comparable for ILT and ECC techniques concerning most values. However the percentage of predicted VO2peak was lower in patients with an intra-atrial lateral tunnel. These results show that reduced exercise capacity remains an important issue in Fontan patients. The ECC modification might have a slightly more favorable outcome for exercise capacity at medium term follow up.

O1-6
Direct Intrapulmonary Injection of Iloprost in the Pulmonary Artery for Testing Vasoreagibility in Pulmonary Hypertension

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Introduction: Diagnosis of severity of pulmonary hypertension (PH) requires right heart catheterization; gold standard for vasoreagibilty testing is the use of pulmonary vasodilators. We present results of our test protocol and offer the alternative of direct intrapulomary application of Iloprost.
Methods: Retrospective analysis of 33 patients (23 female, mean age 15.1 ± 11.9 years, mean BSA 1.7 ± 2.8 m^2) with PH who required invasive vasoreactivity testing between 2007 and 2013. Our standard protocol consisted of baseline measurements, followed by application of 41 oxygen, then added 40 ppm iNO and thereafter applied Iloprost directly into the pulmonary artery (PA). All investigations were performed under deep conscious sedation. Underlying conditions were congenital shunt lesions (n = 22), patients after corrective surgery (n = 8) or suspected primary PH (n = 3). In 24 patients testing was done with oxygen, iNO and intrapulmonary Iloprost. 4 patients did not receive iNO, and 5 no oxygen because of oxygen dependency. Standard measurements were obtained and pulmonary vascular resistance (PVR) and cardiac output (CO) were calculated.
Results: The PVR at baseline was 15.2 ± 9.4 WU*m^2 and could be reduced by oxygen to 13.4 ± 10.0 WU*m^2 (p = 0.002), by iNO to 8.0 ± 3.6 WU*m^2 (p < 0.001) and by Iloprost to 6.8 ± 7.1 WU*m^2 (p < 0.001). The comparison between Iloprost and iNO and oxygen as well as oxygen and iNO was also statistically significant (p < 0.001). The CO rose from 4.4 ± 2.3 l/min at baseline to 4.8 ± 2.6 l/min after oxygen (p = 0.814), to 5.2 ± 2.0 l/min after iNO (p = 0.476) and to 6.4 ± 3.0 l/min after intrapulmonary Iloprost (p < 0.001). The comparison between CO after oxygen and iNO application did not gain any significance (p = 0.495) but the comparison of CO after Iloprost and iNO or oxygen (p < 0.001) did.
Conclusion: Iloprost did effectively reduce the RVP and raise the CO in our patients with pulmonary hypertension even after maximal application of oxygen and iNO. The intravenous application of Iloprost in the PA is certainly as effective as the inhalative application in testing vasoreactivity of the pulmonary circulation and it is much easier to use.

O1-7
Are deficits in emotion processing part of the neurodevelopmental morbidities after transposition of the great arteries (TGA)?

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Introduction: Children with TGA may present neurodevelopmental morbidities including deficits in executive functioning, social cognition and a higher prevalence of subtle autistic-related traits than expected in the general population. Social cognition is a multifaceted neurocognitive ability including the understanding of mental states and the capacity to decode emotions and affective states. No study to date has explored emotion processing outcomes after TGA in spite of its crucial importance in psychosocial and emotional adaptation. The objective of this study is to investigate the different components of emotion recognition and comprehension as part of the social cognition outcomes in children with TGA and to identify potential neonatal medical predictive factors.
Methods: Thirty-eight children (mean age = 7y 4mo) with corrected TGA with or without a ventricular septal defect (VSD) were compared to a control group on standardized neuropsychological assessments of three core aspects of social cognition: the recognition of facial emotional expressions by perceptual identification or by verbal labeling, the understanding other's emotions of increasing complexity in typical social contexts and the advanced understanding of complex affective states. IQ and demographic variables were controlled. Pre-, intra-, and post-operative variables were examined.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Results: All children with TGA had normal IQ scores and did not differ from controls in parental socio-economic status and educational levels (ps $>$ 0.05). No significant differences between the groups were found on the perceptual identification of facial emotion expressions ($p > 0.05$). However, the group with TGA showed significantly worse scores on the verbal labeling of facial emotional expressions ($p = 0.02$), on understanding of age-appropriate emotions in typical social contexts ($p = 0.002$) and on the comprehension of complex mental and affective states ($p = 0.03$; $p = 0.01$). Multivariate regression analyses demonstrated that the presence of a VSD ($p = 0.02$; $R^2 = 0.32$), a younger age at the arterial switch operation ($p = 0.03$; $R^2 = 0.56$) and a prenatal diagnosis of TGA ($p = 0.02$) were significantly associated with better outcomes.
Conclusions: Despite normal intelligence scores, children with TGA may exhibit specific neurocognitive deficits including impaired emotion processing abilities. Preoperative factors associated to reduced neonatal morbidity may have a long term impact on neurodevelopment. Hypotheses concerning the neurological patterns of anomalies resulting in such alterations are discussed.

O1-8

Illness narratives from school-age children with congenital heart defects: children's perspectives on life experiences and coping strategies related to severity of cardiac diagnosis

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Introduction: Understanding children's own views about living with congenital heart defects (CHDs) is key to supporting their successful participation in daily life, school and peer relationships. In this paper we explored children's narratives about living with CHDs and compared qualitatively the views of children with defects that are often surgically corrected during infancy (less severe) and those with complex, palliated defects (more severe).
Methods: A UK-wide cohort of school-age children with CHDs, aged 10–14 years, and their parents completed postal questionnaires about children's health and quality of life; children also answered an open question about what it is like to be a child with 'a heart problem'. Through collaborative thematic content analysis using NVivo software, we explored how children characterise their experiences of living with a CHD and coping strategies they have developed, as well as differences in response by cardiac severity group.
Results: Written narratives and/or drawings were returned by 436 children (248 boys [56.9%]; mean age 12.1 years [SD 1.0; range 10–14]); 313 had less severe and 123 had more severe cardiac diagnoses. Key themes included medical care, positive and negative emotions, social interaction and coping strategies. Within these themes, there were important qualitative differences in the way children in the two severity groups characterised their CHD and some coping strategies were clearly emphasised by one severity group. A key issue for many children was managing revealing their scar in order to minimise the risk of social exclusion.
Conclusion: Children's perceptions of living with CHDs were only partly related to the severity of their cardiac diagnosis, and associated physical limitations. Parents, healthcare staff and teachers responsible for children with CHDs should be aware of and understand the child's perspective of their condition in order to actively support the coping strategies they develop. Managing information-sharing about their condition may be an important step for school-age children with a CHD in taking responsibility for their future health. We have shown that children's written narratives, collected through a postal survey, can be a helpful, innovative child-centred method for capturing these perspectives and adaptive strategies.
Funding: British Heart Foundation, Medical Research Council.

O2-1

Characterization of right ventricular activation in patients with Tetralogy of Fallot and right bundle branch block

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Introduction: A prolonged QRS duration (QRSd) in patients with tetralogy of Fallot (TOF) is considered as a risk factor for sudden death and a possible target for CRT. It has been suggested that QRSd mainly reflects abnormalities of the RV outflow tract (RVOT) rather than the RV body itself. We characterized the RV activation pattern in these patients to better understand the electrophysiological background for arrhythmias and RV dysfunction.
Methods: RV activation sequence was studied in 28 adults (QRSd 153 ± 21ms) referred for either catheter ablation or pulmonary valve replacement late after ToF repair, with application of 3D contact mapping (201 ± 32 sites/patient; Carto 3 - Biosense Webster).
Results: The patients showed a single RV breakthrough in the septum (mid-septal in 79%, septo-basal in 14% and apico-septal in 7%) recorded 28 ± 23 ms after the beginning of the earliest QRS complex traducing a slow left-to-right transseptal activation time.
Two distinct patterns of activation proceeded slowly from the breakthrough site: 1) the first one from the septum to the outflow tract and to the basal portion of the RV free wall; we observed, at the level of the RVOT, presence of fragmented, low voltage, or multiphasic components corresponding to the surgical scars and patches; 2) the second one, from the breakthrough site to the apex and the mid RV free wall with prolonged conduction velocities likely as a result of cell-to-cell conduction and/or anisotropic conduction.
Despite major abnormal electrical signals detected in the RVOT, this area did not influence the localization of the last activated segments. Indeed, the RV activation ended at the mid portion of the RV free wall after a mean RV activation time of 127 ± 20 ms. This RV activation time was correlated to QRSd ($r = 0.72$; $p < 0.001$).
Conclusion: In adults with repaired TOF, we observed slow conduction velocities in the whole RV with a similar pattern of activation than observed in the LV of patients with left bundle branch block. This may have important implications in the understanding of the risk of sudden death, the decision to implant a CRT device and the determination of the optimal pacing sites.

O2-2

Changes in conduction and restriction of expression patterns during sinoatrial and atrioventricular node development: role of ROCK signalling

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Background: Studying the development of the cardiac conduction system can provide useful insight into mechanisms behind specific arrhythmias. Multiple embryonic signalling pathways are re-activated during cardiovascular disease in the adult heart. This could also be the case for arrhythmia prone areas. The RhoA-ROCK signalling pathway is involved in cellular processes like migration, proliferation and myogenic differentiation. RhoA is expressed within the developing cardiac conduction system in chick and disruption in adult mice results in arrhythmias. In this study we provide insight into the electrophysiological and morphological changes within the sinoatrial (SAN) and atrioventricular (AVN) nodal areas during avian heart development and the role of ROCK signalling.
Methods: Electrophysiological changes of developing hearts were assessed in vivo by ex ovo local electrophysiological recordings and in vitro by multi-electrode arrays. The morphology of the developing SAN and AVN areas was studied by expression patterns of the cardiac markers cTnI, Nkx2.5, the gap junction protein Cx43, and the cation channel HCN4. The role of ROCK signalling was studied using chemical inhibition (Y-27632).
Results: Early in development the entire sinus venosus myocardium, including the right-sided SAN and a transient left-sided SAN, has the potential to generate the first electrical activity and a specific expression pattern showing cTnI and Hcn4 expression but no Nkx2.5. Conduction properties differ depending on the initial activation site. Eventually, the electrical potential and cTnI and Hcn4 expression become restricted to the right-sided SAN. We observed a significant increase in heart rate and atrioventricular delay during development, indicative of maturation of the cardiac conduction system. In hearts with diminished ROCK signalling, atrioventricular conduction time changed as expected during development. However, the heart rate did not increase and atrial activation pattern rates were disrupted, suggestive of an immature SAN state.
Conclusions: Significant electrophysiological and morphological remodelling occurs during SAN and AVN development. ROCK signalling may play a role in mechanisms regulating changes in heart rate during development.

O2-3
First experience with a new totally subcutaneous ICD (Cameron Health) in five high risk patients with complex congenital heart disease (CHD)

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Introduction: Implantable cardioverter-defibrillator (ICD) therapy for prevention of sudden cardiac death has been increasingly adopted in the CHD population. In this group, conventional transvenous systems are often not applicable due to specific anatomic situations. Therefore we describe our preliminary experience with a new totally subcutaneous ICD (S-ICD, Cameron Health, USA) in a high risk patients group.
Patients and clinical outcome: In 5 patients (8.9–51.2 years) the S-ICD system (69 cc, 145 gram) was implanted for secondary prevention of sudden death. Patient weights ranged from 34–130 kg. 3 patients with an intracardiac right-to-left-shunt had a contraindication to transvenous lead placement: two with Eisenmenger syndrome and one with pulmonary atresia and VSD. In one patient with Ebstein's anomaly, transvalvular lead passage was not appropriate. In the youngest patient (8.9 y) with severe ventricular dysfunction, transvenous access was limited by a Glenn shunt. No patient had an antibradycardiac pacing indication. 3 procedures were performed with general anaesthesia and two with conscious sedation. In 4 patients the device was submuscular and in one (130 kg) subcutaneous. Post implantation DFT testing showed effective ICD function. Good cosmetic results without patient discomfort were achieved in all. During a median follow up of 11.9 months 4 of the 5 patients did not experience any shocks. In the patient with Ebstein's anomaly with complete right bundle branch block (RBBB), two inappropriate shocks occurred during exercise; this was due to a change of T-wave morphology at higher heart rates leading to 'double counting'.
Conclusions: The new S-ICD is a good choice for complex CHD patients, in whom transvenous lead placement is not applicable. The minimally invasive approach avoids epicardial lead placement via thoracotomy - a major advantage in patients with high preoperative risk factors. Unfortunately the S-ICD has no antibradycardiac pacing option, which substantially limits its use in CHD patients. Despite the bulky size of the device, which restricts the use to patients above 30 kg, good cosmetic results can be achieved if the device is placed submuscularly. For pre-implantation ECG screening, exercise testing should be considered to rule out T-wave oversensing at increased heart rates especially with RBBB.

O2-4
Intraoperative Treatment of Arrhythmias in Adult Patients With Congenital Heart Disease

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Introduction: Supraventricular and ventricular arrhythmias are a major cause of morbidity and mortality in adult patients with congenital heart disease (CHD). Intraoperative ablation offers an alternative for patients that failed ablation procedures or are requiring concomitant surgical intervention. We present the results of our experience with the surgical treatment of arrhythmias in adults with CHD undergoing elective cardiac surgery.
Methods: Between September 2002 and December 2012, 90 consecutive patients with CHD, mean age of 39 years (range 16–72), underwent intraoperative ablation during cardiac surgery for pulmonary implantation (37/90, 41.1%), atrial septal defect closure (22/90, 24.4%), Fontan procedure (18/90, 20%), partial AV canal (4/90, 4.4%), Ebstein or tricuspid valve dysplasia (4/90, 4.4%), scimitar syndrome (2/90, 2.2%), cor triatriatum (1/90, 1.1%), congenitally corrected TGA (1/90, 1.1%), or mitral valve regurgitation (1/90, 1.1%). Significant clinical predictors of arrhythmia recurrence were determined by univariate analysis.
Results: In the study period we performed 44 right-sided Maze procedures, 27 Cox-Maze III procedures and 19 right ventricular ablations. The hospital mortality rate was 5.5% (5 patients) from

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

causes unrelated to ablation. In 85 survivors, the ablation was effective immediately. Over an average follow-up period of 58 months (2–96 months), 3 (3.4%) late deaths occurred: 2 after a Cox-Maze III and 1 after a right-sided Maze procedure. Arrhythmias recurred in 9 (21%) patients after right-sided Maze ablation, 6 (22%) patients after Cox-Maze III procedure, and 7 (37%) patients after right ventricular ablation. Fifteen patients were controlled with medical therapy, 4 patients underwent catheter ablation of the arrhythmia, 1 patient required a permanent pacemaker, and 2 patients had an intracardiac defibrillator implanted. Univariate analysis demonstrated that duration of arrhythmia prior to surgery ($p = 0.003$), right-sided Maze ablation for atrial fibrillation ($p = 0.001$), and atrial fibrillation in patients with tetralogy of Fallot ($p = 0.001$) are risk factor for arrhythmia recurrence.
Conclusions: Intraoperative treatment of unresponsive arrhythmia in adults with CHD is a safe and effective procedure. Freedom from arrhythmias recurrence is 74% after 8 years of follow-up. This procedure should be taken into consideration when transcatheter ablation fails or when elective cardiac surgery is planned.

O2-6
Permanent left ventricular apical cardiac pacing in children: long-term follow-up

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Introduction: To evaluate long-term results in paediatric patients with permanent epicardial pacing from the apex of the systemic left ventricle (LV).
Methods: All consecutive pts <18 yrs of age (N = 50, period 2004–2012) undergoing permanent epicardial pacing from the apex of the systemic LV (median age 1.9, inter-quartile range [IQR] 1.7–4.4 yrs, median weight 12.1, IQR 5.6–15.8 kg) were retrospectively studied. Atrioventricular block was the indication for pacing in 44/50, congenital heart disease was present in 19/50, single-chamber ventricular pacing in 29/50 pts. LV systolic function was evaluated using echocardiography before pacemaker implantation and at last follow-up (median pacing duration of 1.9, IQR 0.5–6.4 yrs) in 39/50 pts with >90% of ventricular pacing.

Parameter	LVEDD z-score mean (SD)	SF [%] mean (SD)	EF [%] mean (SD)
Before cardiac pacing	+0,68 (2,12)	39 (7)	64 (9)
End of follow up	+0,61 (1,38)	37 (6)	62 (7)
P <	NS	NS	NS

LVEDD = left ventricular end-diastolic dimension, SF = shortening fraction, EF = ejection fraction

Results: A subxiphoid approach to the LV apex was possible up to the weight of 34 kg. Four surgical revisions were performed in 3/50 pts (6%). Probability of absence of pacing system revision was 86.1% at 5 yrs after implantation. Battery replacement was necessary in 12/50 pts (24%) with the probability of freedom from battery depletion 84.5% at 5 yrs after implantation. Two of the 50 pts (4%) died during follow-up from causes unrelated to pacing. None of the pts showed signs of pacing-associated LV failure. There was no change in LV size or function during follow-up (Table).
Conclusions: Permanent epicardial pacing from the apex of the systemic LV is associated with a low risk of pacing system revisions, favourable battery longevity and complete preservation of LV function.
Supported by the project (Ministry of Health, Czech Republic) for conceptual development of research organization 00064203 (University Hospital Motol, Prague, Czech Republic).

O2-7
Presentation and follow up of Brugada Syndrome in children

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Background: Brugada syndrome (BrS) is a congenital channelopathy that usually presents in adulthood. Childhood characteristics are less well-known, impetus for this report.
Methods: Data were collected retrospectively from 1996 and prospectively from 2006 by the Brugada team of the University Hospital of Brussels. Patients aged ≤16 years with documentation of spontaneous or drug induced coved type 1 Brugada ECG pattern were included.
Results: Inclusion criteria were met for 43 patients. Median follow up was 2 1/2 years (1 month to 172 months). Six patients were lost from follow up.
Seventeen patients were symptomatic at presentation (M 12/ F 6). Median age was 10 years (7 months to 16 years). Presenting symptoms were: syncope (13 patients - six having an associated conduction disorder) and (aborted) sudden cardiac death (4). Abrupt syncope occurred in 10 cases. In the remaining 3, cause was less clear and implantable cardioverter defibrillator (ICD) implantation was recommended on individual basis, considering the familial background and the repetitive character of the syncope. The circumstances at the occurrence of the symptoms were extremely variable. An ICD was implanted in 15 patients. Appropriate shocks were administered in three patients. Three patients received inappropriate shocks.
Screening of asymptomatic family members revealed 27 patients with BrS. Five were subsequently symptomatic. Three patients had vasovagal syncope so no ICD was recommended. Two patients were referred for ICD implantation.
Ajmaline testing precipitated ventricular fibrillation in 3 patients, requiring CPR.
An electrophysiological study was performed in 28 patients of whom 17 were asymptomatic family members. VT was non inducible in any of the asymptomatic patients, although inducible in 2 out of eleven symptomatic patients.
Conclusion: Symptomatic patients were mostly ≤12-year-old boys. Indication for ICD implantation may be tasking, as most frequent presentation was syncope. Fever did not stand out as a precipitating factor. Ajmaline infusion is potentially dangerous and should be reserved for academic settings. 1/3 was previously diagnosed with atrial conduction disturbances, such as sick sinus syndrome or atrial standstill, suggesting a common denominator. Clinical follow up of asymptomatic family members is useful.

O2-8
Defining Cardiac Ion Channelopathies in Sudden Infant Death Syndrome: A Systematic Review

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Introduction (or Basis or Objectives): Sudden Infant Death Syndrome (SIDS) is the third leading cause of death in infancy. Cardiac ion channelopathies have been implicated in a proportion of the deaths based on retrospective DNA analysis in case reports and SIDS registry material. Our aim was to collate and accurately define the reported contribution of lethal ion channelopathies in SIDS victims that have been robustly characterized and functionally confirmed.
Methods: With a formally constructed search algorithm, we reviewed the National Library of Medicine's MEDLINE database for all putative cardiac ion channelopathy mutations associated with a SIDS death. 614 abstracts were screened for relevance with 128 full-text articles pulled for review. Fourteen original studies that met full inclusion criteria were selected for quantitative synthesis.

Table. aggregate prevalence of functionally significant cardiac ion channelopathies in SIDS victims

Gene/protein	aggregate number screened	averaged prevalence	% of all identified channelopathies
SCN5A/Nav1.5	371	2.2% (8/369)**	17.7%
CAV3/Caveolin-3	134	2.2% (3/134)	17.7%
RYR2/RyR2	134	1.5% (2/134)	12.1%
GPD1-L/G3PD1L	228	1.3% (3/228)	10.5%
KCNQ1/KvLQT1	276	1.1% (3/275)**	8.9%
SCNβ/Navβ	292	1% (3/292)	8%
SNTA1/α1-syntrophin	292	1% (3/292)	8%
KCNH2/Kv11.1	276	0.7% (2/275)**	5.6%
KCNJ8/Kir6.1	292	0.7% (2/292)	5.6%
KCNE2/MiRP1	233	0.4% (1/233)	3.2%
GJA1/Cx43	292	0.3% (1/292)	2.4%
KCNJ2/Kir2.1	201	0%	0%
KCNE1/minK	233	0%	0%
totals:	(cohorts overlap)	12.4%	(~100%)

Results: We identified 36 SIDS victims in 6 countries from 2001–2012 who died from a functionally confirmed mutation in one of 11 cardiac ion channels, namely *SCN5A* (n = 10), *KCNH2* (n = 4), *KCNQ1* (N = 4), *CAV3* (n = 3), *SNTA1* (n = 3), *GPD1-L* (n = 3), *SCNβ* (n = 3), *KCNJ8* (n = 2), *RYR2* (n = 2), *GJA1* (n = 1), and *KCNE2* (n = 1). One SIDS victim was a compound heterozygote for a known pathological *SCN5A* mutation (S1333Y) and a novel *KCNE1* mutation (T20I; no functional data provided). A further two SIDS victims had known pathological mutations in *KCNH2* (T895M) and *SCN5A* (del AL 586–587) with rare polymorphisms in *SCN5A* (G1084S) and *CAV3* (T78M), respectively. Excluding case reports, the prevalence of specific cardiac ion channelopathies was calculated with the denominator being all SIDS victims screened. The aggregate proportion of SIDS victims with functionally confirmed cardiac ion channel mutations was 12.4%. For the averaged prevalence of a specific cardiac ion channel mutation in SIDS victims, please see table.
Conclusions: The specific ion channelopathies identified in SIDS victims differ in their prevalence to that of the general population at large, having implications on the order and priority of which ion channel genes are screened for first.

O3-1
Increased subclinical atherosclerosis in HIV-infected children and adolescents: relations with cardiovascular biomarkers, immune activation/senescence and HIV-related variables

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Background: HIV infection accelerates cardiovascular disease (CVD). The study of subclinical atherosclerosis in subjects without traditional CVD risk factors, such as children and adolescents, may help clarify the role of HIV infection, antiretroviral treatment (ART) and immune activation on the atherogenic process.
Methods: These are the definitive baseline results of the prospective CaroVIH Study. Carotid intima-media thickness (IMT) was measured with a portable echo-device (Phillips CX50) in a group of HIV-infected children and young adults and in a group of healthy subjects of similar sex and age. Cardiovascular biomarkers (hsCRP, IL-6, IL-8, MPO, VCAM, MCP-1, tPA, CD40L) in a random subgroup of 64 HIV+ and 30 HIV- subjects, and T-cell activation (CD38+HLADR+)/senescence (CD27-CD58+) in a random subgroup of 37 HIV+ and 11 HIV- subjects were determined.
Results: 300 subjects were included, 150 HIV-infected patients (97% vertical transmission, 76% on viral suppression, 97% on stable ART) and 150 healthy subjects. Mean age was 14.8 ± 4.9 years, 62% were female. Age, gender, body-mass index (BMI), smoking status, frequency of hypertension or hypercholesterolemia were similar in both groups.
IMT was thicker in HIV-infected subjects compared to healthy individuals (mm) (0.434 ± 0.025 vs 0.424 ± 0.018, respectively, $p < 0.001$). After adjustment by age, sex, BMI, smoking status, triglycerides and non-HDL cholesterol, HIV infection remained independently associated with thicker IMT (>p50 [0.42 mm], OR, 2.3; 95% CI: 1.3–4.1; $p = 0.007$). Among HIV-infected patients, in a multivariate analysis including time with detectable viral load, cumulative ART exposure, CD4 nadir, lipodystrophy, CD4 and CD8 counts, only CD4 nadir remained independently associated to increased IMT (>100 cells/mL, OR, 0.8, 95% CI, 0.7–0.9, $p = 0.033$). Regarding cardiovascular biomarkers, only t-PA and CD-40L were elevated in the HIV-infected patients ($p < 0.05$). HIV-infected subjects presented higher frequencies of activated CD4 T-cells ($p = 0.016$). Viremic patients showed higher frequencies of senescent CD8 T-cells compared to healthy subjects ($p < 0.001$) and aviremic patients ($p = 0.02$).
Conclusions: Structural changes of the vasculature present early in vertically HIV-infected subjects, as well as immune activation and senescence. These patients should be carefully monitored for the prompt detection and early treatment, in order to prevent cardiovascular disease.

O3-2
Pitfalls of the Fontan circulation during exercise; a modeling study

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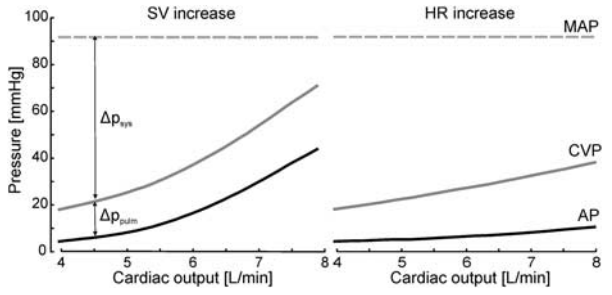
https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Background: The Fontan palliation, used as surgical repair method for many life threatening complex congenital heart diseases, creates a univentricular serial circulation. Large variation of pathologies treated with this palliation makes that current insight in cardiovascular pathophysiology of exercise limitation in these patients is limited. We used a multi-scale computational model of the heart and circulation to explore the pathophysiology of exercise limitation in Fontan patients.
Methods: The model simulates beat-to-beat dynamics of the two cardiac cavities, the valves, and the systemic and pulmonary circulations. The univentricular circulation in rest and exercise was simulated. We evaluated the extreme situations of cardiac output (CO) increase by exclusive increase of either stroke volume (SV) or heart rate (HR).
Results and Discussion: Central venous pressure (CVP) rose independent of the fact whether CO increase was due to HR or SV increase, but CVP rose more with SV increase (Figure). The large end-diastolic volumes that accompanied SV increase, required higher ventricular filling pressure, hence, higher atrial pressure (AP). Because pulmonary resistance was not allowed to change, pulmonary pressure drop and, hence, CVP had to rise to enable the increase in pulmonary flow. Consequently, systemic pressure drop reduced, implying further decrease of either systemic resistance or flow. Limited ability to decrease systemic resistance or to cope with increased CVP may underlie reduced exercise capacity in Fontan patients.
Conclusions: Our simulations suggest that limitation of exercise capacity in Fontan patients is primarily due to increase of CVP. The fact that CVP is more sensitively increased with SV than with HR may explain why bradycardia is not well tolerated.



Increase of cardiac output resulted in increase of atrial pressure (AP) and central venous pressure (CVP). The pulmonary pressure drop (Δp_{pulm}) rose to maintain pulmonary flow, resulting in a vastly decreased systemic pressure drop (Δp_{sys}).

O3-3
Pannexin-1 deficiency results in increased susceptibility for atrial fibrillation and a LQTS phenotype
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Introduction: Pannexin-1 (Panx-1), a cardiac ion channel with structural homologies to connexins, is expressed at the cell membrane and the endoplasmic reticulum. Via Panx-1 ATP and calcium ions are released. In addition, Panx-1 itself gets activated by calcium ions and constitutes the large-conductance cation channel. However, the precise cardiac function of Panx-1 is still largely unknown. We performed cardiac phenotyping of the Panx-1 deficient mouse using different in vivo electrophysiological methods and techniques from cellular and molecular biology.
Methods: Surface ECGs in sedation, telemetric ECGs in unrestrained mice and during swimming and treadmill exercise were analysed. Echocardiography and invasive electrophysiology were in vivo examinations to further elucidate the functional status of Panx-$1^{-/-}$ mice. Histochemistry, western blots and mRNA expression analysis were used for characterization cardiac Panx-1 deficiency on the molecular and cellular level.
Results: Panx-1 is stronger expressed in atria than in ventricles ($n = 5$, $p < 0.001$). Panx-1 deficiency does not alter cardiac histology (i.e. fibrosis or hypertrophy). Echocardiography showed no structural or functional abnormalities in Panx-$1^{-/-}$ mice. Panx-$1^{-/-}$ mice have a significantly higher incidence of AV-block in telemetric ECG analysis during physical activity ($p < 0.05$, $n = 10$). In vivo programmed electrical stimulation revealed no abnormalities in refractory periods and conduction. However, burst stimulation induced atrial fibrillation in all Panx-1 but not in wildtype mice ($n = 8$) with a duration of up to 9 s. Surface ECGs using Avertin or Isofluran for sedation and telemetric ECGs in awake mice showed a significant QT and rate corrected QT prolongation in Panx-$1^{-/-}$ mice compared to Panx-$1^{+/+}$ mice (Telemetry: Panx-$1^{+/+}$ QT: 44.6 ms ± 1.8 QTc 41 ms ± 3.7, $n = 6$, Panx-$1^{-/-}$ QT: 50.2 ms ± 2.1, QTc 47.3 ms ± 2.7, $n = 6$, $p < 0.005$). QTc prolongation was most pronounced at lower heart rates ($p < 0.001$, $n = 6$).
Conclusion: These results are the first evidence of an increased susceptibility of Panx-$1^{-/-}$ mice for atrial fibrillation, a higher incidence of AV-blocks during activity and a LQTS phenotype, therefore further encouraging the functional role of Panx-1 in cardiac electrophysiology. Panx-1 seems to affect automaticity and repolarization in the heart and might be an interesting target for further evaluation in patients with atrial fibrillation.

O3-4
Influence of metabolic syndrome markers upon formation of essential arterial hypertension in adolescents
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Aim: To assess the influence of components of metabolic syndrome (MS) upon formation of essential arterial hypertension (EAH) in adolescents.
Material and methods: We examined 229 adolescents at the age of 12–18 years old with EAH at different stages of its formation. The average age was 14,9 ± 2,0 years. All patients were performed 24-hour blood pressure monitoring. As a result, the following groups were formed: 1 group – patients with "white coat" hypertension – 98 pts (30.1%), 2 group – adolescents with labile arterial hypertension – 108 pts (33.1%) and 3 group – adolescents with persistent hypertension – 93 pts (28.5%). The control group consisted of 27 healthy adolescents. MS marks detection was performed according to the National Cholesterol Education Program Adult Treatment Panel III.
Results: Insuline resistance syndrome (IR) was disclosed in 2.3% of adolescents with EAH, 85.7% of whom had persistent hypertension. Only half of them had abdominal obesity. IR in the form of hyperinsulinemia and increase of insuline resistance index (IRI) of HOMA appeared in 7.4% of adolescents with EAH. Only the third part of the examined patients had overweight. The highest average values of IRI HOMA, in

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

comparison with the control group, were revealed only in the group of adolescents with persistent hypertension, and the difference of values was 1.79 (95% CI 1,01:3,59), $p = 0.049$). Carbohydrate metabolism disorder, which is the indirect indicator of IR, was disclosed in 13.1% of adolescents with EAH. Significant changes of fats in adolescents with essential AH were not disclosed. Clinically significant difference of mean values of triglyceride and very little density lipoprotein-cholesterol ($p = 0.027$ and $p = 0.027$, correspondingly) was discovered in the group with persistent hypertension in comparison with the control group. Hyperuricemia was recorded in 72,1% of patients with EAH and in 22% of adolescents from the control group without any significant correlation between two groups. Clinically significant difference of mean values of uris acid level was recorded only in the group of patients with persistent hypertension - 65,41 mmol/l (95% CI 19,89:110,94), $p = 0.0051$.

Conclusion: MS components start to form long before its clinical implication. Metabolic disorders cause AH stabilization.

O3-5

Impaired function and morphology of the sino-atrial node in a VEGF over-expression model

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Introduction: Vascular Endothelial Growth Factor-A (VEGF) is associated with congenital heart defects. VEGF120/120 mice over express VEGF. Previous studies in this model focused mainly on the outflow tract. In the current study we aimed to asses the effect of VEGF over-expression on the sino-atrial node (SAN) and inflow tract of the heart.

Methods: We performed morphological analyses and embryonic ultrasound to determine cardiac function of VEGF mutants during heart development. High frequency ultrasound was used to measure heart rate (HR) and diastolic function at embryonic day (E) 12.5, 14.5 and 17.5 in VEGF120/120 and wildtype (WT) embryos. Recordings were performed trans-abdominally in sedated (Isoflurane 1.5%) VEGF+/120 pregnant mice under stable vital parameters. The morphology of measured embryos was studied using antibodies for troponineI, Nkx2.5, Wilms tumor1, and HCN4. SAN volume estimations were performed using Carvalier's principle.

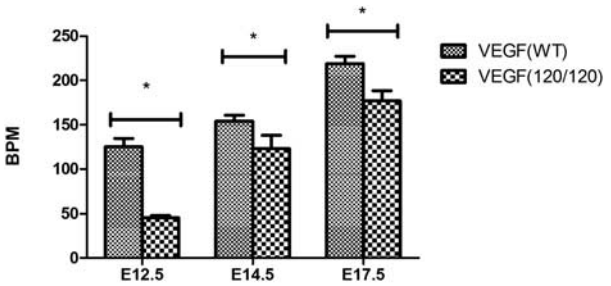

Figure 1: Significant lower heart rate in VEGF mutant versus wild type embryos.

Results: The blindly measured mean heart rate during fetal echocardiography at day E12.5, E14.5 and E17.5 was significantly lower in VEGF120/120 embryos compared to WT controls (figure 1). No irregular heart rhythms where observed during fetal echocardiography. Consistent with these data, morphometric analysis revealed a significant smaller SAN volume at E12.5 and E17.5 in mutants. The SAN of mutant embryos had an aberrant shape, with abnormal vasculature. The inflow phenotype of VEGF120/120 embryos varied between normally septated hearts and atrioventricular septal defects (AVSDs). Dysmorphic atrioventricular valves, and myocardial hypertrophy and signs of diastolic dysfunction were also observed.

Conclusion: Over-expression of VEGF results in an abnormal development of the inflow tract of the heart and function of the SAN. The role of VEGF in development of these structures requires further investigation.

O3-6

Hyperlipidemia and glucose intolerance at late maturity following neonatal hypoxia in rats

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Introduction: There is increasing evidence of early life stress leading to Metabolic syndrome at maturity. Metabolic syndrome is an aggregation of risk factors (overweight, abdominal obesity, hypertension, dyslipidaemia and glucose intolerance) that strongly correlates with cardiovascular disease. We have previously shown that neonatal hypoxia is associated with an increase in arterial blood pressure in male rats that persists to late maturity. The aim of this study was to investigate the effects of hypoxia for the first ten days of life on the lipid profile and glucose tolerance at late maturity in male rats.

Methods: Experiments were conducted on adult male Sprague-Dawley rats aged 8 months. An experimental group ($n = 8$) was raised in hypoxia ($FiO2 = 0.12$) for the first ten days of life and subsequently raised in normoxia (Neonatal Hypoxia). A second group ($n = 13$) was reared in normoxia without exposure to hypoxia (Control). At 8 months rats from each group were fasted overnight. Venous blood was obtained for measurement of triglycerides, cholesterol and HDL. A glucose tolerance test was performed (0.5g/kg glucose i.v.) with venous sampling of glucose at 1, 5, 10, 15, 20, & 30 minutes and the area under the glucose response curve calculated for each animal. Two-tailed, unpaired t-tests were performed ($p < 0.05$). All data are presented as mean ± SEM.

Results: Fasting serum triglycerides, cholesterol and HDL were significantly elevated in the 8 month old neonatally hypoxic rats compared to controls (3.01 ± 0.73 vs. 0.94 ± 0.15, 2.84 ± 0.59 vs. 1.52 ± 0.07, 1.95 ± 0.32 vs. 1.39 ± 0.06 mmol/l). The response to the glucose challenge was also significantly impaired in the neonatally hypoxic rats compared to controls (area under the curve 151 ± 18 vs. 233 ± 32).

Conclusion: Our results indicate that neonatal exposure to hypoxia in the rat is associated with hyperlipidemia and glucose intolerance at late maturity. Together with our findings of increased arterial pressure this suggests the possibility of neonatal programming of adult Metabolic syndrome by early life hypoxia. This raises the question of long-term cardiovascular risk factors incurred by hypoxemia in early life in adult survivors of cyanotic congenital heart disease unrelated to surgical repair or residual cardiac defects.

HSF Quebec & MUHC-RI

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

O3-7

Does application of mechanical stress lead to a more mature phenotype of stem cell derived cardiomyocytes?

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Introduction: Pluripotent stem cells can be differentiated to cardomyocytes (ES-CM) and are therefore an attractive cell source for cell replacement strategies. However, current knowledge on the mechanisms of cell integration and processes of physiological reconstitution as well as mechanical and electrical coupling after transplantation into the host tissue is still fragmentary. One major obstacle for a successful integration is the immaturity of the transplanted cells; ES-CMs have an immature phenotype compared to native cardiomyocytes of comparable age. There is cumulating evidence reporting beneficial effects on differentiation processes by inducing mechanical stress on myocytes. Aim of this study was therefore to investigate whether applied mechanical stress during differentiation will result in more mature phenotype of ES-CMs.
Methods: ES-CM clusters were generated from transgenic ES cells expressing puromycin resistance and enhanced green fluorescent protein (GFP) cassettes under control of a cardiac-specific promoter a-myosin heavy chain. Confluent ES cells were trypsinized and resuspended in the differential medium and maintained on a shaker for 2 days inititating the differentation to cardiomyocytes. Mechanical stress application was performed by a shockwave inducer (swiss dolorcast). Shock waves were applied at day 12,14 and 16 of differentiation with a frequency of 2 Hz at 1.2 bar for a duration of 1000 impulses. Cell survival was assessed by propium iodid staining. Size and percentage of cardiomyocytes (GFP+ cells) was measured by FACS analysis. Beating rate of ES-CM clusters were recorded before and after treatment.
Results: Treatment of ES-CM clusters resulted in a higher yield of cardiomyocytes (90.5% ± 5.86, n = 16) compared to untreated ES-CM clusters (79.48% ± 6.98, n = 13). Treatment was not associated with a higher percentage of cell death. Beating rates did not differ between the groups. FACS analysis showed that the treatment groups consists of bigger cardiomyocytes with a denser granulation.
Conclusion: Our results indicate that shockwave application results in a higher yield of cardiomyocytes and that the mechanical is not associated with a higher cell death. Graphical display of the FACS analysis suggests that mechanical stress leads to bigger cardiomyocytes with more pronounced granulations. This could indicate a more mature phenotype, nevertheless additional experiments are required to confirm a more pronounced maturity.

O3-8

Ultrasound assessment of genetic mutated mouse models, Vegf+/120 a model for Tetralogy of Fallot

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Background: Although hemodynamics play an important role in cardiogenesis and abnormal blood flow through the developing heart might result in congenital heart disease, up till now studies in embryos/fetuses of genetic mutated mouse models mainly focused on the cardiac morphology and the availability of hemodynamic data is limited. High-frequency ultrasound is a promising technique for assessment of morphology as well as hemodynamics along mouse heart development. Here we performed ultrasound experiments in fetuses of the Vegf+/120 mouse model, that has a cardiac phenotype resembling Tetralogy of Fallot.
Methods: A timed breeding program was initiated with Vegf+/120 mice, resulting in fetuses of following genotypes: Vegf+/+ (wildtype), Vegf+/120 (heterozygous) both showing normal heart development and Vegf 120/120 (mutant, Tetralogy of Fallot like phenotype). Trans-abdominal high-frequency ultrasound assessments were performed in fetuses of sedated (Isoflurane 1.5%) pregnant Vegf+/120 mice under stable vital parameters (heart and respiratory rate and body temperature). Hemodynamic and morphological assessments were performed in fetuses at 14.5 and 17.5 days post conception (dpc). Following the ultrasound experiments of 17.5 dpc the fetal hearts were harvested for genotyping and ex-vivo immunohistochemical studies, including 3-dimensional reconstructions.
Results: Whereas the heart rate in wildtype and Vegf+/120 increases along fetal development that of Vegf120/120 fetuses decreased significantly. Morphological assessment could be performed of the different cardiac segments. The presence of structural heart malformations like ventricular septal defects during the ultrasound experiments in Vegf120/120 were confirmed by ex-vivo morphological studies at 17.5 dpc. Furthermore, reliable pulsed-wave Doppler flow recordings could be performed across the developing mitral, tricuspid, aortic and pulmonary valves. An abnormal course of Doppler flow patterns were observed in Vegf120/120, for example a decreasing trend of the E/A-ratio across the developing tricuspid valve.
Conclusion: High-frequency ultrasound is a useful method for longitudinal assessment of (ab-) normal heart development in fetuses of a genetic mutated mouse models. In the future, this technique will help to further unravel the role of hemodynamics in the development of congenital heart disease.

Financial support by the Dutch Heart Foundation grant no: 2009T070

O4-1

Percutaneous tricuspid valve implantation – Initial two centre experience

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Introduction: Tricuspid valve replacement is indicated for severe tricuspid valve dysfunction as the last therapeutical option, if a surgical valve plasty is not feasible. A biological prosthesis in TrV position has a limited durability. Percutaneous tricuspid valve

implantation (PTVI) may be an alternative to repeated surgical valve replacement.
Methods and Results: Since July 2008 ten patients (female 5, diagnosis: Ebsteins's anomaly 3, functionally univentricular heart 3, other 4) with biological valve dysfunction in tricuspid position were treated by PTVI at two centres. Median patient age was 31.2 years (5–71 years) and median weight was 54.3 kg (17.7–97.5 kg). Prevailing severe regurgitation of the bioprosthesis was the indication for treatment in 10/11 procedures. All eleven valves were delivered successfully (Medtronic Melody valve 9, Edwards 26 mm Sapien 1, Edwards 29 Sapien 1) in these 10 pts and early after the intervention only mild residual regurgitation was assessed by TEE in all. Pre-stenting was done in 8/11 procedures. There were no major periprocedural complications. One femoral venous disruption occurred in the smallest patient (17.7 kg) without clinical relevance. One early recurrent tricuspid regurgitation (after 18 months) led to surgical replacement and subsequent repeated PTVI (6 motnhs after repeated surgery). Follow-up ranges from 1 month to $3^1/_2$ years.
Conclusion: PTVI can be done with a low periprocedural complication rate. On short to medium term follow-up valve function is acceptable in 10/11 implanted valves. PTVI may be a good alternative to surgical valve replacement.

O4-2
Cardiac catheterization in patients with congenital heart disease on ECMO: Aimless action or helpful tool? – A review of 31 cases
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Introduction: Hemodynamic instability after surgery for congenital heart disease (CHD) may be treated symptomatically by ECMO. Immediate diagnostic on ECMO is warranted to evaluate the cause of hemodynamic instability and to initiate timely surgical or interventional treatment. Cardiac catheterization (CC) may detect otherwise not identifiable causes of hemodynamic instability and offers the possibility of simultaneous intervention. We aimed at evaluating the diagnostic and therapeutic value of CC in patients (pts.) with CHD on ECMO and to assess the mortality and morbidity related to CC.
Methods: Data of 68 surgical patients, and 6 non-surgical pts., who were treated with ECMO between 2006 and 2012 were analyzed. Median age was 8 months (range: 5 days–51 years), median weight was 4.9 kg (range: 2.0–126 kg).
Results: Indications for ECMO were failure to wean from cardio-pulmonary bypass (n = 29), low cardiac output syndrome on the ICU (n = 18), cyanosis (n = 9), and ongoing cardio-pulmonary resuscitation (n = 18). 33 CC on ECMO were performed on 31 pts. There was no intraprocedural mortality, no major bleeding, and no complication related to the CC. All interventions were performed successfully. In 13 pts. CC was followed by medical treatment only. Eight pts. had interventions during CC. Four pts. received an intervention and a reoperation. Six pts. underwent reoperations. The following procedures were performed on ECMO: Stenting of the pulmonary artery (n = 3), of an aortopulmonary shunt (n = 1), of the superior caval vein (n = 1), of a coronary artery (n = 3), local lysis in a coronary artery (n = 1), in a pulmonary artery (n = 1), in the A. mesenterica superior (n = 1), ballonatrioseptostomy (n = 2), angioplasty of an aortopulmonary shunt (n = 1). Three pts. underwent >1 intervention. Hospital survival of pts. with CC on ECMO was 35% vs. 43% of all pts. on ECMO ($p < 0.05$).
Conclusion: CC and interventions can be performed safely and effectively on ECMO. The findings revealed by CC led to interventional, surgical, or medical treatment in 32% of the pts. Cardiac catheterization is an important diagnostic and sometimes therapeutic tool, especially in these critically ill patients. However, hospital survival of patients with CC on ECMO is lower compared to all pts. on ECMO.

O4-3
Incidence, Diagnosis and Outcomes of Coronary Artery Compression During Percutaneous Pulmonary Valve Implantation
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Introduction: Coronary compression may occur during percutaneous pulmonary valve implantation and is potentially life threatening when undiagnosed before right-ventricular outflow tract stenting. We sought to evaluate its incidence, diagnosis and outcome.
Methods: All consecutive patients who underwent transcatheter right-ventricular outflow tract treatment from May 2008 to December 2011 in 4 institutions were studied. Baseline demographics, diagnosis and outcomes of coronary compression were reviewed with analysis of risk factors.
Results: Coronary compression occurred in 6 out of 100 patients (6%) at a median age of 24 (13 to 49) years, with right-ventricular outflow tract conduit stenosis as the primary lesion in all cases. The initial congenital heart disease was pulmonary atresia-ventricular septal defect (n = 3), complex transposition of the great arteries (n = 2) and critical aortic stenosis status-post Ross operation (n = 1). The right-ventricular outflow tract initial median conduit diameter at surgical implantation was 23 (17 to 24) mm and conduit types were homograft (n = 3), bioprothesis (n = 2) and a pericardial patch (n = 1). Coronary compression was diagnosed by coronary angiogram during balloon dilation of the right-ventricular outflow tract in all cases whereas it was suspected on pre-procedure computed tomography in only 2 cases. Compression occurred on the left main coronary artery in 5 cases and on a single coronary artery in one patient. No risk factor was found but there was a significantly higher incidence of coronary compression in one of the 4th institutions ($p = 0.04$). Coronary compression was well-tolerated and resolved after the balloon was deflated in all the cases. No patients underwent right-ventricular outflow tract stenting or percutaneous pulmonary valve implantation. Surgical conduit replacement was electively performed in 3 cases. Two patients with moderate residual right-ventricular outflow tract stenosis are followed. One patient with encephalopathy and respiratory insufficiency died 9 months after catheterization.
Conclusions: Coronary compression is efficiently diagnosed by coronary angiogram during balloon dilation in a small proportion of patients undergoing transcatheter interventions on right-ventricular outflow tract. Diagnosis by pre procedure computed

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

tomography is not accurate. No specific risk factors exist. Surgical conduit replacement is indicated when balloon dilation fails to improve the right-ventricular outflow tract obstruction.

O4-4
Transcatheter Fenestration in Fontan Failure: single center experience?

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Background: Total Cavo-Pulmonary Connection (TCPC) is the final step of the palliative separation of the circulations in children with a univentricular heart. Fenestration between the systemic venous conduit and the common atrium might be a life saving approach in acute or chronic failing Fontan or even necessary for electrophysiological treatment.
Methods: Since 2005, 23 percutaneous catheter-based fenestrations were performed in 20 Fontan patients; 18 patients had an extra cardiac conduit, 2 a lateral tunnel. The perforation of the conduit/lateral tunnel patch was performed with a Brockenbrough technique; one closed surgical fenestration was re-opened by HF-(Byliss), and two with a stiff wire technique. After crossing the conduit or patch, gradual balloon dilatation was performed. Then a 6/8F long sheath was advanced to the atrium during deflation of a 6–8 mm balloon. Hand-crimped or pre-mounted stents (Genesis, Valeo, Formula, Jo-Med) were placed in the newly created fenestration in a diabolo shape in all, but one.
Results: A fenestration of a TCPC tunnel was newly created in 12 patients; re-opening of a surgical or catheter-based closed fenestration was successfully performed in 8 (+3) patients. In 14 patients, catheterizations were performed as a high urgency procedure because of and still during clinical and hemodynamic instability in order to avoid or to treat an already failing Fontan. After stent placement mean arterial oxygen saturation decreased from 92,5% (n = 22; SD 5,5%) to 84,2% (n = 21; SD 5,5%). Transcatheter fenestration approach was performed with a median fluoroscopy time of 17.3 minutes. Ten of the 14 acute treated patients improved immediately within one week. In all, pleural effusions and ascites diminished corresponding with decreasing central venous and pulmonary artery pressures. In three patients clinical improvement took longer than a week. Serious complications, especially bleedings were not observed. In 6 patients the fenestration closed meanwhile spontaneously.
Conclusion: The percutaneous approach is a low risk procedure even in TCPC with an extra-cardiac conduit. It has a big advantage comparing high risk and complex operation.

O4-5
One year follow-up of the PREMIER multicenter registry for the Edwards SAPIEN pulmonic transcatheter heart valve

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Introduction: PREMIER is a single arm, multi-center registry assessing the safety and efficacy of the commercially available Edwards SAPIEN^{TM} Pulmonic Valve for treatment of patients with conduit failure in the right ventricular outflow tract (RVOT), or moderate to severe pulmonary regurgitation with or without stenosis. A minimum of 100 consecutive patients treated at 10 sites or more have to be included in order to complete the enrollment. One year follow-up has been completed for the first 39 patients and an interim analysis was carried out.
Methods: Edwards SAPIEN^{TM} Pulmonic 23 mm and 26 mm valves were implanted in the pulmonary position in patients with a dysfunctional RVOT. Prospective clinical and echocardiographic data are being collected annually throughout 5 years. If valves were implanted before the registry initiation, data are recorded retrospectively starting with the first commercial implant at the site.
Results: The mean patient age was 27.5 ± 12.4 years, and 38.5% (n = 15) of patients were female. A total of 22 patients (56.4%) underwent prior conduit implantation, 5 patients (12.8%) underwent the Ross procedure, 11 patients (28.2%) underwent prior pulmonary valve repair. NYHA functional class was ≥ II in 67.6% of patients, and pulmonary regurgitation was grade 3+ or 4+ in 68.6% of patients. The procedural success rate was 100%. The mean procedure and fluoroscopy time was 185.9 ± 63.6 min and 44.2 ± 29.0 min, respectively. The freedom from all cause mortality at 1 year was 100%. Of the 11 adverse events that have been reported, only one was considered serious (i.e., vascular – stenosis). No valve stent fractures have been observed. There were no re-interventions or reoperations. At 1-year, most subjects with complete echocardiographic evaluation (16/26 or 61.5%) did not have any pulmonary regurgitation and the remaining subjects (10/26 or 38.5%) had trace or mild pulmonary regurgitation.
Conclusions: This interim reports suggests that Edwards SAPIEN^{TM} can be implanted safely in the pulmonary position with very low risk and significantly improve pulmonary regurgitation at 1-year. Further evaluation and long term follow-up is going on to further validate the clinical implications of this promising treatment for patients with dysfunctional RVOT.

O4-6
Predictors and Outcomes of Right ventricular Outflow Tract Conduit Rupture during Percutaneous Pulmonary Valve Implantation: A Multicentric Study

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Background and Objectives: Conduit rupture is a complication encountered during percutaneous pulmonary valve implantation (PPVI). There is insignificant data on the risk factors for this potentially life threatening complication. We sought to evaluate the incidence, and predictors of conduit rupture during right ventricular outflow tract (RVOT) transcatheter treatment.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Methods: All consecutive patients who underwent transcatheter RVOT treatment from May 2008 to December 2011 were prospectively studied. Baseline demographics along with incidence, predictors and outcomes of conduit rupture with various transcatheter therapies were reviewed.
Results: Conduit rupture occurred in 9 out of 99 patients (9.09%). All conduit ruptures occurred universally during balloon dilatation of the RVOT. Significant risk factors included heavy calcification ($p < 0.05$, OR = 16 [1.87–357]), conduit type (homograft vs. others; $p < 0.05$, OR = 5.37 [1.1–27.39]) and conduit stenosis as the primary lesion ($p < 0.05$). Other factors such as prolonged time interval between prior surgical RVOT repair and interventions, use of high-pressure balloons, balloon diameter, and overexpansion of conduit statistically failed to show any association. All patients were managed in the cardiac catheterization laboratory. None required surgery. There were no delayed complications during a mean follow up period of 2.3 ± 0.95 (SD) years. Conduit rupture had no impact on the mid-term outcomes.
Conclusions: Conduit rupture is a serious complication noted in a small proportion of patients undergoing transcatheter interventions on RVOT. Heavy calcification, homograft conduit and stenosis as primary lesion were significant predictors for conduit rupture. Immediate diagnosis with use of targeted interventional therapies should be attempted before proceeding with PPVI to avoid urgent surgical repair.

O4-7
Transcatheter Melody Valve Implantation in Native Ventricular Outflow Tracts

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Background: Percutaneous pulmonary valve implantation is now considered feasible and safe. Native ventricular outflow tract (OFT), without surgically implanted valved conduit, is currently considered off-label positioning for the Melody valve use.
Objective: To report the extended application of percutaneous melody valve implantation in right and left native ventricular OFT.
Methods: Retrospective review of melody valve implanted (MVI) in patients with conduit-free ventricle OFT, from 3 different centres.
Results: 14 patients. Nine females and five males, age range 1–32 years (median 12 years), and weight range 9–66 kg (median 35 kg). Thirteen patients had undergone a surgical procedure involving Right Ventricle OFT. Ten had different Fallot Tetralogy variants, with previous surgical repair using a transannular patch (9) or infundibular patch (1) One patient had dTGA, repaired via Jatene procedure. One patient (Noonan Syndrome) had pulmonary stenosis (PS) resolved by surgical valvuloplasty. One patient had undergone a Kawashima procedure for correction of Double Right Ventricle OFT. Ten had severe Pulmonary Regurgitation (PR), 2 had severe PR and moderate PS, and 1 isolated severe PS. The last patient suffered severe aortic regurgitation in the context of Berlin-Heart ventricular-assistance-device waiting for transplantation. Sizing of the narrowest systolic diameter of the OFT was performed by angio (13) or with AGA sizing-balloon (1); median diameter 15.85 mm (ranged 11–21.75 mm) correlation with previous MRI measurements was $r = 0.97$. The procedure was performed via femoral vein (10), jugular vein (2), left thoracotomy and through left ventricle Berlin-Heart cannula (1), In all cases pre-stenting was performed (4 CPstents and 10 Andrastents XL). Median fluoroscopy time was 37.6 min. The implantation procedure was uneventful. ECMO was necessary in the patient with Berlin-Heart, but this patient died the day after procedure, non-procedure and non-valve related causes. During follow-up (24 h–30 months) freedom from relevant PR was 100% and from significant PR 90% (one patient had moderate stenosis with pseudoaneurysm of the pulmonary trunk, a valve-in-valve melody implantation was performed 6-months post-procedure).
Conclusion: PMVI in native ventricular OFT is feasible in selected patients. The OFT measurement accuracy with echo, angiography and MRI is mandatory.

O4-8
Prospective study of patients with percutaneous implantation of Melody valve in pulmonary position: incidence of infective endocarditis and evaluation of risks factor for infective events

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Introduction: Percutaneous pulmonary valve replacement (PPVI) is used to treat dysfunctional right ventricular outflow tract. The mid-term hemodynamic results of the Melody valve are good but there are increasing reports of infective endocarditis (IE) with some fatal outcome.
Objective: We sought to review the patients who received a Melody valve in pulmonary position from 2009 to 2012. We identified patients with IE and studied the potential risks factors.
Results: 86 patients were eligible. 5 patients had previous history of IE on the tube stented with a Melody. 5 cases of IE were identified during survey. Overall, mean age at implantation was 23,8 years. 58% were male. Mean follow-up was 25 months. Mean residual RV/PA gradient after procedure was 12 mmHg. In the group of patients with IE, mean age was 29,8 years and mean follow-up at the time of IE was 14 months, there was no history of previous IE. 4 patients were male. Mean residual RV/PA gradient was 12 mmHg. 3 presented with severe obstruction, 2 patients died within 48 hours and one needed cardiac catheterisation to relief obstruction then surgery. The two remaining patients were treated medically. Bacterial identification was possible in all cases and probable source of infection was found in four cases. When reviewing potential risks factors, we found no association with previous stent implantation or with the length of procedure. When looking at the incidence of invasive procedure done post Melody implantation (cardiac catheterisation or other medical invasive procedure) we found statistical difference between the 2 groups. There were four death during follow-up: two due septic shock, one due to biventricular heart failure in a patient who had a Melody valve endocarditis and one in a patient with biventricular failure who refused to have heart transplantation. In this review, death was associated with IE.
Conclusion: IE is a real issue with PPVI, a relatively new technology. Risk factors include male sex, post implantation invasive procedure and not previous IE. Careful follow-up of these patients is required. In case of IE, aggressive treatment should not be delayed as fatal outcome is possible.

O5-1
Aortic coarctation: a feasible prenatal diagnosis

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Introduction: Aortic coarctation may be associated with significant neonatal morbidity and mortality. Prenatal diagnosis may improve survival and reduce morbidity but it is still associated with a high rate or false negatives/ false positives.
Methods: Between 2002–2008, we carried out a retrospective review of collected echocardiography data base for all fetuses evaluated at our unit in whom a suspicion of coarctation was the cause of referral. From 01/01/2008 to 31/08/2012 the data was collected prospectively. Fetuses with abnormalities of the cardiac connections were excluded. Of the 65 fetuses who fulfilled these inclusion criteria, 4 were excluded because incomplete outcome data. All of them had a postnatal echocardiography/autopsy study. The diameters of the atrioventricular valves, pulmonary/ aortic valves and their ratio were measured. We obtained the diameter of the aortic isthmus and duct in the 3-vessel and trachea view; the isthmal to ductal diameter ratio, and their Z scored related to gestational age were calculated. Receiver operating characteristic (ROC) curves were created for all these measures. Diagnoses of ventricular septal defect (VSD), bicuspid aortic valve (BAV) and persistent left superior caval vein were identified; logistic regression (LR) was used to test their association with fetal coarctation. Aortic arch hypoplasia (transverse aortic arch z-score <2) was studied.
Results: Coarctation was confirmed in 35/61 neonates (57,3%). Mean gestational age at presentation was higher in normal fetuses as compared to fetuses with coarctation ($p < 0.005$). Good separation was found of isthmal/ductal ratio, pulmonary/aortic valve and isthmal z-score for cases with coarctation from falses positives ($p < 0.001$). ROC curves showed an excellent area under the curve (AUC) for isthmal-to-ductal ratio (0,927, IC 95% 0,86–0,99) and for isthmal z-scores (0,852, IC 95% 0,74–0,96). Stepwise LR model only included SAB and VSD and this combination, showed an excellent AUC (0,92, IC 95% 0,85–0,991). Aortic arch hypoplasia was found in 74,7% of true cases.
Conclusions: The isthmal to ductal ratio and the isthmal z-score, combined with gestational age at diagnosis and secondary cardiac defects as VSD and BAV, may improve the accuracy of fetal coarctation diagnosis and reduce false positives. The rate of aortic arch hypoplasia is high in fetal/neonatal coartaction.

O5-2
Fetal Congenital Heart Disease Associated With Maternal Gestational Diabetes

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Introduction: It is estimated that maternal type 1 diabetes mellitus infers an increased risk of congenital cardiac disease (CHD). The association with gestational diabetes (GDM) is more controversial. Metanalyses have suggested a slightly higher incidence in women with GDM compared to normal cohorts, though the risk is lower than women with pregestational (type 1/2) diabetes.
Methods: This retrospective study was undertaken to analyse the demographics and outcomes of woman with GDM referred for detailed fetal echocardiography to our tertiary centre between 1st January 2009 and 31st December 2011. Women with Type 1 or pre-existing Type 2 diabetes were excluded from the study.
Study population: 195 women with GDM were assessed in the 3 year period and form the study population.
Results: The median age of women referred with GDM was 34 years (Range 21–44 years, Mean 33.2 years). Of the 195 women, 86 (44.1%) were controlled by diet; 30 (15.4%) by oral medication; 73 (37.4%) by insulin and 6 (3.1%) by a combination of insulin and oral medication.
Of the 195 referrals, a diagnosis of definite CHD was made in 6 cases (3%). These were: atrioventricular septal defect; tricuspid atresia/VSD; double inlet left ventricle/TAPVD/Pulmonary atresia; double outlet right ventricle/TAPVD; transposition of the great arteries; ventricular septal defect. The referral reasons in the 6 cases were abnormal views of the heart (3), increased nuchal translucency (1), inadequate views (1) and fetal arrhythmia (1).
149 women had the presence/absence of extracardiac anomalies (ECA) documented in their notes. Of these documented, 11 (7.4%) had an ECA noted on the fetal anomaly scan. Of the 141 women in which the nuchal translucency (NT) was documented only 1 (0.7%) was noted to be raised in early pregnancy.
Conclusion: Our data shows that there is an increased risk of fetal congenital heart disease in women with GDM. Detailed fetal echocardiography should be considered in this group.

O5-3
Epicardium derived cells drive differentiation of the left and right ventricle in normal and TGFbeta2 mutant mice

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Introduction: In the adult heart, morphological and functional differences of right ventricle (RV) and left ventricle (LV) are apparent. We hypothesized that a different contribution to the myocardium of epicardium-derived cells (EPDCs) during development might contribute to these differences. In the current study, we aimed to assess normal and disrupted formation of the compact myocardial layer of the RV and LV.
Methods: Epicardial sheet formation and contribution of EPDCs were studied in wildtype and TGFβ2-null embryonic mice (E9.5–14.5) using expression patterns of WT-1 and a Cre-activated WT-1 reporter model.
Results: After epicardial covering of the heart tube EPDCs were observed first in the inner curvature and RV wall. At E13.5, WT-1 expressing cells were abundantly observed in the wall of both ventricles, more pronounced in the LV correlating with a significantly thicker LV compact myocardial wall as compared to the RV. In TGFβ2-null mice, formation and migration of EPDCs were diminished, although an epicardial covering was formed. Differences in RV and LV myocardial thickness as observed in wildtype, were absent in TGFβ2-null mice.
Conclusions: Spatio-temporal differences in contribution of EPDCs to RV versus LV myocardium were observed during development. Compact myocardial layer formation starts upon migration of EPDCs into the ventricles and is more pronounced in the LV. Disruption of EPDC migration results in absence of normal LV compact myocardial thickening. The observed different EPDC-myocardium interaction in the LV versus RV may explain the occurrence of lateralized cardiomyopathies as isolated LV non-compaction and can prove relevant for development of cell-and drug based therapies.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

O5-4

Prenatal diagnosis of right aortic arch: A spanish multicenter study

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Introduction: the 3-vessels trachea (3VT) view has led to a higher prenatal detection of aortic arch anomalies. Early diagnosis of right aortic arch (RAA) is important as it maybe associated with cardiac, extracardiac anomalies and chromosomopaties.
Methods: Retrospective, multicenter, echocardiographic analysis of all cases diagnosed prenatally of RAA from 1/2010–12/2011. Type of RAA and branching pattern were assessed. Different diagnosis were sought; vascular ring: RAA with anomalous left subclavian artery (ALSA) and double aortic arch (DAA), no vascular ring: RAA with mirror image branching (MIB) and RAA undetermined type (when the branching pattern was impossible to determine). Intracardiac anomalies were studied. Gestational age at diagnosis, karyotype and outcome data were assessed. Fisher's exact test was used.
Results: 73 patients had a RAA. 33 had a RAA with ALSA, 5 DAA (all with a predominance of the RAA), 28 RAA and MIB and 6 RAA and undetermined branching. Mean gestational age at diagnosis was 22 weeks. A RAA was associated to intracardiac anomalies in 32 cases (44%). There was a significant association to other heart defects in the case of MIB (78,6%, $p < 0.001$). Most fetuses in the groups of ALSA (84,8% $p < 0.001$), undetermined type (66,7%) and double aortic arch (60%) had normal hearts. Karyopype was available in 39/73 cases, 8 had anomalies, 4 were 22q11 deletions (2/33 with RAA and ALSA and 2/28 with RAA + MIB). Extracardiac malformations were detected in 6/73 cases, 66,7% of which were RAA and MIB vs 33,5% RAA with ALSA. Pregnancy was interrupted in 14 cases. The rest of on going pregnancies were assessed postnatally with a correct diagnosis in 93% of cases. 3/73 patients with a RAA were symptomatic during the first year of life, 2 of whom had a vascular ring (DAA and RAA + ALSA) and the other had a RAA and MIB. There were 2 deaths related to intracardiac anomalies.
Conclusions: Prenatal diagnosis of RAA and vascular ring is feasible. RAA and MIB is strongly associated to intracardiac anomalies and it may be associated with extracardiac and chromosomal anomalies. Most cases of RAA + ALSA and DAA are asymptomatic during the first year of life.

O5-5

Brain growth is impaired in fetuses with congenital heart disease- MR volumetric assessment of the fetal brain

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Introduction: In the current era of excellent surgical results for congenital heart disease (CHD) focus has become directed on quality of life for these children. Previous studies have shown that neurodevelopmental outcome in CHD is impaired. The mechanisms for this are complex and not well understood but there is increasing evidence that the origins of this are in fetal life. We aimed to describe the in utero brain growth in a cohort of fetuses with CHD and relate this to the circulatory abnormalities on fetal echo.
Methods: Pregnant women with a fetus with CHD were prospectively recruited in a fetal medicine centre. The congenital heart defect was phenotyped using fetal echocardiography and patients subdivided into 5 physiological groups: Left heart obstructive lesions with forward flow (1) and reversed flow (2) in the transverse arch, Right heart obstructive lesions (3), Transposition of the great arteries (4), Shunt lesions (5).
Fetal brain MRI was performed. In addition to biometric measurements SVR (snapshot to volume reconstruction) was used to construct a 3D data set from the oversampled raw data. From these 3D volumes the total brain volume and ventricular volumes were measured by manual segmentation.
12 patients had a second MRI scan in pregnancy.
Results: 31 women were recruited who had a fetus with CHD. Comparison was made with 53 normal controls. The median gestational age of the CHD cohort when analysed by the last scan in pregnancy was 28.5 and 29.3 weeks in controls. ($p = 0.83$). Cases and controls showed a similar linear increase in biparietal diameter and transcerebellar diameter with gestation.
There was a slower rate of brain growth and smaller brain volumes in fetuses with CHD ($p = 0.029$). Analysis by diagnostic subgroup showed a trend towards slower brain growth in all subgroups.
Conclusions: Fetuses with CHD have a slower rate of brain growth compared to normal controls. In addition to that previously described in hypoplastic left heart and transposition of the great arteries, this study suggests that there may also be differences in other congenital heart lesions. Further research into a broader spectrum of congenital heart defects is needed.

O5-6

Patients with prenatally diagnosed coarctation of the aorta may be at increased risk of re-coarctation

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Introduction: Prenatal diagnosis of coarctation of the aorta (CoA) has improved by comparing the aortic and ductal arch sizes and flow disturbance in three-vessel and tracheal view. While antenatal diagnosis of CoA may improve perioperative survival, little is known regarding longer term morbidity of children detected prenatally.
We compared children with antenatal (A) and postnatal (P) diagnosis of CoA undergoing surgical repair. We hypothesized that detection of CoA prenatally selects patients with more complex aortic arch anatomy that are at increased risk for re-intervention following repair.
Methods: Retrospective study of all patients undergoing surgical repair of CoA within the first year of life between 2003–2011. Patients were excluded if there were associated major cardiac abnormalities other than mild aortic stenosis, atrial or ventricular septal defect. All operative and medical records were assessed. Re-coarctation was defined as 20 mmHg difference between upper and lower limb blood pressure and diastolic tail on echocardiography.
Results: One-hundred-and-thirty-eight children met inclusion criteria, 48 (34.8%) in group A. Twenty-seven (19.6%) had pulmonary-artery-banding. There were 7 deaths (5.1%).

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Group P had a higher risk of preoperative ventilation ($p = 0.005$), lower pH ($p = 0.0024$), older age at operation ($p < 0.0001$) than group A. There was no statistical difference in mortality ($p = 0.67$).
Predictors for mortality included associated aortic valve stenosis ±ASD or VSD (HR 26.5, 95% CI 1.6–449.9, $p = 0.023$), and pulmonary-artery-banding (HR 6.03, 95% CI 1.3–27.0, $p = 0.019$). Eighteen of 138 (13.0%) required re-intervention for aortic arch obstruction. Univariable risk factors for re-intervention included prenatal diagnosis (HR 3.7, 95% CI 1.4–10.0, $p = 0.011$), age at operation ≤7 days (HR 3.9, 95% CI 1.4–11.1, $p = 0.011$), subclavian flap procedure (HR 2.9, 95% CI 1.0–8.4, $p = 0.049$) and male gender (HR 0.3, 95% CI 0.1–0.9, $p = 0.026$). Multivariable analysis showed age at operation ≤7 days (HR 2.6, 95% CI 0.8–8.8) and prenatal diagnosis (HR1.2 95% CI 0.7–7.0) had increased hazard ratio but were not statistically significant.
Conclusions: Children with postnatal diagnosis of CoA have increased morbidity at initial presentation; however an antenatal diagnosis of CoA confers increased risk for surgical re-intervention which should be reflected in prenatal counseling. These findings may reflect the more successful antenatal detection of arches with long segment hypoplasia.

O5-7
Prognosis of severe congenital heart diseases: do we overestimate the impact of prenatal diagnosis?

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Introduction: Congenital heart diseases (CHD) are the most common and serious among birth defects (neonatal incidence 0.8%). Prenatal screening is both time-consuming and costly in terms of organization. It is subject to controversy because of unproven benefits on morbidity and mortality associated with a significant impact on the continuation of the pregnancy, starting with the psychological aspect.
Method: From 2003 to 2009 we retrospectively collected all pregnancies and children aged less than 1 with a diagnosis of CHD in our tertiary center database. Our study population was then limited to serious or complex CHD: lethal cases, leading to medical termination of pregnancy (MTP), and CHD requiring surgery, interventional catheterization or hospitalization during the first year of life. Primary endpoint was 1- year mortality among alive neonates.
Results: 322 severe CHD were included. 62.1% had a prenatal diagnosis with an excellent screening predictability of the heart defect severity. We observed significant differences between prenatal (group 1) and postnatal (group 2) CHD diagnoses comparing: type of heart disease (hypoplastic left heart syndrome 7.8% vs. 0.8%, $p < 0.05$), frequency of ductal-dependent heart defect (34.3% vs. 28.7%, $p < 0.05$) and association with chromosomal abnormality or malformation syndrome (31.1% vs. 28.8%, ns). We counted 96 MTP of 200 prenatal diagnosis (48%). Among the 224 alive neonates 15.2% died before the age of 1. Mortality at 1 year was not different between both groups (16.7% vs. 13.9%, $p = 0.13$). Major prognosis morbidity variables were not significantly different in both groups (medical treatment, duration of hospitalization, neurological, respiratory, or infectious complications).
Conclusion: like in many tertiary care CHD centers, we have strongly promoted fetal diagnosis over the 2 past decades throughout southern France. In our center prenatal diagnosis of severe CHD has an impact on the decision of MTP but not on prognosis in terms of 1-year mortality and morbidity. However, the overall severity of the prenatal diagnosed group is higher than the postnatal one, which moderates this conclusion. We recommend prospective multicenter studies with assessment of neurological prognosis and quality of life of patients. These studies will be facilitated by the use of standardized CHD registries.

O5-8
Prenatal evolution of heart defects detected during 11–13+6 weeks' scan in a Referral Centre of Perinatal Cardiology

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Introduction: It is possible to evaluate fetal heart during 11–13 + 6 weeks' scan in a referral fetal cardiology centre. Early diagnosis allows further genetic testing and better planning of antenatal care. There is still little knowledge about prenatal evolution of heart defects and about limitations of first trimester diagnostics.
Methods: Between 2004–2009 in our referral centre 797 fetuses were examined between 11–13+6 w.o.g. according to international U/S examinations safety regulations. In every case a follow-up ECHO exam in the second and/or third trimester was performed. In cases of prenatal diagnosis of CHD a further genetic testing was offered.
Results: 42 fetal heart defects were diagnosed (30[72%] in the first trimester) including both complex (HLHS, AP with VSD) and simple (AVSD, VSD) cardiac lesions. In 13[31%] fetuses also a chromosomal aberrations were confirmed. There were 18 TOP, 9 IUD and 15[36%] neonates were life-born (including fetuses with trisomy 21 and trisomy 18). There was a statistically significant correlation between increased nuchal translucency and the presence of congenital heart defects, extracardiac malformations (ECM) or chromosomal abnormalities ($p < 0.001$). ECM were diagnosed in another 9 fetuses (acrania, omphalocele, megacystis, NIHF).
We observed a progression of cardiac failure in a fetus with trisomy 18, PS and VSD (live born) and a fetus with CHB and left isomerism (heterotaxy) who died in utero. However in a fetus with a large disproportion between LV and RV in the first trimester, there was a final diagnosis of CoA in neonate and a correction with good result was undertaken.
We also reported problems with diagnosis of truncal anomalies (i.e. TOF) and small ventricular defects in the first trimester. In our study group the final diagnosis was confirmed after 16. week of gestation.
Conclusions: The first trimester echocardiography is feasible in a referral prenatal cardiology centre. Because both the progression of fetal circulatory system failure and regression of cardiac symptoms were noticed there is a necessity of further cardiac evaluations throughout the pregnancy after the first trimester ECHO. There is a strong need of genetic counseling of patients due to correlations of heart defects with chromosomal aberrations.

O6-1
Liver stiffness: a new, rapid and non-invasive method of central venous pressure evaluation in patients with congenital heart disease

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Background: Transient elastography is a rapid, noninvasive and reproducible approach to assess liver fibrosis by measuring liver stiffness (LS). However, because the liver is enveloped by a capsule, any variation in parenchymal fluid content could theoretically affect LS. LS has been correlated to central venous pressure (CVP) in an animal model. We aimed to determine the correlation between LS and CVP in children and adults with congenital heart disease.
Methods: In this ongoing prospective study, all patients referred for right heart catheterization were included. Measurements of mean right atrial pressure were obtained under general anesthesia (Fi02 = 21%) using an Optitorque 5 French catheter. The patients underwent 10 LS measurements (median value taken as representive) by transient elastography (Fibroscan®, Echosens, France) within the 24 hours before catheterization. The results of LS are expressed in kilopascals (kPa).
Results: Twenty eight (mean age = 9 ± 6 years old, 64% male) and 22 adults (mean age = 34 ± 17 yo, 66% male) have been included so far. Catheterism indications were pulmonary angioplasty (n = 10), Melody valve implantation (n = 4), fenestration occlusion after a Fontan procedure (n = 2), aortic coarctation stenting (n = 2), atrial septal defect closure (n = 8) and pre operative assessement of a complex congenital heart defect (n = 24). Mean right atrial pressure was 8,2 ± 3,3 mmHg and mean LS was 8,1 ± 4,4 kPa. Correlation between LS and mean right atrial pressure was excellent for these first 50 patients ($r = 0.86$, $p < 0.001$).
Conclusion: Liver stiffness is a new, rapid and reliable method to evaluate CVP in patients with congenital heart disease. This non invasive parameter could potentially be usefull for patients in whom CVP play a key role, especially in patients with a Fontan circulation.

O6-2
The impact of age on right ventricular morphology and function late after repair of Tetralogy of Fallot: a cardiac magnetic resonance study

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Background: Cardiac magnetic resonance (CMR) is the gold standard to evaluate right ventricular (RV) hemodynamics after repair of tetralogy of Fallot (TOF). CMR markers such as severe RV dilation and biventricular dysfunction are independent predictors of death, sustained VT and heart failure. However, the relationship between patient's age at surgical correction and RV function is not fully understood. A better understanding of determinants of outcome after repair of TOF could improve prediction of adverse events and clinical management.
Methods: All patients who underwent hemodynamic evaluation by CMR late after repair of TOF in two pediatric cardiology centers, between 2011 and 2012, were included in the study. Previous surgical repair included either transannular infundibular RV outflow tract reconstruction. Surgical data (including era and details of repair) and CMR parameters (RV volumes and function, pulmonary and tricuspid regurgitation, pulmonary branches stenosis) were collected.
Results: The study enrolled 165 patients (65% males) aged 18 ± 6.4 (range 5–38) years who had undergone repair at the age of 1.4 ± 1.7 (range 0.2–12 years) years. RV end-diastolic volume indexed (RVEDVi) was correlated both with PRF ($p = 0.001$, $r = 0.526$) and RVOT obstruction and/or bilateral stenosis of pulmonary arteries ($p = 0.01$). On the other hand RVEDVi was not correlated with patient's age or time interval from surgical repair. RV ejection fraction (RVEF) decreased in relation to RV dilatation ($p = 0.01$; $r = -0.462$), as expected. There was a significant relationship between RVEF and left ventricular ejection fraction ($p < 0.001$). Older age at repair significantly affected RVEF ($p = 0.04$; $r = -0.235$), which conversely did not show any relationship with patient age or time interval from surgical repair.
Conclusion: Patient's age and time interval from surgical repair might not have a fundamental role in RV dilation, which is mostly related to PRF. In contrast RV dysfunction is significantly related to an older age at surgical repair, independently of ventricular dilation. Larger longitudinal follow-up studies are needed in order to extend our observations and define the exact role of each element in predicting the adverse outcome of patients with repaired TOF.

O6-3
Right Ventricular Systolic Function in Hypoplastic Left Heart Syndrome: A comparison of Velocity-Vector-Imaging and Magnetic Resonance Imaging

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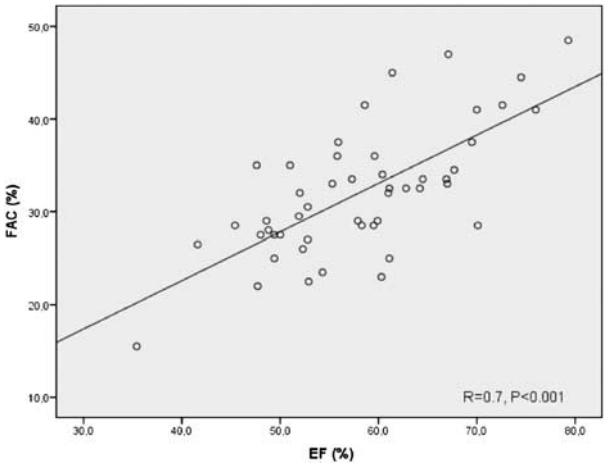
Background: Velocity-Vector-Imaging (VVI) is an echocardiographic technique based on speckle tracking which has been validated in left ventricle. It has not been critically evaluated to assess the systemic right ventricle (RV) in patients with hypoplastic left heart syndrome (HLHS). The aim of this study was to evaluate whether VVI measurements reliably reflect RV systolic function in patients with HLHS using magnetic resonance imaging (MRI) derived ejection fraction (EF) as a gold standard.
Methods: Forty nine patients with HLHS underwent transthoracic echocardiogram and cardiac MRI under the same general anesthetic as part of routine assessment between the different stages of palliative surgery, both prior to and after completion of Fontan. Global RV fractional area change (FAC), strain (S) and strain rate (SR) were analyzed from apical 4-chamber view using VVI technique (Syngo USWP 3.0, Siemens). MRI EF was calculated in the usual manner from a short axis cine stack of images.



Picture. Comparison of MRI derived EF and VVI derived FAC.

Results: All parameters measured with VVI correlated significantly with EF measured with MRI (FAC $r = 0.7$, $p < 0.001$;

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S r = 0.5, P = 0.001 and SR r = 0.5, p < 0.001). Intraobserver and interobserver reproducibility was high for all VVI-parameters (Intra/interobserver interclass coefficient for FAC 1.5%/5,7%, S 5.5%/9.6% and SR 3.0%/14.0%; respectively).
Conclusions: VVI provides a reliable tool for quantification of global RV systolic function in patients with HLHS.

O6-4
CaroVIH Study: Cardiovascular risk and ventricular function evaluation in HIV-infected children and young adults
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Background: Since the introduction of antiretroviral therapy (ART) HIV-infected patients have a higher rate of aging-related diseases, including cardiovascular disease (CVD). Studies in HIV-infected adults have evidenced the presence of premature atherosclerosis and ventricular dysfunction, due to the virus and prolonged ART. Ultrasound techniques, like carotid intima-media thickness (IMT) measurement and Speckle Tracking Echocardiography (STE) could serve as early markers of CVD.
Methods: Multicentre study including vertically HIV-infected children and young adults matched with controls by age and sex. Clinical and analytical variables were recorded. A portable echo-device (Philips CX50) was used during the complete study to measure IMT and to perform a complete echocardiography: M-Mode (shortening fraction (SF) and ejection fraction (EF)), 2D-echo, Doppler, tissue Doppler and STE.
Results: 300 subjects were included (150 HIV-infected and 150 controls). Mean age was 14.8 ± 4.9 years, 62% were female. Age, gender, body-mass index, smoking status, hypertension and hypercholesterolemia was similar in both groups. IMT was thicker in HIV-infected subjects compared to healthy individuals (mm) (0.434 ± 0.025 vs 0.424 ± 0.018, respectively, p < 0.001). A complete echocardiographic study was done in148 participants, 77cases and 71 controls: HIV-infected subjects showed a lower systolic function (SF 36,3% (SD 6,41) and EF 66,2% (SD 8,39)) versus (SF 40,6% (SD 6,88) and EF 71,3% (SD 7,51)) (p < 0.001) (all values within normal ranges). No differences were found in diastolic function and tissue Doppler examination. Ventricular torsion was greater in HIV-infected: 6,06° (SD 2,25) versus 5,49° (SD 1,97) (p = 0.09). Longitudinal strain was analyzed in 54 subjects (28 cases and 24 controls), being −21,54% in HIV-infected and −22,29% in cases (p = 0.299).
Conclusion: Since childhood, cardiovascular risk, determined by IMT, is increased in HIV-infected subjects. Also EF and SF are lower in comparison to controls. Longitudinal strain impairment, which correlates with atherosclerosis, is not present in our cohort. Ventricular torsion is increased, in accordance with a senescent myocardium. Our results suggest that at adolescent age, HIV-infected patients have a premature myocardial tissue aging and not yet affection in tissue perfusion even though higher cardiovascular risk is present.

O6-5
Diagnostic and interventional MRI catheterisation: A 10-year single centre experience
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Introduction: Hybrid fluoroscopic (X-ray) and MR imaging-guided cardiac catheterisations (XMR) provide high definition anatomy and haemodynamics in a single combined procedure whilst minimizing radiation exposure. We report a 10-year, single institution experience of XMR catheterisation, while demonstrating the evolving applications of this approach in the diagnosis and management of congenital heart disease and evolution of solely MR-guided interventions.
Methods: Retrospective review of XMR and MR-guided catheterisations between Feb 2002 and Feb 2012 at a single institution. Data collated on patient demographics, XMR procedural data and findings, impact on patient management and outcomes. Procedural complications were noted. Institutional Ethics and UK regulatory authority approval were obtained.
Results: 221 studies were performed in 196 patients. Median age and weight was 4.5 years (range 4 days to 64.7 years) and 15.4 kg (range 2.3–106 kg), respectively. 201 were combined XMR catheterisations. 20 were solely MRI-guided cardiac catheterisations, of which 7 were part of the first-in-man clinical trial on MRI-guided cardiac interventions. 57 patients had a functionally univentricular heart, of whom 18 were post-Fontan procedure. 176 patients had pulmonary vascular resistance (PVR) studies. Median total PVR was 2.4 $wu.m^2$ (0.4–66). 65 had an elevated PVR > 3 $wu.m^2$ (median 4.6 $wu.m^2$; range: 3–66 $wu.m^2$). 54 patients had a pharmacological stress study to assess cardiac output and haemodynamic responses. 151 patients went on to have an intervention (medical, catheter or surgical) based on the XMR data, at a median interval of 46 days (range 0–763 days). Accurate PVR assessment led to risk stratification and fenestrated rather than complete closure of septal defects (n = 8). 23 patients were assessed pre-liver transplant, with 12 put forward for liver transplantation. 4 of these had cardiac lesions requiring intervention and repeat XMR before being accepted for transplant. There were 2 immediate complications and 1 late complication, with no procedural deaths.
Conclusions: XMR catheterisations provide additional information on cardiac anatomy, physiology and haemodynamics to facilitate risk stratification and management planning for patients with suspicion of raised PVR and complex anatomy. Recent advances in interventional MRI have led to the performance of the first-in-man clinical trial on MR-guided percutaneous cardiac interventions.

O6-6
Isovolumic Acceleration at Rest and During Exercise in Children after Heart Transplant
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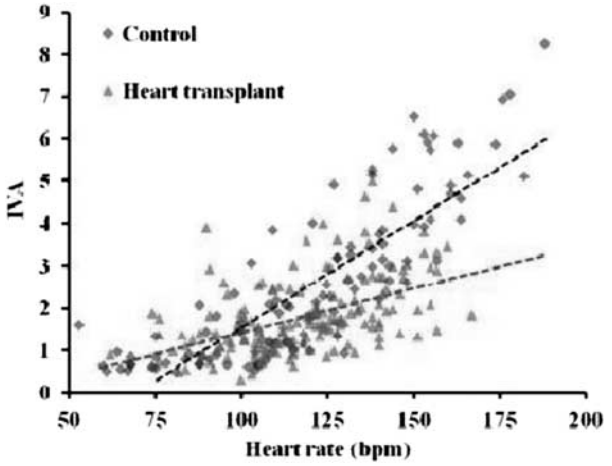
Background: Progressive graft failure is an important clinical problem in pediatric heart transplantation. The myocardial systolic and diastolic reserve during exercise has been poorly studied in the transplanted heart while it could provide important

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

information. Left ventricular (LV) myocardial acceleration during isovolumic contraction (IVA), is a TDI derived index, while correlates well with indices of myocardial contractility. It can be ued to study the force frequency relationship (FFR), which reflects the increase in contractility with increasing heart rate. The aim of the current study was to evaluate myocardial contractile response to exercise in children after HTX using semi-supine cycle ergometry stress echocardiography (SSCE).
Materials and Methods: A total of 43 pediatric HTx recipients and 24 age and gender matched controls were included. Median age at transplantation was 9 years (birth to 15 years) and median time since transplant was 4.9 years (0.5 to 15.4 years). A stepwise SSCE protocol was used. LV IVA was measured in all the subjects at rest and at incremental heart rates. FFR was constructed by plotting LV IVA against heart rate.
Results: Resting HR (mean ± SD) was higher in the HTx group than in the controls (90 ± 14 vs 72 ± 10 bpm, $p < 0.001$) and peak HR was lower in the HTx group than in CON (141 ± 12 bpm vs 165 ± 15 bpm, $p < 0.001$). LV IVA values were significantly higher at rest in the transplant group (Htx 1.22 ± 0.64 cm/s vs. 0.79 ± 0.31 cm/s $p = 0.001$) but were significantly lower at peak exercise (2.4 ± 1.02 cm/s vs. 5.2 ± 1.41 cm/s $p = <0.001$). The contractile response as studied by the FFR, was significantly blunted in HTX compared with controls, $p = <0.001$. (See figure).
Conclusions: Our data suggest a significantly different LV contractile response to exercise in HTX compared with controls when studies using the FFR. This suggests subclinical myocardial dysfunction in the grafts, which could have important prognostic implications.



O6-7
Neo-Aorta and Aortic Arch after Arterial Switch Operation for Tansposition of Great Vessels: A Morphometric and Geometric Study
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Introduction: In the current era, neonatal arterial switch operation (ASO) is the preferred surgical method for repair of transposition of great arteries (TGA). Among the late complications of ASO are aortic root dilatation and acute angulation at the level of the arch. These morphological changes might influence the patterns and severity of aortic arteriosclerotic disease and have an impact on future surgical or endovascular intervention involving head and neck vessels. Our aim was to quantify the morphometric differences between ASO patients and healthy controls.
Methods: A total of 20 ASO patients and 20 age and BSA matched healthy controls were enrolled in this study. All individuals signed informed consent. Cardiac magnetic resonance imaging (MRI) was performed in all subjects with a 1.5 Tesla MRI scanner. Using standard MRI sequences, balanced steady state free precession (bSSFP) 3D-wholeheart images were acquired to reconstruct a three-dimensional (3D) model of the left heart of each individual which included: left ventricle (LV), left ventricular outflow tract, aortic root, ascending aorta, aortic arch and descending aorta to diaphragm level. The geometrical analysis assessed: (i) the angle between the line connecting the valve centre with the LV apex and the aortic valve plane; (ii) the angle between the aortic valve plane and the sinotubular junction plane; (iii) the indexed length of the vessel centreline starting from the centre of the aortic valve to the level of the diaphragm; (iv) the indexed curvature of the aortic arch, from the inverse of the radius (r) of the maximum circumference fitted at the highest point of the centreline ($= 1/r$), and (v) the aortic root volume. Independent samples T-test was used to compare mean differences.
Results: Geometric differences between the two groups are shown in Figure. The two groups were closely matched for age (15.3 ± 1.8 vs. 14.9 ± 1.9 years for ASO vs. controls) and BSA (1.8 ± 0.2 vs. 1.6 ± 0.2 m^2 for ASO vs. controls).
Conclusions: This work highlights quantifiable differences in the 3D morphology of the thoracic aorta in ASO patients compared to healthy controls. This geometrical abnormality might reflect in the development of unusual flow patterns and potential arteriosclerotic disease.

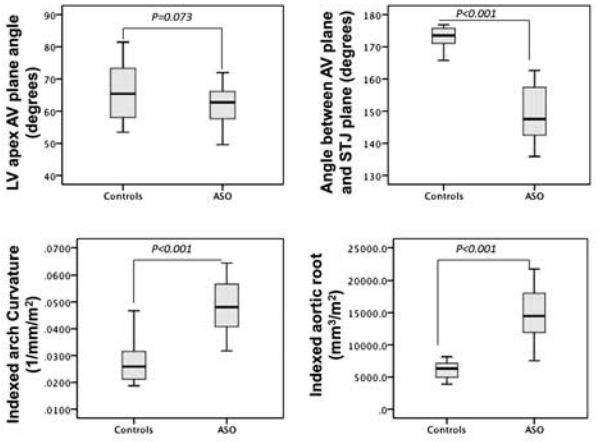

Figure
Box plots of geometric differences between arterial switch and controls with p<0.05 showing statistical difference (LV- left ventricle, AV- aortic valve, STJ- sino-tubular junction).

O6-8
Visualisation of flowpatterns in the Fontan Circulation by 4-dimensional respiratory- and ECG-triggered phase contrast magnetic resonance imaging
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Background: Evaluation of blood flow characteristics in total cavo-pulmonary connection (TCPC) with CMR remains difficult due

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

to its strong modulation by respiration, and is not yet entirely understood. New approaches using 4D phase contrast magnetic resonance imaging (4D PC MRI) are promising and can contribute to the understanding of hemodynamics in the Fontan Circulation. Our objective was to visualize and compare flowpatterns in the TCPC using respiratory- and ECG triggered 4D PC MRI.
Methods: 10 children with hypoplastic left heart syndrome were evaluated after surgical completion of the Fontan-circulation (TCPC with lateral intra-atrial tunnel) in a single center. In all patients one respiratory (80–100 phases) and one ECG (30 phases) triggered 4D PC MRI covering the whole thorax, voxel size ranging from isotropic 1.5^3 to 2.0^3 mm^3 were acquired during a single CMR examination. Dedicated commercial and custom-made software was used for detailed analysis and visualization of flowpatterns.
Results: Respiratory-triggered acquisitions revealed significantly higher maximum and lower minimum flow, maximum and minimum velocity in the inferior vena cava and tunnel compared to ECG-triggered 4D PC MRI. Flowpatterns, e.g. expiratory backflow from the left pulmonary artery to the lower intra-atrial tunnel (see fig. 1), that could not be detected on ECG-triggered 4D PC MRI could easily be visualized by respiratory-triggered 4d PC MRI.
Conclusion: Respiratory-triggered 4D PC MRI of the TCPC avoids averaging of flow and velocity over the respiratory cycle, resulting in significant differences of blood flow volume and flow velocities to solely ECG-triggered acquisitions. This study suggests that hemodynamics in the TCPC are mainly dependant on respiration, while ventricular function causes only minor modulations of flows in the TCPC connection. 4D PC MRI adds to our understanding of hemodynamics and fluid mechanics in the Fontan-circulation and may help to identify patients at risk for failure.

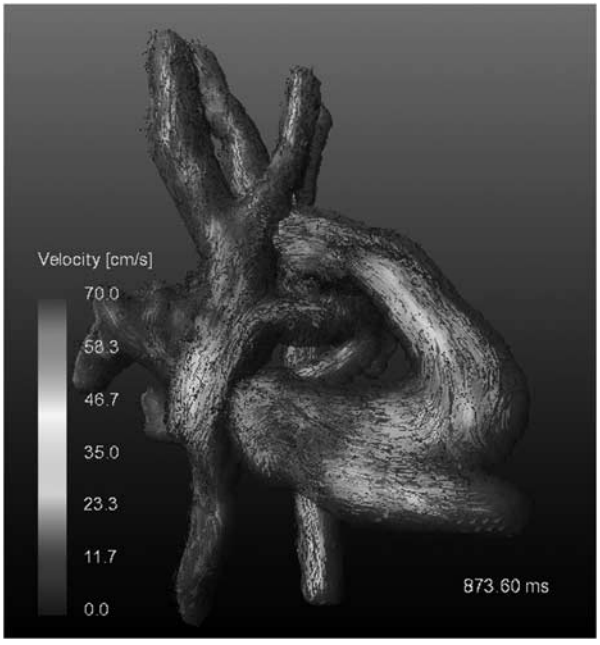

Fig. 1: Expiratory backflow from LPA

O7-1
Abnormal myocardial rotation is a non-invasive marker of rejection in paediatric heart transplant recipients
Grotenhuis H., Nyns E.C.A., Kantor P.F., Greenway S.C., Holtby H., Chaturvedi R., Yoo S.-J., Grosse-Wortmann L.
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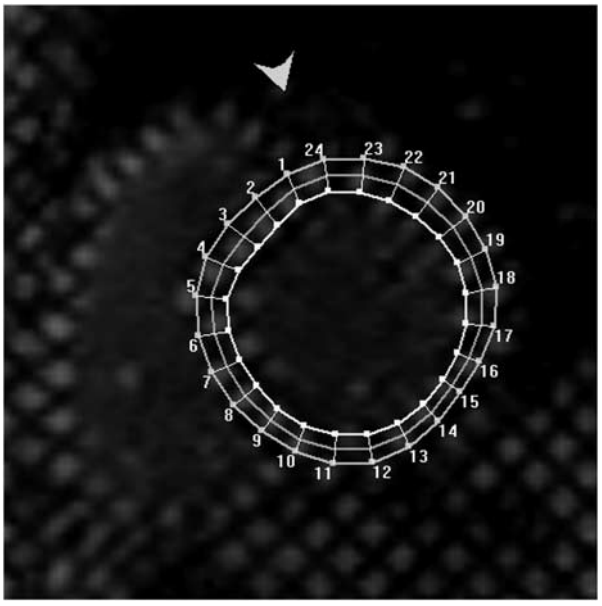
Background: Graft function may be compromised for a variety of reasons after heart transplantation, including rejection. The objective of the current study was to assess cardiac function and the relationship with the presence and degree of rejection as determined by endomyocardial biopsy in paediatric heart transplant recipients.
Methods: Cardiac magnetic resonance (CMR) imaging was performed in 14 patients (11 male; mean age 13.9 years ± 4.7 (range 2.4–17.9 years), 1.2 years ± 1.3 (range 12 days–5.0 years) after heart transplantation. CMR was obtained at the time of routine endomyocardial biopsy for rejection surveillance. A total of 18 combined CMR studies/biopsies were performed). In addition to biventricular systolic function and dimensions, left ventricular (LV) circumferential strain, rotation, twist and torsion were measured using myocardial tagging (Figure). The results were compared to those of 9 age-matched controls. All transplant patients also underwent routine cardiopulmonary exercise testing.
Results: Heart transplant patients showed lower LV and right (RV) ejection fraction (EF): LVEF 55 ± 8 vs. 61 ± 3%, $p < 0.01$, RVEF 48 ± 7 vs. 53 ± 6%, $p = 0.03$) and increased LV mass, indexed to body surface area (67 ± 14 vs. 55 ± 13 g/m^2, $p = 0.01$). Global LV circumferential strain (−13.5 ± 2.3 vs. −19.1 ± 1.1%, $p < 0.01$), basal strain (−13.7 ± 3.0 vs. −17.5 ± 2.4, $p < 0.01$), mid-ventricular strain (−13.4 ± 2.7 vs. −19.3 ± 2.2, $p < 0.01$) and apical strain (−11.8 ± 7.2 vs. −19.9 ± 2.0, $p < 0.01$) were significantly reduced in patients when compared with controls. In addition, LV rotation (6.1 ± 1.65 vs. 7.8 ± 1.13°, $p < 0.01$) was decreased and basal rotation was abnormal (−2.0 ± 2.1 vs. −5.0 ± 2.0°, $p < 0.01$) in patients compared with controls. Transplant patients also showed decreased LV torsion (6.1 ± 1.65 vs. 7.8 ± 1.13°, $p < 0.01$). Heart transplant recipients had reduced predicted peak work load (58 ± 12%), predicted peak oxygen consumption (PVO_2) (55 ± 12%) and predicted PVO_2 at aerobic threshold (57 ± 14%) as compared to a paediatric reference population. The severity of rejection correlated inversely with LV twist ($r = -0.53$, $p = 0.02$) as well as basal ($r = -0.48$, $p = 0.04$), mid-ventricular ($r = -0.56$, $p = 0.02$), and apical rotation ($r = -0.65$, $p < 0.01$).
Conclusion: Paediatric heart transplant recipients have mildly reduced systolic global biventricular function, increased LV mass, abnormal rotation, strain, torsion and twist, as well as impaired exercise performance. Abnormal rotation appears to be a non-invasive marker of graft rejection.



https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

O7-2

Usefulness of Cardiac Magnetic Resonance in the Assessment of Myocardial Inflammation and Fibrosis in Children Born to Mothers with Anti-SSA/Ro Antibodies: A Prospective Study of 26 Cases and 6 Controls

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Background: Mothers known to have anti-SSA/Ro or anti-SSB/La antibodies are at risk of delivering babies with cardiac neonatal lupus erythematosus syndrome (NLES). Besides congenital heart block (CHB), other manifestations include endocardial fibroelastosis (EFE) and dilated cardiomyopathy. Autopsy data suggests that myocardial lesion is under-diagnosed by fetal and post natal echocardiography. No data are available regarding the interest of cardiac magnetic resonance (CMR) to evaluate inflammation and fibrosis in children exposed in utero to anti-SSA/Ro antibodies (Ab).
Patients and materials: Children born to mothers with anti-SSA/Ro Ab were enrolled in a prospective study performed from December 2010 to October 2012. The control group consisted of 6 healthy children born to mothers without anti-SSA /Ro Ab. All the children had a complete cardiac testing and a CMR with intravenous administration of gadolinium: High-signal on T2-weighted imaging corresponded to inflammation whereas late myocardial enhancement depicted fibrosis.
Results: A total of 26 children exposed to anti-SSA/Ro Ab were included. Children were separated in group 1 with cardiac manifestations of NLES, and group 2 with no cardiac NLES.
Group 1 (N = 16):
Cardiac manifestations included conduction defect (n = 14, complete CHB in 11), supraventricular tachycardia (n = 2), and/or EFE diagnosed on echocardiography (n = 7, fetal echocardiography was not available in 1 case).
CMR was abnormal in 11 children: inflammation (n = 3), fibrosis (n = 7), and both (n = 1). Only 3 of those 11 children had an abnormal postnatal echocardiography.
Among the 14 children with conduction defect, 9 had an abnormal CMR. The other 5 CMR were normal in 3 and incomplete in 2 since injection could not be performed.
Group 2 (N = 10): CMR was abnormal in one 3 months old child positive for inflammation and fibrosis.
Control group (N = 6): CMR was always normal in anti-SSA-Ab non exposed children.
Conclusion: Since late cardiomyopathy contribute to the morbidity and mortality of cardiac NLES, it is necessary to identify the predictors of its occurrence in NLES. EFE, especially when it involved the left ventricle, is probably one of them. CMR was more sensitive to detect persisting cardiac inflammation and/or fibrosis than postnatal echocardiography, and can potentially be a valuable tool to assess this risk.

O7-3

Dramatic Improvement of the prognosis of idiopathic PAH in the young during the last 3 decade - Predictive factors from a single center experience with 92 cases -

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Purpose: Idiopathic pulmonary arterial hypertension (iPAH) is an intractable and the prognosis treated at a single center with advanced therapy (AT) has not been well reported. Herein, we studied retrospectively for the significant prognostic factors of iPAH.
Subjects and Methods: Consecutive 92 patients with iPAH (47 F/45 M) since 1978 were enrolled. The age at onset was 11.7 ± 8.4 y/o, and the median follow-up was 70 months (ranging 0.4m - 28y). Nearly 1/4 of patients were found by healthy screening in the school. We reviewed medical records for their clinical parameters such as the CTR, BNP, NYHA-FC, mode of advanced therapies, genetic analysis including BMPR2, ALK1, ALK6, and Smad 8, and the outcome. Lung transplantation and the mortality were defined as an event. To assess the correlation between available therapeutic options and the prognosis, patients were divided into 4 subgroups depending on the era when vasodilator therapy was started as follows; that is, group A (~1993), B (~1999), C (~2003), and D (2003~). We started oral PGI2 from 1993, IV-PGI2 from 1996, Sildenafil from 2003, and Bosentan from 2005, Tadarafil from 2010, Ambrisentan from 2011.
Results: At June 2012, the 5y-survival of group A to D were 14.3 ± 13%, 61.5 ± 12%, 85.0 ± 15%, 91.7 ± 8%, respectively ($p < 0.0001$). Sixty-four patients (69.6%) were alive including 7 (7.6%) after lung transplantation (LT). Twenty-eight (30.4%) deceased including 1 death after LT. The actuarial survival at 3, 5, 10 years are 88.8 ± 11%, 74.1 ± 26%, and 59.7 ± 40%, respectively. Cox regression analysis revealed that CTR before treatment was the significant independent predictor of event-free survival (RR of 1.09, $p = 0.003$). Their survival rate was significantly better than those with cardiomegaly of 50% or more (K-M curve with Log-rank test; $p = 0.02$). Starting the AT with BNP < 200 and NYHA-FC ≤ II, patients found by school healthy screening, and negative for ALK1 gene also showed excellent results.
Conclusions: We conclude that the prognosis of iPAH in the young has been improving significantly because of ATs, especially last 10 years. The prognosis is significantly affected by CTR, BNP, NYHA-FC before treatment and mode of combination therapy and genetic background.

O7-4

Limited preload reserve during exercise limits exercise capacity in healthy Fontan patients

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Introduction: Factors controlling cardiac output in a Fontan circulation are still poorly understood. This study aimed at evaluating changes in exercise hemodynamics using a novel cardiac magnetic resonance (CMR) methodology during mild, moderate and strenuous exercise.
Methods: Ten Fontan patients (6 male, age 20 ± 4 years, NYHA 1–2) underwent CMR at rest and during supine exercise on a programmable cycle ergometer. Systemic ventricular volumes were obtained at rest (heart rate 72 ± 14 bpm) and during mild (100 ± 10 bpm), moderate (122 ± 15 bpm) and strenuous (144 ± 15 bpm) exercise. Images were acquired using an ungated, free-breathing real-time CMR sequence (12–18 contiguous 8 mm slices). Software was developed to allow for synchronization of short and long-axis images with compensation for respiratory phase translation. Endocardial borders were delineated

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

using a bi-plane model. Simultaneously, radial and pulmonary artery pressures were measured
Results: Cardiac output (CO) increased continuously during exercise (6.8 ± 1.6 vs 10.0 ± 3.2 vs 11.8 ± 3.2 vs 12.5 ± 3.1 l/min; P < 0.0001). The increase in CO depended on a 106 ± 49% increase in heart rate as stroke volume (SV) did not change from rest to mild exercise and decreased during moderate and strenuous exercise (95 ± 19 vs 100 ± 21 vs 96 ± 20 vs 87 ± 16 ml; P < 0.0001). End-diastolic volume (EDV) decreased during strenuous exercise (169 ± 38 vs 174 ± 39 vs 170 ± 38 vs 162 ± 36 ml; P = 0.029), whereas end-systolic volume (ESV) did not change during exercise (74 ± 31 vs 74 ± 31 vs 73 ± 32 vs 75 ± 32 ml; P = 0.944). Ejection fraction (EF) decreased during strenuous exercise (57 ± 10 vs 58 ± 10 vs 59 ± 11 vs 55 ± 11%, P = 0.029). Pulmonary artery pressures (9 ± 3 vs 14 ± 4 vs 17 ± 4 vs 21 ± 5 mmHg; P < 0.0001) and mean systemic artery pressures (81 ± 7 vs 92 ± 7 vs 100 ± 6 vs 106 ± 4 mmHg; P < 0.0001) increased during exercise. Arteriovenous oxygen difference increased from 5.5 ± 1.2 to 9.6 ± 1.9 ml/100 ml (P = 0.001).
Conclusions: A decrease in SV and EDV during exercise despite a blunted heart rate response indicates a limited ventricular preload reserve. The decreased preload reserve in a Fontan circuit is an important determinant limiting exercise capacity.

O7-5
Sildenafil Improves Exercise Hemodynamics in Fontan patients

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Background: Reduced preload reserve, especially with increased heart rate, is a fundamental limitation of the Fontan circulation. Sildenafil may increase exercise capacity in patients with Fontan physiology, but the underlying mechanism is still unclear. This study tested the hypothesis that sildenafil would improve exercise hemodynamics, especially ventricular filling, in Fontan patients using a novel cardiac magnetic resonance (CMR) methodology.
Methods: Ten Fontan patients (6 male, age 20 ± 4 years) underwent CMR at rest and during supine exercise on a programmable cycle ergometer before ("baseline") and after a single dose of sildenafil (50 mg oral). Systemic ventricular volumes were obtained at rest and during mild (104 ± 11 bpm), moderate (127 ± 16 bpm) and strenuous (147 ± 15 bpm) exercise. Ventricular filling rate (VFR, ml/msec) was defined as stroke volume corrected for RR-interval. Bi-plane cine images were acquired using an ungated, free-breathing real-time CMR sequence (12–18 contiguous 8 mm slices) and analyzed using software developed enabling retrospective gating for cardiac phase and respiratory translation. Endocardial borders were delineated using a bi-plane model. Simultaneously, radial and pulmonary artery pressures were measured.
Results: Under resting conditions as compared with baseline, sildenafil reduced pulmonary artery pressure (9 ± 3 to 8 ± 3 mmHg, P = 0.029) and increased cardiac output (6.8 to 8.1 L/min, P = 0.006) and VFR (113 ± 27 to 134 ± 35 ml/sec, P = 0.006). During exercise sildenafil resulted in improved hemodynamics as compared with baseline. Pulmonary artery pressure decreased (mean difference 1.6 ± 1.0 mmHg, P = 0.006), whilst cardiac output (mean difference 1.7 ± 0.8 L/min, P = 0.001), ejection fraction (mean difference 4.8 ± 2.8%, P = 0.003), stroke volume (mean difference 6.0 ± 5.6 ml, P = 0.039) and VFR (mean difference 28 ± 14 ml/sec, P = 0.001) all increased.
Conclusion: In patients with Fontan physiology, sildenafil improves cardiac output during exercise despite a reduction in pulmonary artery pressures. This implies that pulmonary vasodilation is a potential physiological target for improving exercise hemodynamics, the clinical significance of which warrants further study.

O7-6
Long term follow-up after heart transplantation in very young children

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The aim of this study was to review long-term follow-up of heart transplanted small children to assess prognosis and outcomes
Material and methods: Patients who underwent orthotopic heart transplantation (OHT) within the first 3 years of life were included in the study. Demographics, clinical data, events, outcomes and survival were assessed.
Results: Among 96 paediatric heart transplantations performed in a French single-centre, 25 were included in the study (10 males, 15 females). Among them, 10 (40%) were on VAD support at the time of OHT. Age at OHT was 1.5 ± 0.9 years (median 1.2). Underlying cardiac disease was congenital in 4 (16%) or idiopathic cardiomyopathy in 21 (84%). Post-transplant follow-up was 7.1 ± 7 years (range 1 day to 22.7) and was > 10 year in 7 cases (28%). Three patients died at 1st day, 2nd year and 4th year post-transplant. Mean age of survivors at the time of the study was 9.1 ± 7.3 years (range 1.5 to 23.6). One acute rejection episode occurred in 2 patients, at 1st month and one at 11st year. One post-transplant lympho-proliferative disease occurred at 14th year post-transplant and was successfully cured. Graft coronary disease occurred in 2 cases (8%), who underwent 2nd-heart and kidney transplantation at 16th and 22th year after first transplant. All others were free from coronary disease with normal graft function. End-stage renal failure occurred in the 2 re-transplanted cases. Severe renal dysfunction was present in 3 cases (no dialysis), moderate in 3 cases, and 17 had normal renal function. Linear growth was normal in all patients, except the 2 cases with end-stage renal failure, despite maintenance low dose steroids in 80% of the survivors. All are in NYHA class I, except the 2 re-transplanted cases who were in class IV at the time of 2nd transplant. Patient survival was 96% at 1-year, 90.7% at 3-year and 83% at 10-year post-transplant. Graft survival was respectively 96%, 90.7%, 83% and 66% at 1, 3, 10 and 16-year follow-up.
Conclusion: Long-term survival of very young heart transplant recipient is acceptable, with low incidence of graft coronary disease and optimal functional status and growth.

O7-7
Cardiac and Multi-Organ Transplantation for End-Stage Congenital Heart Disease

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Introduction: Cardiac transplantation for patients with complex congenital heart disease (CHD) has been associated with increased morbidity and mortality compared to other transplant patients. We report our single center experience with patients

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

who had cardiac and multi organ transplantation for end stage CHD.
Methods: We reviewed records for all patients with CHD transplanted at the Mayo Clinic in Rochester, Minnesota, USA, from January 1990 through June 2012. Patients with cardiomyopathy (including hypertrophic cardiomyopathy) were excluded.
Results: Overall, 45 patients had cardiac transplantation for end stage CHD (mean age 26.1 ± 18.4 years; range 1 month to 65 years). Congenital diagnoses included single ventricle physiology (N = 16), d-transposition of the great arteries (N = 8), ventricular or atrial septal defects with subsequent functional deterioration (N = 4), Ebstein anomaly (N = 6), tetralogy of Fallot (N = 4), congenitally corrected transposition (N = 4), and complex left sided lesions (N = 3). Patients had a mean of 2.6 (0–8) prior cardiac operations, including 37 (82%) with a prior sternotomy and 15 (33%) with a prior thoracotomy. There were 7 (16%) with a history of Fontan palliation prior to transplantation. Two patients had combined heart/liver transplantation; one had heart/kidney transplantation. Ten patients (22%) required additional procedures (most commonly pleurodesis or wound exploration). Patient survival at 1, 5, and 10 years was 88%, 86%, and 69%, while graft survival at 1, 5, and 10 years was 88%, 86%, and 58%. Over the same era, ISHLT reported patient survival in patients with cardiomyopathy was 85%, 72%, and 56%. Over a mean of 8.7 ± 6.2 years of follow-up, rejection requiring treatment was documented in 35 patients (78%). Eleven patients (24%) have been diagnosed with neoplasia (8 skin, 2 lymph, 1 other). Three patients (7%) have required retransplantation. Four patients (9%) have developed significant coronary vasculopathy; one was retransplanted, while three died 10 ± 4 years after transplantation.
Conclusions: With appropriate patient selection and post transplant monitoring, survival for patients with complex end stage CHD can be equivalent to patients with cardiomyopathy. Multi organ transplants are an option for selected patients with CHD.

O7-8
The role of perinatal autopsy in prenatal interventions patients

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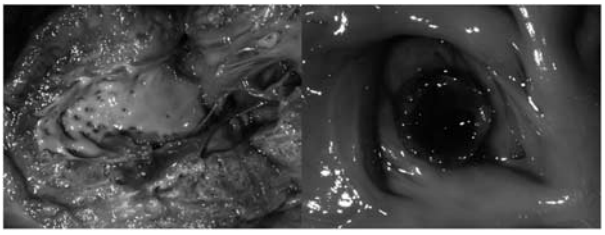
Introduction: Knowledge about results of prenatal interventions and causes of perinatal mortality related to them remains insufficient. There are no collected autopsy data regarding patients who died in utero or in the early neonatal period published so far. We report autopsy results of foetuses and newborns who died after prenatal interventions.
Methods: Between 2011–2012 19 fetal procedures were performed: balloon aortic valvuloplasty (BAV) -13, pulmonary balloon valvuloplasty (PBV) -2, balloon atrial septoplasty (BAS) - 2. In one fetus with hydrops due to severe heart failure and polihydramnios, BAV, BAS and implantation of stent to the interatrial septum (SIAS) were done. Interventions were performed between 20 and 31 week. 3 fetuses and 1 neonate died: 2 foetuses after BAV died in utero (late placental insufficiency unrelated to BAV - 1, placental abruption after transplacental BAV - 1), 1 foetus after BAS died in utero due to cardiac tamponade, 1 neonate born in 30 weeks after BAV, BAS and SIAS in 8th day of life due to multi-organ failure. Autopsy was performed in all 4 cases.
Results: In foetuses and the newborn after BAV the autopsy revealed dysplastic, bifoliate aortic valves and confirmed the technical success of BAV. The left ventricle of one foetus was severely dilated and thin-walled. In the other foetus after BAV and in the newborn after BAV, BAS and SIAS extensive endocardial fibroelastosis and diffuse myocardial fibrosis were observed. In foetus after BAS the perforation in the interatrial septum was almost entirely obstructed. Thickened left atrial and pulmonary venous walls were found. The stent in the interatrial septum in newborn after BAV, BAS and SIAS was in good position. The thrombus attached to its wall appeared to be responsible for postnatal stent obstruction and unfavourable outcome.
Conclusions: In all foetuses and newborns who died after prenatal interventions the autopsy should be obligatory. It allows to check results of procedure and explain causes of perinatal mortality. Autopsy data from all prenatal intervention centres should be collected and correlated with clinical and embryological studies.



O8-1
Comparison of percutaneous dilatation and surgical valvotomy in infants with aortic stenosis: a single center survey

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Objective: The management of critical aortic stenosis between balloon valvotomy and surgical valvotomy remains a controversial topic. Here,we report our experience with these two techniques in a single tertiary referral center over a period of 24 year.
Methods: Retrospective analysis of files of 155 infants (<1 year) at the time of first intervention, and born between May 1986 and November 2009. Clinical data were collected: sex, age, birth weight,transvalvular aortic gradient in mmHg and semi-quantitative aortic regurgitation before/after procedure, aortic annulus size,fractional shortening, aortic valve morphology and associated cardiac malformations.
Results: 84 infants with aortic stenosis underwent surgical valvotomy (S - 43 newborns) and 71 infants underwent balloon valvotomy (D-51 newborns). 29% in the surgical group and 37% in the dilated group had associated cardiac malformations. The procedure was successful in 82% D and in 92% S with significant decrease in mean aortic gradient (47 ± 21% in D and 52 ± 17% in S). LV dysfunction was more frequent in newborns (42 vs 22%, $p = 0.02$) and was, with associated mitral valve malformations, the main risk factor for increased early and total mortalities in newborns (16% vs 3%, $p < 0.02$ and 30% vs 7%, $p < 0.001$). If newborns with LV dysfunction or with mitral valve

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

malformations were excluded, no difference was found between dilated (n = 21) and operated (n = 36) patients for the rate of re-interventions (21.5%), aortic valve replacement (8.5%) and the delay of re-interventions (median of 3.5 years). Re-interventions for aortic regurgitation with/without stenosis was not significantly different (14% vs. 3%, $p > 0.05$) but overall mortality was higher in the dilated group (29% vs 3% $p < 0.05$). After 10 years follow-up, 50% of survivors remained free of any type of re-intervention in both groups.
Conclusion: In this single center retrospective study, there was no significant difference in outcomes for the two techniques in infants without LV dysfunction and mitral valve malformation although overall mortality was higher in the dilated group.

O8-2

When Routine RF [RFA] - or Cryoablation [CRYO] Fails: Application of the Alternative Ablation Mode [either RFA or CRYO] for Successful Ablation Therapy in Pediatric Patients and Patients with Congenital Heart Disease [pts]

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Introduction: Transvenous cryoablation for septal tachycardias might allow a lower incidence of AV block and cryoadhesion can facilitate ablations through better tissue contact. RFA though is considered as more effective and efficient than CRYO and deeper lesions can be produces using irrigated tips.
Aim of the Study: We report our results of the usage of RFA and CRYO in those pts where the initially chosen ablation mode failed.
Patients and Methods: From 1/2004 until 12/2012 503 pts were ablated. Ablation mode was single use of RFA in 76.7% (n = 386) and CRYO in 18.6% (n = 85) alone. In 6.4% (n = 32) a combination therapy of RFA and CRYO [COMBI] was used.
Results: For COMBI, median pts age was 12.9 yrs. (range 6.5–28.4) and mean weight was 48.4 ± 15 kg.31% of the pts. had CHD. Dx were WPW/AP in 15; AET in 7, AVNRT in 5, VT in 3 and other in 3 pts. There were 46 arrhythmogenic substrates (1.4/pt.). Procedure time was 272 ± 105 min, radiation dose was 1587 cGy × cm^2. Overall success rate was 84.4% (27/32 pts), there were no complications. In 3 pts RFA and CRYO were used for two different targets selectively with a success rate of 100%. In the other pts initial intention to treat with RFA (n = 13) was unsuccessful, but CRYO was effective in 69%. In 15 pts initial CRYO failed, but RFA could cure the arrhythmia in all cases (see table for substrates).

Intention to treat (single tachycardia substrate):	*Success with RFA*	*Success with CRYO*
CRYO failure (n = 19)	*100% (n = 19)*	
RFA failure (n = 15)		*66.7% (n = 10)*

Conclusions: 1. Combination of RFA and CRYO in pts with more than 1 arrhythmogenic substrate is effective and safe. 2. In pts where the initial ablation mode - either RFA or CRYO - fails, the additional usage of the other ablation mode can effectively improve the success rate. 3. The additional usage of RFA after failed CRYO seems to be more effective than CRYO usage after RFA failure. 4. COMBI improves outcome in difficult tachycardia substrates in children and pts with CHD.

O8-3

Mid- to long term follow-up of coronary artery bypass grafting in children – a single centre experience

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Background: Literature about coronary artery bypass grafting (CABG) of children and young adults is rare.
Methods: This is a 20 year-single centre retrospective observational analysis of all pediatric patients (pts) who underwent one or more CABG less than 18 years old. We reviewed diagnosis, therapy, long term surgical results and outcome.
Results: 16 pts with CABG were identified since 1992 at our centre. CABG was necessary after surgeries due to CHD (n = 10), coronary artery malformation (n = 3) and stenosis subsequent to Kawasaki disease (KD) (n = 3) at a median age of 4.8 (0.04–16.75) years. The left internal mammary artery (LIMA) was used in 8 pts; the right internal mammary artery (RIMA) in 2. 1 pt with RIMA graft needed reactivation with a saphenous vein after graft closure. A venous graft was used in 3 pts. One pt with KD received LIMA, RIMA, and a venous graft. In 2 pts the bypass type was not mentioned in their surgery report. After surgery all pts received continued oral anticoagulation.
Median follow-up was 9.8 (2.8–22.3) years. Four pts were NYHA class II-III in their last follow-up. In these four pts angiography showed graft closure with collateral formation (n = 1, after Ross procedure), stenosis (n = 1, after Ross procedure), graft closure due to/with normal antegrade coronary artery perfusion (n = 1, after arterial switch), normal perfusion (n = 1, after arterial Switch). Magnetic resonance imaging (MRI) revealed no sign of ischemia in 3 of them, one pt with graft closure showed perfusion defects. The remaining 9 pts are doing well with no restriction to their daily lives and activities. MRI scan demonstrated small myocardial perfusion defects in 3 of them, in 2 no MRI scan was performed. The patency rates of the LIMA grafts were 67% (n = 6), RIMA 67% (n = 2), venous graft 40% (n = 2).
Conclusion: Despite small vessels,narrow anastomotic sites at implantation, and pts growth, CABG using arterial grafts (LIMA/RIMA) show good overall mid- to long term results in children in typical lesions as after cardiovascular surgery, Kawasaki disease and coronary malformation. Venous grafts have a lower patency rate.

O8-4

MRI catheter stress haemodynamics in hypoplastic left heart syndrome after Fontan completion

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Introduction: Effort intolerance and signs of a failing circulation are common following the Fontan procedure. We studied the haemodynamics of the Fontan circulation using MRI catheterisation at baseline and maximal pharmacological stress.
Methods: Prospective study of children with hypoplastic left heart syndrome (HLHS) post Fontan referred for MRI catheter due to signs or symptoms of a failing Fontan, poor cardiopulmonary exercise test performance or abnormalities on routine MRI. Quantification of volumetrics and flow was from MRI with

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

simultaneous invasive catheter pressure measurements. Measurements were made during baseline conditions (Stage 1), dobutamine at 10mcg/kg/min (Stage 2) and 20mcg/kg/min (Stage 3). Control data was from 10 healthy adults with biventricular hearts without invasive measurements.
Results: Eleven patients recruited; median age 9.47years (3.54–11.1 yrs). All reported exercise intolerance. Two had plastic bronchitis, of whom 1 also had protein losing enteropathy. One patient only received 10mcg/kg/min dobutamine due to an elevated blood pressure response. There were no reported adverse events.
Median heart rate (HR) increased from 80 bpm to 162 bpm ($p < 0.05$) in HLHS, against 67bpm to 119 bpm ($p < 0.05$) in controls. In HLHS, indexed cardiac output (iCO) was 2.8 L/min/m^2, increasing by 53% from baseline to 4.3 L/min/m^2 ($p < 0.05$) at stage 2 matching controls (Figure 1). However there was no significant further increase at stage 3 ($p = 0.21$), with iCO of 4.4 L/min/m^2 despite increased HR and ejection fraction (EF). There was a significant fall in indexed end diastolic volume (iEDV) during stage 2 and 3 not seen in controls (Figure 2). The maximal fall in indexed end systolic volume (iESV) from baseline was similar in both groups at 50% in HLHS and 54% in controls.
Mean baseline transpulmonary gradient in HLHS was 3.5 mmHg ± 1.12 with a pulmonary vascular resistance (PVR) of 1.63 wu.m^2 ± 0.55. Mean resting right ventricular end diastolic pressure (EDP) in HLHS was 6.5 mmHg ± 2.8.
Conclusion: There is good systolic reserve in response to dobutamine stress. The rise in cardiac output is blunted at higher dose dobutamine in the HLHS Fontan. As the resting ventricular EDP is normal, we hypothesize that this is due to an inability to appropriately increase blood flow through the pulmonary circulation and hence ventricular preload despite low resting PVR.

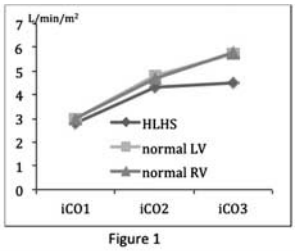

Figure 1

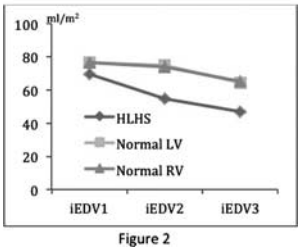

Figure 2

O8-5
Hybrid Approach for Infants with Pulmonary Atresia, Ventricular Septal Defect, extreme Hypoplasia of Central Pulmonary Arteries and Major Aortopulmonary Collaterals
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Objectives: to report our early experience with treatment of pulmonary atresia with ventricular septal defect (PAVSD), severe hypoplastic central pulmonary arteries (PAs) and major aorto-pulmonary collaterals (MAPCAs), by a hybrid approach.

Background: PAVSD and MAPCAs is a complex congenital heart defect that can vary according both to PAs anatomy and source of pulmonary blood flow. Management of this lesion is still controversial. Early establishment of a forward flow through the native pulmonary arteries could promote their growth; however results from both surgical and percutaneous approaches are often disappointing when required in small infants.
Methods: the department registry was retrospectively searched for all patients affected by PAVSD, MAPCAs and severe hypoplastic PAs (< 2 mm) and treated with a hybrid approach between February 2007 and March 2012.
Results: a perventricular implantation of a right ventricle (RV) to PA stent was attempted in five hypoxemic infants (mean age, weight and saturation were 65 days, 3,5 Kg and 60%, respectively). The procedure was performed under general anesthesia, throughout a midline sternotomy. Under direct vision a 22 Gauge needle was advanced towards the free wall of the RV into the pulmonary trunk. After angiography, a 0.014 inch floppy guide wire was inserted into a pulmonary branch and a 3Fr sheath placed over that wire. A Taxus Libertè 3.5 mm × 19 mm stent was implanted. The procedure was successful and final angiography showed a well-placed stent in all cases. Mean total procedural time was 175 minutes with a mean X-ray time of 8 minutes. The post-operative course was uneventful in all infants but one in whom we had an early major complication. At a mean follow-up of 43 months all patients are alive. Four of them had a new opened RV to pulmonary artery conduit (VSD left opened), with increased PAs size and an oxygen saturation above 82%. Only one patient was considered unsuitable for other surgical procedure, having showed complete stent occlusion at CT scan control, without any evidence of central PAs.
Conclusions: hybrid approach allows to obtain growth of native PAs even in small infants with the most unfavorable PAs anatomy.

O8-6
Optimal alignment of the Figulla® Flex Occluder (FFO) to the atrial septum before release in patients with atrial septal defects
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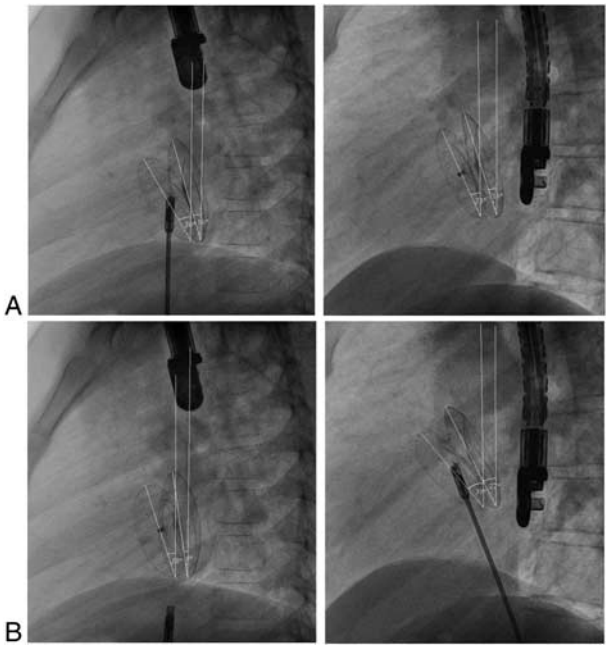
Introduction: Closing atrial septal defects (ASD's) may be challenging for several reasons including deficient or floppy rims, abnormal anatomy of the ASD, the absolute ASD size in correlation to the atrium or the proximity to other relevant intracardiac structures such as coronary sinus or the AV-valves. Optimal alignment is those cases is crucial to visualize the presumed final position of the device. The Figulla® Flex device has a mobile joint connecting the delivery cable in a tilted angle of 45 degrees without significant stress on the implant. We report the results of the change in orientation before and immediately after device release.
Patients: Over the past 2 years we implanted 85 FFOs in our patients for interventional ASD closure (age 1 to 48 years, weight 6 to 100 kg). All investigations were performed under deep conscious sedation under TEE and/or TTE guidance (below 10 kg bw). Standard fluoroscopy included a strict lateral projection during the release process of the device (see pictures). The angles of the left and right atrial discs before (A and B) and

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

after (C and D) the release of the device were measured and the difference (A-C and B-D) calculated thereafter.



Examples in 2 patients: note the minimal changes of the angles before and after release of the device

Results: The angles before release were A: 28,2° (±7,2°) and B: 42,6° (± 7,2°) before and C: 18,1° (±5,3°) and D: 26,5° (± 6,6°) after release, resulting in a change of the left atrial disc in 10° (±4,3°) and the right atrial disc in 16,1° (±6,4°) only.

Discussion: Whereas other devices with a simple screw system to connect the delivery system with the device cause a significant torsion and tension of the device, the delivery system of the FFO allows with the tilt of 45° an ideal adaptation to the septal tissue. All devices could be placed in the final position without adverse tension or stress caused by the delivery wire. We think that this device offers superior anatomical adaptation even in challenging anatomy.

O8-7

Safety of transcatheter closure of atrial septal defect with a fenestrated Amplatzer septal occluder in patients with pulmonary hypertension or heart failure

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Purpose: In patients with atrial septal defect (ASD) complicated with moderate-to-severe pulmonary hypertension or heart failure, complete closure of the defect may carry significant risks. A fenestration was generally created in the occluder for gradual reduction of shunt to close the ASD in those high risk patients.

Methods: During a 10.5-year period, 44 patients (10 males) with ages ranging from 7 to 81 years underwent transcatheter closure of ASD with a fenestrated device. Of them, 39 patients had moderate-to-severe pulmonary hypertension, 4 had heart failure and 1 had pulmonary atresia intact ventricular septum with a right atrial pressure above 15 mm Hg after balloon test occlusion. A fenestration was created about 1/3 to 1/4 of the diameter of the Amplatzer septal occluder. The techniques of device deployment are similar to those reported.

Results: Of the 44 patients, the mean pulmonary artery systolic pressure was 66 ± 18 mmHg & mean Qp/Qs ratio was 2.7 ± 1.4. The mean device diameter used was 30 ± 6 mm. Implantation was initially successful in all 44 patients. Immediately after implantation, shunt flow across the fenestration was observed in all 44 patients. However, 1 developed embolization of the device several hours later. The patient was sent for emergent surgery. After a mean follow-up period of 42 ± 15 months, majorities of patients had improvement in symptoms, regression of right heart dilation & decrease in pulmonary artery pressure. Three patients underwent a second procedure to close the residual defect because of presence of significant shunt 12 months later. Twelve patients received Sildenafil. Six patients had very small residual shunt noted on the most recent echocardiography.

Conclusions: Transcatheter closure of ASD in patients with moderate– to-severe pulmonary hypertension or heart failure using a fenestrated device is safe and effective.

O8-8

The EDWARDS VALEO LIFESTENT® for treatment of cardio-vascular lesions in children

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Background: The EDWARDS VALEO LIFESTENT® is a stainless steel, premounted, open cell stent. Easy dilation to large diameters and low profile are advantages in growing children. Radial force is poor.

Methods and results: between 4- 2011 and 12- 2012, 37 VALEO® stents were placed during 31 procedures, including 28 transcatheter (group 1) and 3 peroperative procedures (group 2). Data were retrospectively analyzed. Median age at implantation was 2.8 years (5 days–23 years), weight 10 kg (2–53). Indications were: pulmonary artery stenosis in 18, pulmonary vein stenosis in 1, sub-hepatic vein thrombosis in 1, ductus arteriosus stenting (hybrid approach) for hypoplastic left heart in 6 and for interrupted aortic arch in 2 patients.

In group 1, access was femoral in all except 5 (4 jugular, 1 transhepatic). Stent placement was achieved in all but 1. Predilation was performed in 12, postdilation in 4. Immediate results were satisfactory in all but 1 requiring covered stent placement for stent fracture and vessel tear. Acute complications were hemoptysis in 2, reperfusion edema in 1 and stent dislodgement from balloon in 2 (1 stent advanced without long sheath). Median follow-up reached 2.4 months (1 day–16 months). Eight patients were recatheterized, a median of 2 months after initial procedure. All stents remained fully patent, except 1 ductal stent obstructed due to neointimal proliferation. Surgery performed in 4 patients (interval 1.9 to 3.4 months), showed completely endothelialized and patent stents.

In group 2, stents were secured with a single proximal stitch and flared at the proximal end. All 3 had early post-op recatheterization (median 6 months), with balloon redilation to achieve better wall apposition.

No obstructive stent fractures were seen on chest X-ray, CT scan or fluoroscopy performed in 50% of the patients during follow-up.

Conclusion: the VALEO® stent is a useful stent in growing children. Low radial force is counter balanced by high flexibility, allowing implantation in distal and tortuous lesions. Early fractures may occur. Caution should be taken when advanced

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

without a long sheath as dislodgement is possible. When used per-operatively, early re-catheterization is warranted to improve wall apposition. Longer-term follow-up is needed.

O9-1
The Incidence of Unrecognised Life-Threatening Congenital Heart Disease in Newborns Discharged from Hospital in Scotland

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Aims: To determine the proportion of infants with life-threatening congenital heart defects being discharged from hospital before diagnosis, over a 10 year study period in Scotland.
Methods: Retrospective data analysis of all infants born from 1st January 2001 to 31st December 2010 in Scotland with prenatal, postpartum, postdischarge or post-mortem diagnosis of life-threatening congenital heart disease.
Results: 491 infants were born with life-threatening congenital heart defects in Scotland from 2001–2010. In 98 cases the diagnosis was made after the infant was discharged from hospital, equating to 20.0% of all diagnoses. Of these 98 infants, 6 (6.1%) were dead, 30 (30.6%) were critically ill, 60 (61.2%) were unwell and only 2 (2.0%) were well at the time of presentation.
Conclusions: 20.0% of all diagnoses of life-threatening congenital heart disease in Scotland from 1st January 2001 to 31st December 2010 were made after the infant was discharged from hospital. In 98% of these cases, the infant was either unwell, critically unwell or deceased at presentation. As clinical examination of newborns continues to perform poorly in identifying infants with life-threatening congenital heart disease, improvement in detection is likely to come from other sources: increasing antenatal diagnoses, educating health professionals about the limits of physical examination and implementing routine pulse oximetry in the postnatal period.

O9-2
Heart Catheterization (HC) complications in children with Pulmonary Hypertension (PH) as reported from the global TOPP Registry (Tracking Outcomes and Practice in Pediatric PH)

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Objectives: PH is a significant cause of morbidity and mortality in children. Hemodynamic assessment via HC remains the "gold-standard" confirming diagnosis and evaluating disease severity. We collected data from the global pediatric TOPP registry regarding complications associated with HC in pediatric PH.
Methods: Of 568 patients (age $\leq$18 years at diagnosis) enrolled in TOPP (HC required to confirm diagnosis; 34 sites, 20 countries) between 1/2008–2/2012, 486 patients (86%) fulfilled the criteria to confirm PH (HC: mean pulmonary artery pressure $\geq$25 mmHg, pulmonary capillary wedge pressure $\leq$12 mmHg, pulmonary vascular resistance index $\geq$3 WU*m^2).
Results: 908 HCs were performed in 555 patients (554 diagnostic; 354 follow-up HCs; conscious sedation in 257 (46%), general anesthesia in 291 (54%)). PH etiology: 263 (47%) idiopathic/familial, 202 (36%) congenital heart disease (CHD) associated, 57 (10%) chronic lung disease. 32 patients had either associated pulmonary arterial hypertension other than CHD (n = 27), or Venice Group 4 (n = 2) or 5 (n = 3). Clinically significant complications were reported in 37 patients (6.7% of 555): cardiac arrest (n = 5), systemic hypotension requiring intervention (n = 17) and PH crises (n = 10). Frequency of complications was similar in 16 follow-up HC (6.8% of 325 patients). Five HC-related deaths were reported in TOPP (0.6%). Factors identified with increased risk for complications included younger age ($<$1 yr 8/73 [11%]; 1–2 yrs 4/46 [8.7%]; 2$\leq$12 yrs 16/288 [5.6%]; $\geq$12 yrs 9/147 [6.1%]) ($P < 0.001$, Fisher's Exact test) and worse WHO functional class (FC): FC I 3/72 [4.2%]; FC II 14/259 [5.4%]; FC III 15/185 [8.1%], and FC IV 5/38 [13.2%] ($P < 0.001$, Fisher's Exact test). Study results are limited as some patients who died during/shortly after HC may not have been included in TOPP due to its design (informed consent required).
Conclusions: In TOPP, $<$1% HC-related mortality was reported in $>$900 HCs. Complications occurred in 6.7% of procedures and increased with lower age and worse WHO FC. HC is required to establish valid hemodynamic data for confirmation of diagnosis and evaluation of treatment. To minimize complications, expertise and experience in the pathophysiology and clinical management of pediatric PH are paramount for all members of the pediatric PH team.

O9-3
Subtype mitral stenosis/aortic stenosis is a risk factor for cerebral thromboembolic events in patients with hypoplastic left heart syndrome (HLHS)

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Introduction: In patients with HLHS the risk of thrombosis is increased. Only a few case reports exist speculating that the native aortic root is a potential source of thrombus formation and consequently cerebral embolism. We retrospectively analysed data on cerebral thromboembolic events (CTE) in our patient cohort focussing on thrombus formation in the native aortic root.

	n	Thrombus in NAR and/or CTE	%	native aortic root/BSA [mm/m^2]
overall	187	9	4.8	23.8 (9.0–57.3)
MA/AA	74	0	0	19.6 (9.1–39.5)
MS/AS	54	9	16.6	26.1 (12.2–57.3)
MS/AA	42	0	0	21.2 (12.9–35.3)
MA/AS	17	0	0	29.3 (17.9–37.8)

Methods: Data of 187 survivors with HLHS (born 1996 to 2012) after hemifontan or fontan operation were analysed. All patients received anticoagulation treatment, low dose aspirin was given as standard. CTE were identified as clinical signs of stroke and infarction seen in cerebral imaging. Lesions suggestive of diffuse ischemia were excluded as well as CTE that occured after

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

resuscitation, during sepsis, or with known thrombophilia. Size of aortic root was measured in angiocardiography.
Results: CTE occured only in the anatomic subtype of mitral and aortic stenosis (MS/AS; n = 7) (P = 0.0001) and in two of them a thrombus in the native aortic root could be identified. In another 2 patients with this subtype a thrombus in the native aortic root was found without signs of cerebral embolism. The size of the native aortic root was significantly larger in patients with aortic stenosis than aortic atresia (P < 0.001, available in 88 patients).
Conclusion: CTE and thrombus formation in the native aortic root occurred in nearly 17% of HLHS survivors who belonged all to the anatomic subtype of MS/AS. Larger anatomical size of the native aortic root as well as abnormal flow characteristics within this area may contribute to the increased risk for cerebral embolism in this subtype. It needs special attention during echocardiographic follow up and requires optimal anticoagulation treatment.

O9-4
Early remedial services use in children with transposition of the great arteries: prevalence and associated factors

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Objectives: There is growing concern regarding neurocognitive screening and remediation strategies for children with congenital heart disease (CHD). Some developmental delays and cognitive impairments may be observed before school entry and may require early preventive or treatment interventions. No data is currently available on the early utilization of these services after CHD. As early remediation may be essential for better long term prognostics, the objective of this study is to characterize the prevalence of early remedial service use and its associated demographic, medical and cognitive factors in children aged 4 to 6 years with corrected transposition of the great arteries (TGA).
Methods: Neurocognitive outcomes after TGA, the prevalence and characteristics of children's educational and/or rehabilitation services utilization were prospectively evaluated. Forty-five eligible patients (mean age = 5 y 4 mo; 67% males) underwent formal neuropsychological testing including IQ and a comprehensive battery of executive functions (EF) including motor and interference control, short-term memory and working memory as well as cognitive flexibility. Parental reports on the children's behavior and executive functions were also evaluated. Demographic factors, pre-operative, intra-operative and post-operative factors as well as cognitive factors were examined according to the current use of remediation.
Results: Twenty-four (53%) patients were receiving remedial services that included educational supports, speech, psychology and occupational therapy and neurological follow-up. Male gender (p = 0.04), a postnatal diagnosis of TGA (p = 0.03) and a longer postoperative ICU stay (p = 0.04) were significantly associated to remediation use. Children currently receiving remediation had lower EF scores (ps < 0.01), had more severe EF deficits as observed by formal testing (p = 0.001) and were rated by their parents as having more behavioral daily-life difficulties (p = 0.01). However, in the group without remediation, 13 children (43%) also displayed EF deficits rated as moderate to severe.
Conclusions: Educational and rehabilitation resources are frequently used by young children with TGA. Associated demographic (male gender) and medical factors (postnatal diagnosis of TGA and longer postoperative ICU stay) could help identify children at higher risk for neurocognitive delays. Evaluation of EF from an early age is necessary as it may influence prompt referral for remediation.

O9-5
Long-term survival and reinterventions after surgical correction of truncus arteriosus: a population based study

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Introduction: We evaluated long-term survival and reinterventions of patients operated for truncus arteriosus (TA) in Sweden.
Methods: Patient files were studied and survival was cross-checked against the National Population Registry in Sweden as of Jan 1st 2012. From Jan 1st 1994 to Dec 31st 2011, 80 patients (42 girls/38 boys) with TA were operated in Sweden. Median age and weight at first surgery was 29.5 days (3–510) and 3.4 kg (2.0–8.5). Mean gestational age was 38.5 ± 2.4 weeks with 15 (18.8%) born prematurely. Mean cardiopulmonary bypass time and aortic cross clamp time were 177.2 ± 40.3 minutes and 83.8 ± 27.6 minutes, respectively. An aortic homograft was used in 35, a pulmonary homograft in 36 and a Contegra graft in six patients. The mean size of the original right ventricle to pulmonary artery conduit was 11.4 ± 1.8 mm. Interrupted aortic arch was corrected in 14 patients (17.5%) and surgery on the truncal valve was performed in 11 patients at the time of initial repair.
Results: One early and seven late deaths occurred (1.3% and 8.8%). Median follow-up time was 6.9 years (44 days to 18 years). Reinterventions were performed in 54 patients, altogether 167 surgical or catheter procedures. In 48 patients conduit replacement was performed with 14 needing a second conduit replacement. The first conduit replacement was done at a mean time of 4.3 ± 3.7 years after initial repair. Freedom from conduit replacement was 87.5%, 61.3% and 47.5% after one, five and 10 years. Surgery for pulmonary artery branch stenosis was performed at 21 occasions. Aortic valve replacement was performed in 21 patients. Freedom from any catheter treatment was 87.5%, 78.8%, 72.5% after one, five and 10 years. Ballon dilatation or stenting of the conduit was performed at 21 occasions and of a pulmonary artery branch at 28 occasions. Three patients had balloon dilatation or stenting of the aortic arch.
Conclusions: Despite early complex surgery and a high frequency of reinterventions, mid- and long-term survival after repair for truncus arteriosus was good in this complete 18-year national cohort.

O9-6
Effect of left ventricular outflow tract obstruction on long-term survival with childhood hypertrophic cardiomyopathy: impact of modern management

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Background: Studies from tertiary adult cardiology centres have suggested that the presence of left ventricular outflow tract obstruction (LVOTO) in hypertrophic cardiomyopathy (HCM) increases risk of sudden death and symptom progression, but it nevertheless remains controversial whether symptom-free LVOTO should be an indication for treatment. We have studied long-term survival on unselected paediatric HCM-patients to establish effect on long-term prognosis.
Methods: The complete cohort of patients diagnosed with HCM below age 19 yrs at all five of Sweden's regional centres of paediatric cardiology were collected (n = 117), and classified as LVOTO if there was a gradient >16 mm Hg at rest assessed with Doppler or cardiac catheterisation (n = 65). Management strategies in the early era (from 1970–1991) consisted of surgical myectomy for patients with symptoms on conventional doses of beta-blockers (or verapamil). From 1992 a more aggressive medical strategy was pursued with LVOTO treated with high-dose beta blocker therapy (propranolol equivalents of 6 mg/kg/day or more) with addition of disopyramide if gradient persisted, and myectomy (or short AV-delay pacing) was reserved for those with remaining gradients >50 mm Hg on maximal medical therapy, even if symptom-free. Follow-up of surviving patients ranged between 1-49 years (mean 13.4 yrs).
Results: Kaplan-Meier survival curves showed patients with LVOTO at rest having a significantly worse survival than those without with a hazard ratio of 3.29 [95% CI 1.44–5.37, p = 0.002]. Without LVOTO there was 88% 5-year survival, 80% 10-year survival and 80% 20-year survival. With LVOTO the 5-year survival was 78%, but thereafter the curves diverge further, with 70% 10-year survival and 51% 20-year survival. There is an obvious era-effect, with the hazard ratio for death being 2.97 [1.04–6.28; p = 0.04] in the 1970–1991, compared with 1992-present. In the early era 42% required myectomy, in the later only 29%. The only long-term survivors from the LVOTO group in the early era were patients whose gradients had been completely controlled either with medication, myectomy or pacing; in 7 high-dose propranolol and disopyramide had been added. After 1992 48% of patients received disopyramide.
Conclusions: LVOTO has serious impact on long-term prognosis, and aggressive management even of symptom-free LVOTO seems justified.

O9-7
Multidisciplinary approach to multivessel lesions in Takayasu's arteritis: the Gaslini Institute experience

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Introduction: Takayasu arteritis (TA) is a chronic inflammatory disease affecting the large arteries and their branches; its etiology is still unknown. In individuals suffering from TA, arterial inflammation progresses to stenosis and/or occlusion, leading to organ damage and affecting survival. The disease is often unrecognized, heterogeneous in presentation, progression and response to therapy. Most patients require repeated and prolonged treatment including aortic surgery or transcatheter procedures. TA usually affects young female individuals during the second and third decades of life but it has also been reported in young children. We describe our experience of eleven young patients with TA.
Methods: we retrospectively reviewed, as per the 2010 EULAR/PRINTO/PRES criteria, a cohort of 11 Caucasian patients (pts) (9 female, 2 male) referred to our Institution with TA over the last decade. The median age of the disease onset was 13.9 years (range 5.1–21 years). The arterial lesion was classified according to Hata et al. (1996) who suggested five types of vessel damage involved.
Results: According to TA-criteria we observed angiographic abnormalities (11pts), pulse deficit or claudication (5pts), discrepancy of blood-pressure (2pts), bruits (5pts), hypertension (3pts), abnormal acute phase reactants (11pts). In particular, the angiographic abnormalities were classified according to Hata criteria as follows: type I (1pt), Type IIA (3pts), Type IIB (1pt), Type III (1pt), Type IV (1pt), Type V (4pts). Medical treatment included steroids (11pts), Cyclophosfamide (7pts), Methotrexate (11pts) Infliximab (9pts), Adalimumab (4pts), Azathioprine (3pts). Surgery was required in 3 pts: kidney revascularization (2pts), ascending aorta replacement (1pt) associated with aortic arch angioplasty. Two additional artery renal percutaneous angioplasty was required for residual stenosis in one patient and for bypass failure in another patient. Seven patients are still in medical therapy, without symptoms.
Conclusions: Takayasu arteritis is a complex and rare condition requiring a multidisciplinary approach. Although medical treatment can sometimes improve the course of the condition, an invasive approach with surgery or transcatheter treatment could be required when the involvement of the major arterial system makes the patients susceptible to significant medical sequelae including stroke, hypertension, congestive heart failure, and myocardial infarction.

O9-8
Heart-lung Interaction in Infants with Heart Insufficiency after Cardiac Surgery

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Introduction: Heart-lung-interaction, particularly important in heart failure patients, is discussed on the basis of the existing literature and our own studies on infants after heart surgery.
Materials and methods: We investigated respiratory and circulatory conditions after extubation (1st study, n = 60), under post extubational non invasive ventilation (NIV) (2nd study, n = 48) and the effects of chest physiotherapy with expiratory flow acceleration (EFA) in ventilated patients (3rd study, n = 51). Parameters measured: respiratory: tidal volume (TV), inspiratory pressure (PIP), positive endexpiratory pressure (PEEP), lung compliance (C), airway resistance (R), functional residual capacity (FRC), circulatory: stroke volume (SV), cardiac output (CO), ventricular contractility (ICON), all measured by electrical velocimetry (EV), mitral velocity time integral (MVTI) measured by Doppler ultrasound, blood pressure, urine output, biological: Lactate, base excess (BE), brain natriuretic peptide (BNP), a hormonal marker for ventricular afterload.
Results: 1st study: BNP rises from 554 to 1165 pg/ml (p < 0.001), SV decreases from 5.2 to 4.7 ml (p = 0.006), pH moves from 7.41 to 7.38 (p < 0.001) with BE from 1.5 to –0.05 mmo/l (p = 0.005), indicating increased afterload, reduced SV and thus impact on metabolism. BNP cut off 379 pg/ml for negativation of BE and of SV 354 pg/ml.
2nd study: 6 h after extubation (patients < 5 kg): BNP increases from 716 to 1350 pg/ml (p < 0.001) without and from 867 to 1030 (p = ns) with NIV, cardiac output decreases from 0.70 to 0.54 l/min (p < 0.05) without NIV and remains stable at 0.7 l/min in the NIV group, indicating hemodynamic stability of extubated patients under post extubational NIV.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

3rd study: BNP increases from 305 to 358 pg/ml ($p = 0.001$) without and decreases from 391 to 358 pg/ml ($p < 0.001$) with physiotherapy. SV remains at 5 ml without physiotherapy whereas it rises from 5 to 6 ml ($p = 0.001$) corresponding to a significantly better left ventricular filling measured by MVTI. Respiratory parameters: significant amelioration of C, R and alveolar recruitment (FRC augmentation), correlating well with the hemodynamic parameters, suggesting a positive influence of chest physiotherapy on heart-lung-interaction.
Conclusion: Infants with postsurgical heart failure present with distinct heart-lung-interaction features that can be positively influenced by post extubational NIV and respiratory physiotherapy.

O10-1
Reduced exercise capacity in patients operated for ventricular septal defect

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Introduction: Ventricular septal defects (VSDs) are generally closed very simply, and postsurgical the patients are considered healthy with normal physical capacity. However, if this is actually true, is still not clarified, and is therefore the aim of this study.
Methods: We tested cardiopulmonary exercise capacity in 21 patients and 11 healthy control subjects on an ergometer cycle. Pulmonary ventilation and gas exchange were simultaneously measured breath by breath with Jaeger MasterScreen CPX®. Each test was performed as a maximal incremental test. The graded cycling test protocol was chosen individually to ensure test time to be approximately the same for all participants. During the test session respiratory gas exchange was measured along with heart rate, blood pressure, and EKG. Endpoints were: maximal oxygen uptake, maximal workload, and anaerobic ventilatory threshold. For the last-mentioned both absolute and relative thresholds were measured using V-slope. Before each test a spirometry was made to measure FVC, FEV1 and PEF.
Preliminary results: VSD patients had a median age at surgery of 2.6 years (1.5–4.1 years) and 21.1 years (19.8–23.2 years) at the time of examination. Compared to controls they had a markedly, impaired maximal oxygen uptake, median 38.0 ml $O2\,kg^{-1}\,min^{-1}$ (31.6–40.8 ml $O2\,kg^{-1}\,min^{-1}$) vs. 45.8 ml $O2\,kg^{-1}\,min^{-1}$ (41.1–49.9 ml $O2\,kg^{-1}\,min^{-1}$) in control subjects, $p < 0.01$. Furthermore, absolute and relative anaerobic thresholds were reduced in VSD patients, median 22.1 ml $O2\,kg^{-1}\,min^{-1}$ (17.5–25.9 ml $O2\,kg^{-1}\,min^{-1}$) and 60.0% (54.0–72.7%), respectively, vs. 33.5 ml $O2\,kg^{-1}\,min^{-1}$ (25.1–41.6 ml $O2\,kg^{-1}\,min^{-1}$) and 76.1% (64.0–86.4%), respectively, $p < 0.05$ for both parameters. Lastly, maximal workload were significantly reduced, median 3.2 W kg^{-1} (2.7–3.6 W kg^{-1}) vs. 4.1 W kg^{-1} (3.2–4.3 W kg^{-1}) in control subjects, $p < 0.01$.
Conclusion: Patients with a surgically closed VSD had a markedly reduced cardiopulmonary exercise capacity compared to healthy controls; findings include effort-independent measurements.

O10-2
The Current and Future Profile of the Adult Single Ventricle Patient Group

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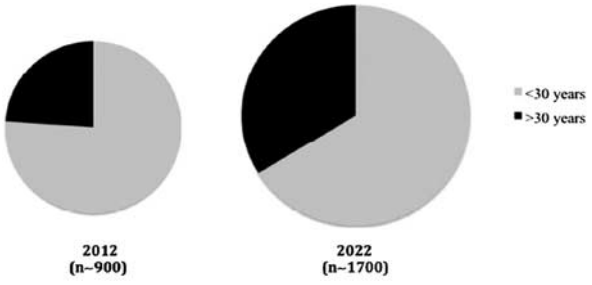
Introduction: The medium term outcome for patients born with single ventricle physiology has improved substantially with progress in Fontan surgery. However, deterioration in ventricular function is arguably inevitable. Medical therapies remain unproven, ventricular assist devices are expensive and transplantation is scarce and complicated in this cohort. It is difficult to address the clinical needs of this growing patient group when the size and status of the population is unknown.
Objective: This study aims to determine the size and status of the United Kingdom (UK) adult single ventricle population currently and in the next decade.
Methods: The number of surviving individuals (>16 years) with single ventricle physiology in a defined area of Northern England (resident population 2.9 million, 4.7% of UK) was identified primarily from the Northern Congenital Abnormality Survey (NorCAS). Conditions included double inlet ventricle, tricuspid and mitral atresia, hypoplastic left heart syndrome and others with Fontan surgery. Adult prevalence of single ventricle physiology was calculated. NYHA status was assessed from last clinical contact. The current paediatric population (5–15 years) was similarly determined. The UK population was extrapolated and population growth predicted by applying 10-year mortality, based on literature, to the defined populations. Migrants in and out of the region were excluded: extrapolation thus led to the lowest possible UK adult population estimates.
Results: 46 adult single ventricle patients were identified in the NorCAS region. Principal diagnoses included tricuspid atresia ($n = 16$) and double inlet ventricle ($n = 15$). The majority had undergone total cavopulmonary connection ($n = 30$). Few were >40 years ($n = 8$). All patients over 30 years were NYHA class 2 or greater ($p = 0.018$). Regional adult and live birth prevalence of single ventricle physiology were 2 per 100 000 adult population and 35 per 100 000 livebirths respectively. A current UK caseload of over 900 adults increasing to over 1700 patients by 2022 was estimated.
Conclusion: The UK single ventricle population will consist of approximately 1700 adults at the end of the next decade. Data on functional status supports symptomatic decline over time. Detailed, multi-centre study is required to address availability, applicability and utilization of resources.



Current and predicted United Kingdom adult single ventricle population.
190x114mm (300 x 300 DPI)

O10-3
Hypoplastic Left Heart Syndrome: an audit of clinical outcomes in patients surviving beyond the age of 10 years

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Introduction: Hypoplastic left heart syndrome (HLHS) has seen dramatic shifts in management over the past two decades with increasing numbers of children surviving surgical palliation. However, little research has been conducted into the long-term outcome functional status of these individuals.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Methods: Audit of clinical outcome beyond the age of nine years at a single tertiary centre based on care record review.
Results: A total of 105 patients underwent staged Norwood palliation at our centre more than nine years ago (pre 2003) of whom 58 are still alive. The median age at time of audit was 12 years (range 9–17). All case records were available for review. The median height lay on the 9th centile, and median weight on the 25th centile. Exercise tolerance was subjectively reduced in 42 (72%) of our patients and formal ETT/CPET, where performed, revealed 45–85% predicted V¬max across the population. 39 patients (67%) were referred to medical services with extra-cardiac symptoms (72%) of these with more than one complaint. 26% of the total population complained of respiratory symptoms including wheeze, chronic cough and recurrent RTI. 23 patients (40%) had concerns over their educational performance, also, 4 (7%) had been diagnosed formally with ADHD and 4 (7%) with an autistic spectrum disorder. 7% of patients suffered some form of imaging-confirmed cerebral infarction, 3 (5%) of our patients were registered partially sighted and 2 patients (3%) were registered as deaf. Only 9 patients (16%) had no documented referral to medical/psychiatric services, however even within this group, 6 had reduced exercise tolerance and 4 of these had made an application for, or were in receipt of, a Disability Living Allowance.
Conclusions: Survivors of HLHS surgery appear to be at risk for adverse clinical outcome with adverse developmental and psychological outcome particular causes for concern. We aim to follow up this pilot study with further controlled studies to elucidate the impact of HLHS on quality of life in patients surviving to adolescence and beyond.

O10-4
Long-term survival and functional status of adult patient with Eisenmenger Syndrome

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In the context of new target therapies, this study aimed to assess the functional status and long-term outcomes of patients with Eisenmenger Syndrome reaching adulthood.
Material and methods: This is a single-centre retrospective review of all patients diagnosed with Eisenmenger Syndrome. Demographics, clinical data, underlying cardiac disease, functional status, therapeutics and outcomes were collected.
Results: 159 patient were included (94 females: 59%), aged 27.7 ± 14.8 years at end-follow up, and 60 with Down syndrome (38%). Underlying cardiac disease was: AVSD in 30%, VSD in 35%, ASD in 9%, PDA in 5%, associated shunts in 5%, complex CHD in 10%, left heart obstruction in 2.5%, pulmonary veins anomaly in 2.5% and TGA in 1%. CHD was native in 122 cases (77%), 7 had palliation (4%) and 30 complete repair (19%). Pulse oxygen saturation was 84 ± 12% (range 44 to 98%), lower in non-operated or palliated cases (81%) than in repaired cases (92%, $p = 0.002$). Patients were in NYHA class I (18%), class II (42%), class III (37%) or IV (3%), not different with previous repair or not. Target therapy agents were given in 35% of the cases (1 agent in 20%, 2 associated in 13%, intravenous epoprostenol in 1.5%). Death occurred in 26 patients (16%) at the age of 29.3 ± 17.8 years. Complications occurred in most of the cases (64%) including: hemorrhages events, syncopes, thrombo-embolia, cerebral abscess, infective endocarditis, heart failure or arrhythmias. NYHA class did not differ between patients with or without target therapy. SpO2 was 82% in untreated cases compared to 86% in treated cases (NS). Survival rates were: 98% at 10-years, 93% at 20-years, 87% at 30-years, 83% at 40-years, 73% at 50-years and 53% at 60-years of follow-up. Survival was lower in Down patients ($p = 0.0023$), in males ($p = 0.04$) and higher better up to 50-years in patients under target therapy ($p = 0.05$).
Conclusion: Survival rates of adult patients with Eisenmenger Syndrome seem to improve up to 50-year of age with target therapy agents. These results have to be confirmed by larger scale multicentre studies.

O10-5
Frequency of Miscarriage, Stillbirth and Pregnancy Termination in Women with Congenital Heart Defects in Germany, Hungary and Japan

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Introduction: The 2011 'ESC Guidelines on the management of cardiovascular diseases during pregnancy' define maternal predictors for neonatal events (preterm birth, small for gestational age, respiratory distress syndrome, cerebral haemorrhage, foetal/neonatal death) in pregnancies in women with heart disease. It is unknown whether these predictors also lead to an increased number of miscarriages, stillbirths and terminations of pregnancy (TOP), particularly regarding patients with congenital heart defects (CHD). In the general population, miscarriages and stillbirths occur in 15–20%. In the participating countries, stillbirth occurs in below 0.5% of all pregnancies.
Methods: In a multicentre cross-sectional questionnaire-based study, 634 women with CHD (Germany 61%, Hungary 24%, Japan 15%) were surveyed over a period of twelve months concerning courses of pregnancy. 309 out of 634 women were pregnant at least once (578 pregnancies). Patients were grouped into those with and those without existing maternal predictors for neonatal events. The predictors were NYHA > II or cyanosis, maternal left heart obstruction, smoking during pregnancy, multiple gestation, use of oral anticoagulants during pregnancy, mechanical valve prosthesis. The outcomes were miscarriage/stillbirth and TOP (miscarriages and stillbirths were grouped together).
Results: In 75 women with predictors, a total of 141 pregnancies occurred (group I, 24%); In 234 women without predictor, a total of 437 pregnancies occurred (group II, 76%). There have been no significant differences between the participating countries.

	Group I (n=141) n (%)	Group II (n=437) n (%)	p
Miscarriage/ stillbirth	27 (19.1)	70 (16.0)	0.536
TOP	22 (15.6)	24 (5.5)	0.002
Combined	49 (34.8)	94 (21.5)	0.042

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Conclusions: Underlying maternal predictors for neonatal complications do not lead to a significantly higher number of miscarriages or stillbirths. However, TOP occurred significantly more frequently in this group. In presence of maternal predictors for neonatal events pregnancies in women with CHD are less likely to be successful.

O10-6

Characteristics and Survival of Adult Patients with Pulmonary Arterial Disease Associated with Congenital Heart Disease: COMPERA-CHD Registry

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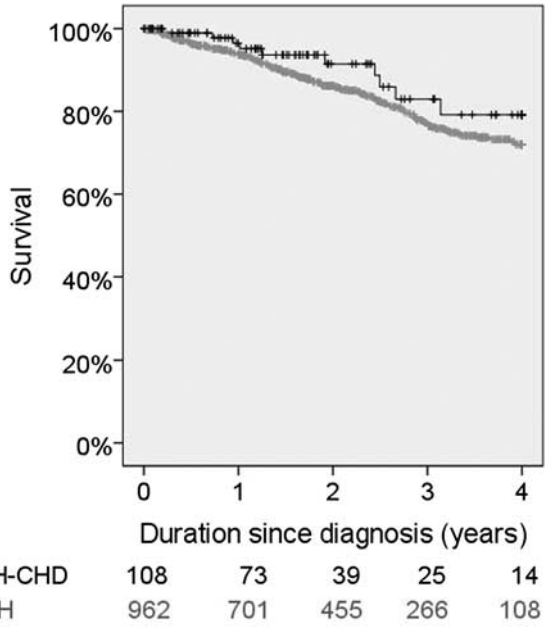
We aimed to document real-life data of adult patients with pulmonary arterial hypertension (PAH) as an important complication in patient with congenital heart disease (CHD, Dana Point Group 1.4.4.).

The COMPERA registry (Prospective Registry of Newly Initiated Therapies for Pulmonary Hypertension, ClinTrials.gov Identifier NCT01347216) documents adult patients with pulmonary hypertension, including those with PAH-CHD, if they are treated with disease-specific drugs (endothelin receptor antagonists ERA, phosphodiesterase-5-inhibitors, prostacyclins). Further, patients with Eisenmenger physiology can be included whether drug treated or not, as well as patients with pulmonary vascular dysfunction or relative PAH (e.g. after cavo-pulmonary anastomosis PCPC, modified Fontan operation TCPC).

As of 16 November 2012, a total of 3642 PAH/PH patients were included in COMPERA. 288 had PAH-CHD (8%), and were documented in 30 centres in 7 countries.

Detailed information from a special CHD case report form in 104 patients (97 Eisenmenger, 7 Fontan) are presented in the following. Mean follow-up was 40 ± 30 (on average 4.2 visits per patient). Patients were 38.6 ± 13.6 years old, in 39% males, in NYHA functional class I 2%, II in 37%, III in 59%, IV in 3%, mean 6-minute walking distance was 370 ± 102 meters. Quality of life on the EQ-5D visual analogue scale (0 extremely bad, 100 excellent) was 51 ± 21 points.



The majority of these 104 CHD patients in the database were treated with PAH specific drugs, usually as monotherapy (80%), less frequently as combination therapy (9%), or without drugs (11%). ERA were given in 45%, PDE-5 inhibitors in 51%, prostacyclines in 1%. Oral anticoagulation was given in only 20% of patients (Eisenmenger 16%, Fontan 86%).

Of the CHD patients who received the PAH diagnosis after start of the registry (1. May 2007), 10 died (mortality rate 9.3%, compared to idiopathic PAH IPAH 15.9%), and the mean 4-year (Kaplan Meier) survival estimate was 79%.

Current COMPERA data indicate that CHD patients compared to IPAH patients

- are younger and have higher exercise capacity
- nonetheless have a reduced quality of life

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

- usually are treated with monotherapy
- receive substantially less frequently oral anticoagulation
- have higher survival rates in line with previous reports.

O10-7

Aortic flow abnormalities contribute to the aortopathy in bicuspid aortic valve disease

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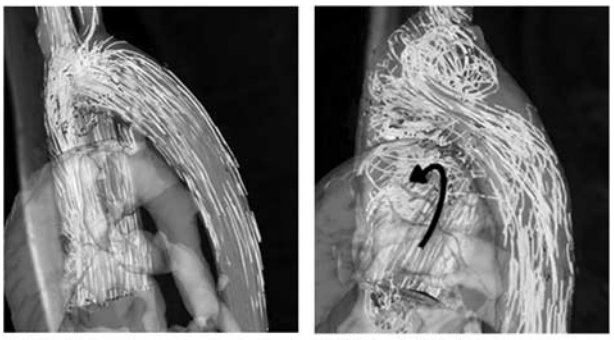
Background: Bicuspid aortic valve disease (BAV) is associated with aortic dilation. We examined the impact of flow abnormalities on measures of vascular function in the ascending aorta.
Methods: We prospectively enrolled 142 participants (95 patients with BAV and 47 healthy volunteers [HV]); mean age 40 years (range 8–70). Cardiac magnetic resonance was used to measure arterial strain, distensibility, pulse wave velocity (PWV), rotational flow values to quantify helical flow, flow angle and wall shear stress (WSS).
Results: Both BAV and HV had similar aortic diameters at the sinuses of Valsalva and the proximal descending aorta. BAV had significantly larger aortic diameters at the sinotubular junction (16.6 vs 15 mm/BSA, $p = 0.001$) and the ascending aorta (18.2 vs 15.2 mm/BSA, $p < 0.001$). There was no difference in PWV across the arch compared to HV (median 4.5 vs 4.7 m/s, $p > 0.05$), distensibility in the ascending aorta (mean 4.1 vs 4.6 1/mmHg, $p > 0.05$) or arterial strain (mean 0.19 vs 0.21, $p > 0.05$). The most common flow pattern in our BAV cohort was a right-handed helical flow in 72%. A normal laminar flow pattern was observed in 11%, complex disorganised flow in 13% and left-handed flow in 4%. The normal flow pattern group had similar ascending aortic diameter, rotational flow values, flow angle and total WSS compared to the HV group ($p > 0.05$). The right-handed flow group had significantly higher ascending aortic diameter (18.3 vs 15.2 mm/BSA, $p < 0.001$), rotational flow values (31.7 vs 2.9 mm^2/s, $p < 0.001$), flow angle (23.1 vs 7.0 degrees, $p < 0.001$) and total systolic WSS (0.85 vs 0.59 N/m^2, $p < 0.001$) compared to the HV group.
The left-handed flow group only contained 4 cases but showed a trend towards higher ascending aortic diameter (20.0 mm/BSA), rotational flow values (−49.7 mm^2/s), flow angle (30.0 degrees) and WSS (1.18 N/m^2) compared to the HV group and right-handed flow group.
Conclusion: Marked differences in ascending aortic flow abnormalities occur with BAV but there were no differences in distensibility or pulse wave velocity. These findings suggest that the ascending aortic flow pattern is an important determinant of adverse aortic outcome.



a) normal flow b) right-handed helical flow

O10-8

Causes of late death after paediatric cardiac surgery in patients with univentricular heart defects

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Objectives: To identify those patient groups at risk for late mortality after surgery for univentricular heart defects by assessing the mode of their late deaths.
Methods: All 306 patients operated for univentricular heart defects at our institution before the age of 18 years between Jan 1st 1994 and Jan 1st 2009 were cross-checked against the Swedish National Population Registry on January 1st 2012 to reliably identify all dead patients. Of 83/306 deceased patients 41/306 (13,4%) had died more than 30 days after the last surgery and 19/306 (6,2%) had undergone a heart transplantation. 2/306 patients (0,7%) were lost to follow-up (emigration). The circumstances of late death were analysed by reviewing clinical charts and autopsy reports. The mode of death was defined as the condition that initiated a clinical course of deterioration leading to death rather than the pre-terminal event causing the patient to pass a terminal threshold in an already marginal physiological state.
Results: The mode of death was considered to be related to the underlying heart defect in 34 and most likely CHD-related in further 5 cases together accounting for 39/41 (95%) of all late deaths. Of the late deceased patients 29/41 (71%) had a systemic ventricle of right ventricular morphology. 23/41 (56%) patients died after the first palliation mainly due to shunt complications (7/23–30%), circulatory failure in the face of infection or pulmonary disease (6/23–26%) and acute, autopsy-negative circulatory collapse in an ambulatory setting (5/23–22%). Other causes included ventricular failure and myocardial ischemia. The main mode of death after the Glenn operation was ventricular failure (5/10) and after the Fontan operation thromboembolic complications in 2/4. 4/41 patients died due to complications after heart transplantation (3 malignancies, 1 rejection). Syndromes and/or relevant congenital defects were noted in 4/41 (9,8%).
Conclusion: Deaths in patients with univentricular hearts were nearly exclusively related to their heart defect and occurred predominantly after the first palliation. The main mode of death was a failing shunt circulation. The majority of patients hade a systemic ventricle of right ventricular morphology. Associated syndromes and/or congenital defects were relatively uncommon.

O11-1

Analysis of the aortic root in Tetralogy of Fallot patients undergoing early repair: form follows function

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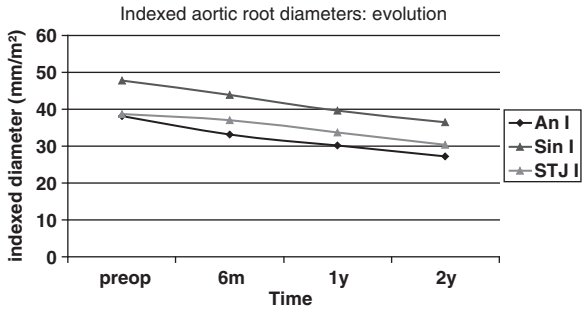
Objective: Aortic root dilatation remains of concern in patients late after repair for tetralogy of Fallot (ToF). The underlying mechanism is unclear - both hemodynamic and intrinsic wall abnormalities have been suggested. In a previous study we demonstrated that indexed aortic root diameters decrease progressively when correction is performed early in life. To test if aortic wall changes are intrinsic or acquired, a histological study was performed, focusing on the ascending aortic wall characteristics in infants at early repair.
Methods: In 24 of 31 consecutive infants (mean age 5,9 ± 4,9 months) undergoing ToF repair, full-thickness aortic biopsies were obtained from the proximal ascending aorta, and evaluated

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

histologically. The aortic root z-values and indexed diameters were then prospectively followed up to 2 years postoperatively.
Results: None of the aortic specimens showed signs of important medial degeneration, increased fibrosis, elastic fragmentation, mucoid accumulation or apoptosis. The aortic root was dilated in all infants at the time of repair (mean indexed diameter of annulus $38 \pm 5{,}7\ mm/m^2$, sinus $47{,}9 \pm 6\ mm/m^2$ and sinotubular junction $38{,}8 \pm 5{,}4\ mm/m^2$), and regressed significantly within 2 years (mean indexed diameter of annulus $27{,}9 \pm 5.1\ mm/m^2$, sinus $37{,}1 \pm 5.6\ mm/m^2$ and sinotubular junction $31{,}1 \pm 6{,}5\ mm/m^2$ at 2 years; $p < 0.0001$).
Conclusions: Infants with ToF undergoing repair around 6 months of age show no histological features of important intrinsic aortic degradation at the time of repair. Prospective echocardiographic follow-up demonstrates progressive and significant reduction in indexed aortic root diameters. These findings support a hemodynamic mechanism underlying late aortic root dilatation and favor a strategy of early surgical repair to prevent ongoing aortic dilatation.



O11-2
Hemodynamic effects of temporary right ventricular resynchronisation in children after surgery for tetralogy of Fallot

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Introduction: Right bundle branch block (RBBB) associated with right ventricular (RV) electromechanical dyssynchrony may contribute to postoperative hemodynamic impairment in tetralogy of Fallot (ToF). We thought to evaluate changes in haemodynamics and tissue oxygenation due to RV resynchronisation during the first 24 hours after surgery.
Patients and Methods: Arterial pressure, cardiac output (PiCCO) and tissue oxygenation (NIRS) were measured during baseline sinus rhythm and after RV resynchronisation using atrial-triggered RV free wall pacing in complete fusion with spontaneous activation in 10 patients (median age 8.7 months). Studied variables were compared in a cross-over design in four 5-minute intervals (baseline rhythm and stimulation 2x each) and results were averaged and statistically analyzed using RM ANOVA.
Results: Resynchronisation reduced the QRS complex duration from 96 (±12) to 64 (±8) ms ($p < 0.001$) and increased arterial systolic (median +4.3, range $+2.1 \mp 13.0\%$ $p < 0.001$), mean (median +4.0, range $+1.0 \mp 9.7\%$, $p < 0.001$) and pulse pressure (median +4.0, range $-2.9 \mp 22.6\%$, $p = 0.007$), improved left ventricular contractility (dP/dT max, median +4.6, range $-2.3 \mp 8.7\%$, $p = 0.002$) and splanchnic (renal) oxygenation (median +1, range 0 ∓ 3 saturation points, $p < 0.05$). There were no statistically significant changes of central venous pressure, cardiac output and cerebral oxygenation.
Conclusion: RV resynchronisation improved hemodynamic parameters and renal oxygenation in children early after surgery for ToF and may be an important part of low cardiac output management in selected patients.
Supported by the project (Ministry of Health, Czech Republic) for conceptual development of research organization 00064203 (University Hospital Motol, Prague, Czech Republic)

O11-3
Therapy of low cardiac output syndrome after cardiac surgery in infants and children: a double blind randomized study comparing Dobutamine and Milrinone

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Background: Low cardiac output syndrome (LCOS) after cardiopulmonary bypass is usually treated with intravenous inotropic and afterload-reducing agents. Dobutamine (D) is well established for prevention and therapy of postoperative LCOS, similar effects can be achieved with milrinone (M). The aim of this study was to compare safety and efficacy of D versus M, which to our knowledge has not been done in a similar population yet.
Methods: Fifty non-selected children, median age: 1.2 years (range 0.2–14.2), median weight 8.6 kg (range 3.4–35.5) with non-obstructive congenital heart lesions including single ventricles undergoing open-heart surgery were randomized to continuous infusion of D or M for 36 h after cardiopulmonary bypass. Maximum dosis: D 6 μg/kg/min, M 0.75 μg/kg/min.
Results: Need for additional inotropic support did not differ between the two groups (D 39% vs. M 33%, $p = 0.71$). Sodium nitroprusside was used significantly more often in the D group (42% vs. 13%, $p = 0.019$). Systolic blood pressure at 1 and 36 h after ICU arrival was higher in the D group (106 ± 18 vs. 94 ± 23 mmHg, $p = 0.042$ and 99 ± 13 vs. 92 ± 17 mmHg, $p = 0.024$). Similarly, early (8 h) postoperative heart rate was higher in the D group (143 ± 16 vs. 131 ± 26 bpm, $p = 0.039$). No significant differences were found in central venous oxygen saturation, serum lactate levels, urine output, duration of chest drains, and length of mechanical ventilation, ICU stay and hospital stay. The cardiac function evaluated by echocardiography was consistently good in both groups. Both drugs were well tolerated, no serious adverse events occurred.
Conclusions: Milrinone has at least an equal efficacy and safety as Dobutamine for the treatment of LCOS in paediatric patients undergoing heart surgery for congenital heart disease. Milrinone demonstrated a trend to be more efficient in afterload reduction and might have less chronotropic effects. An individual selection of the appropriate inotropic agent may be justified.

O11-4
Evaluation of potential risk factors for prolonged periods of decreased cerebral tissue oxygen saturation after the Norwood procedure for Hypoplastic left heart syndrome

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Objectives: Lower cerebral tissue oxygenation has been observed by near infrared spectroscopy during the early postoperative course after the Norwood procedure. We evaluated potential preoperative and postoperative risk factors for prolonged periods of decreased cerebral tissue oxygen saturation.
Methods: Cerebral (cSO2) and somatic (sSO2) tissue oxygen saturations and routine intensive care monitoring data were recorded from 68 HLHS patients for 24 hours before and 48 hours after the Norwood procedure. Average values of the last 4 preoperative hours (baseline) and of the first and last 4 postoperative hours (early and late course) were calculated. The absolute duration of cSO2 below 40% was evaluated and patients who had cSO2 values below 40% for 60 minutes or longer were classified to have sustained a prolonged period of decreased cSO2. Risk factors were evaluated with binary logistic regression and validated by bootstrepping.
Results: Baseline, early and late cSO2 values were $58 \pm 7\%$, $52 \pm 10\%$ and $61 \pm 7\%$. Early postoperative values were significantly lower compared to baseline and later course ($p < 0.001$). Postoperatively, cSO2 was <40% for 50 (0–1040) minutes. 32 patients had a cSO2 below 40% for ≥60 minutes. Preoperative cSO2 (OR 0.84 [0.74–0.94], $p = 0.004$), age at operation (OR 1.44 [1.05–1.97], $p = 0.027$) and postoperative diastolic blood pressure (OR 0.88 [0.78–0.99], $p = 0.038$) were associated with a cSO2 <40% for ≥60 minutes in a logistic regression model. After validation by bootstrapping preoperative cSO2 ($p = 0.001$) and postoperative diastolic blood pressure ($p = 0.023$) remained in the model. A trend was observed for age at operation ($p = 0.056$), but the mean cSO2 in the early postoperative course was lower in patients operated later than 4 days of life ($49 \pm 11\%$ vs. $55 \pm 8\%$, $p = 0.010$). BT-shunt size, duration of cardiopulmonary bypass and selective cerebral perfusion were not associated with a cSO2 <40% for ≥60 minutes.
Conclusions: cSO2 during the early postoperative course of the Norwood procedure is lower compared to preoperative baseline. Older age at the Norwood procedure and a longer preoperative period of poorly balanced circulations, indicated by lower preoperative cSO2, may predispose to a more vulnerable cerebral vasculature and an increased risk for a more pronounced decrease of postoperative cSO2.

O11-5
Reintervention Rate and Aortic Valve replacement in Critical Aortic Stenosis: Comparison of Patients who Underwent Primary Surgical or Balloon Valvuloplasty

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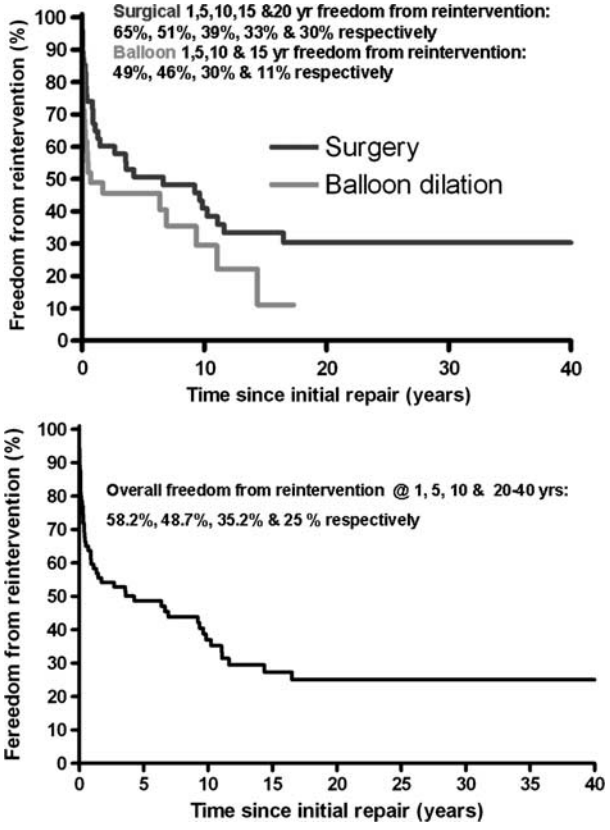
Introduction: Surgical and catheter intervention can be undertaken with initial success in patients with critical aortic stenosis (CAS). The majority require further interventions, long-term outcome and follow up remains uncertain.
Methods and Results: Over 40 years (1970–2010) 96 patients required intervention for neonatal (<30 days) CAS. Surgery (SX) was undertaken in 61 patients and balloon dilation (BD) in 35 patients. Before 2000, SX was more common (56/63 cases) while BD was favoured after 2000 (28/33 cases). There were 29 (32.9%) reported deaths. Twenty (22.7%) patients died without any further intervention, one patient had a heart transplant and another had a single ventricle palliation. Overall survival rate at 1, 5, 10, 15 and 20 years was 73%, 71%, 71%, 66%, and 66% respectively. Survival rates were better for BD patients (survival 5–20 years was 87.2% versus SX 5,10,15 & 20 yr survival rates 62, 62, 57 & 57%, $P = 0.009$). Among the 57 survivors, 52 (87%) had at least one reintervention. Sx 1,5,10,15 & 20 year freedom from reintervention was 65%, 51%, 39%, 33% & 30% versus BD 1,5,10 & 15 yr freedom from reintervention was 49%, 46%, 30% & 11% ($P = 0.01$). Further reintervention was required in 21 (36.8%). There were 33 aortic valve replacements (AVR) in 32 patients, 31 (35.2%) had a Ross or Ross-Konno procedure. Freedom from AVR at 5, 10 and 20 years was 78%, 63%, and 44%. The median time interval between initial procedure and AVR for the BD group was 1.32 years (mean 3.9 ± 4.7, range 1 day–14.4 years) this was shorter than for the SX patient group, 8.9years (mean 7.24 ± 5.1, range 1 day–16.5 years). For patients presenting for initial procedure since 2000, AVR was undertaken in 14/34, with a median time to AVR of 1.3 yrs.
Conclusions: This study highlights the need for repeat intervention on the aortic valve and the acute and long-term mortality associated with CAS. Catheter based intervention has become more common but is still associated with early mortality. Reintervention is highly likely in survivors, over 50% of survivors requiring surgical AVR, and a trend towards earlier surgical AVR at our institution in recent years.



O11-6
Analysis of pre-operative condition and interstage mortality in Norwood and Hybrid procedures for Hypoplastic Left Heart Syndrome using the Aristotle scoring system

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Introduction: The "Hybrid" procedure, consisting of surgical banding of the pulmonary arteries with intraoperative stenting of

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

the arterial duct, was developed as primary palliation in Hypoplastic Left Heart Syndrome (HLHS) avoiding the risks of cardiopulmonary bypass (CPB) and circulatory arrest. It is generally reserved for low birth weight, premature or unstable neonates in whom the risks of CPB are increased. Its role in unselected cases of HLHS has yet to be defined.
Methods: The pre-operative condition of all patients with HLHS who underwent either the Hybrid or the Norwood procedure for HLHS between 2005–2011 was analysed retrospectively, using a modified comprehensive Aristotle score (AS). We then compared the early (< 30 day) and inter-stage mortality for each cohort.
Results: Of 138 patients with HLHS, 27 had Hybrid and 111 underwent Norwood procedures. The Hybrid group had significantly higher scores (mean AS 4.1 vs. 1.8; $p < 0.001$); however there was no significant difference in the early post-operative mortality (< 30 days, 33% vs. 28%; $p = 0.64$) or overall inter-stage mortality (44% vs 37%, $p = 0.51$).
Conclusions: We conclude that the Hybrid is an appropriate choice for primary palliation of HLHS in higher risk patients, with comparable mortality to the Norwood procedure performed on a lower risk cohort. More prospective work is needed to establish whether the Hybrid is an alternative to the Norwood in all HLHS patients, and whether more complex outcomes linked to CPB, such as neurodevelopmental status, could be improved by taking this approach.

O11-7
Strategies for biventricular outflow tract reconstruction in complex forms of transposition (TGA) of the great arteries (TGA) with ventricular septal defect (VSD) and left ventricular outflow tract obstruction (LVOTO) – midterm results

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Objectives: The optimal surgical solution for the correction of complex forms of TGA with VSD and LVOTO is discussed controversially. Long-term outcome of the most widely used Rastelli procedure is unsatisfactory while experience with newer techniques such as the modified Nikaidoh procedure is still comparably limited. We present our single center experience with different surgical approaches to biventricular outflow tract reconstruction.
Methods: From 2000–2013, 42 patients with TGA, VSD and LVOTO have been operated in our institution. Modifications of the Nikaidoh procedure (aortic translocation n = 15, en bloc rotation n = 4, double switch with Senning procedure n = 2), the Rastelli procedure (n = 13) and arterial switch (ASO) with LVOTO relief (n = 8) were performed. Median age at operation was 9.5 months, median weight 8.7 kg. Sufficient pulmonary valve (PV) diameter with Z-score > −2 was present in 16 patients, in 10 of them the entire PV or PV leaflets could be preserved (8 ASO, 2 Nikaidoh).
Results: Median follow-up was 2 years (range 0.04–12.5). There were 3 early deaths (7% early mortality, Nikaidoh n = 2, ASO n = 1) and 2 late deaths (5% late mortality, Rastelli n = 1, Nikaidoh n = 1). On discharge, good cardiac function and outflow tract performance was observed. All autografts after modified Nikaidoh remained intact during follow up. Reoperation for LVOT reobstruction was required in one Rastelli patient while no LVOT reoperations were necessary after Nikaidoh or ASO. Freedom from significant reobstruction of the RVOT (dp >50 mmH) at 5 years was 100% for ASO, 78% for Nikaidoh and 39% for Rastelli, respectively. Subsequent RVOT reoperation was necessary in 2 Nikaidoh patients (10%) and 3 Rastelli patients (23%).
Conclusions: All surgical approaches to this complex lesion offer good early functional outcome. Midterm results of the Nikaidoh procedure are superior to the Rastelli procedure, though long-term results have to be evaluated. Inevitable conduit failure remains a major concern and cause of reoperation in both techniques. En bloc rotation preserves PV growth potential and may reduce need for RVOT reoperation. Given the favorable reoperation rates, the performance of ASO with LVOTO relief or en bloc rotation should be considered in cases with PV Z-Scores < −2.

O11-8
Pulmonary artery growth and interventions after bilateral branch pulmonary arterial banding

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Background: Bilateral branch pulmonary artery banding (bilatPAB) is used to control pulmonary blood flow in the hybrid palliation for ductus dependent lesions. The long-term impact on pulmonary artery growth and the need for interventions has not been assessed.
Methods: We performed a retrospective review of all newborns (2001–2012) undergoing bilatPAB at a single institution (n = 46); these were compared to a random group of contemporaneous newborns undergoing the Norwood operation (n = 48). Pulmonary arterial growth as determined by angiography prior to hemi-Fontan and Fontan operation and the need for pulmonary artery interventions (surgical arterioplasty, balloon angioplasty, or stent implantation) was assessed.
Results: Left and right pulmonary artery were banded to the same diameter in 34 (73.9%) of 46 patients; 7 patients required subsequent band adjustment. Bands were in place for a median of 79.5 days (1–229 days). Mean pulmonary artery branch diameter prior to hemi-Fontan was smaller in the bilatPAB group (RPA: 4.0 mm vs. 5.2 mm, $p = 0.03$ and LPA: 3.7 mm vs. 4.1 mm, p = NS). There was a trend toward increased likelihood of lobar branch obliteration in the bilatPAB group (hazard ratio 2.8, 95% CI 0.5–15.2). Surgical and catheter interventions were more frequent in the bilatPAB group compared to the Norwood group ($p = 0.02$; hazard ratio 1.9, 95% CI 0.8–4.2). The bilatPAB patients frequently required multiple interventions (9/46 vs. 2/48, $p = 0.02$). Duration of banding, and the need for band adjustment did not appear to affect the likelihood of subsequent intervention.
Conclusions: Bilateral branch pulmonary artery banding appears to reduce pulmonary artery growth and carries the risk of lobar branch obliteration resulting in increased need for pulmonary artery interventions.

PW1-1
Percutaneous closure of coronary artery fistulas in paediatrics

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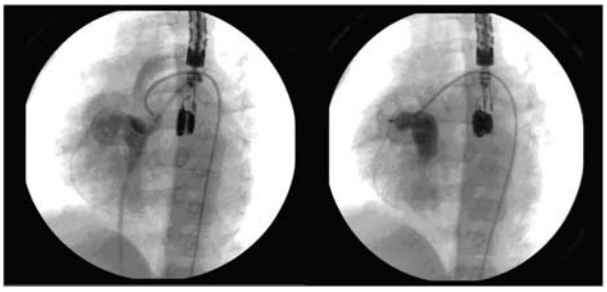
Introduction: The prevalence of coronary fistulas is poorly known in the pediatric population. However, complications can be serious

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

with : heart failure, sudden death, arrhythmias, endocarditis. In adults percutaneous treatment is an alternative to surgical treatment. In pediatric population, a few publications exist on the feasibility and effectiveness of this treatment.
Objective: Evaluation of the feasibility of percutaneous closure of coronary fistulae in paediatrics, follow up and occurrence of complications.
Materials and methods: Retrospective observational multicenter national study including all patients under 16 who underwent transcatheter closure of a congenital coronary fistula (complex cardiopathies were excluded).
Results: Population: 61 patients (36 girls, 25 boys), mean age at diagnosis 2.6 years.
Initial signs: precordial murmur (90%), congestive heart failure (11%), non specific ECG abnormalities (4,9%), left or right ventricular dilatation (31%).

ORIGIN	Left coronary (28)	Right coronary (28)	Double (5)
DRAINAGE SITE	Right atrium (12) Right ventricle (12) Left atrium (2) Left and right ventricle (1) Pulmonary artery (1)	Right atrium (12) Right ventricle (13) Pulmonary artery (2) Left atrium (1)	Right ventricle (4) Pulmonary artery (1)

At procedure completion : 35 patients had no resisual flow through the fistula, 16 a trivial flow, 5 a moderate and 5 a persistent flow (failure).
Efficiency: complete occlusion at hospital discharge in 82%.
Complications: no death. 3 transient STEMI, 3 Coil embolization, 1 ventricular fibrillation recovered, 5 procedures failures including an urgent surgical closure (refractory heart failure).
Combination therapy: antiplatelet (46%), mean duration 4.1 months.
Evolution: No recanalization in 87% at 2.3 years (n = 39), 4 reoperations (one surgery). No cardiovascular complication.
Conclusion: this largest world series shows that the percutaneous closure of coronary fistulas in the pediatric population appears to be effective and safe.



PW1-2
Radiofrequency Catheter Ablation – A Novel Concept For The Treatment of Hypertrophic Obstructive Cardiomyopathy
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Introduction: The subaortic obstruction observed in hypertrophic obstructive cardiomyopathy (HOCM) can be relieved by either surgical myectomy, alcohol septum ablation or by pharmacological treatment. Reduction of septal hypertrophy may also be achieved through radiofrequency catheter ablation (RFCA), which is a novel strategy in the management of HOCM. We report our preliminary experience using this technique in children and adolescents.
Patients: Four patients aged 2 to 16 years (weight 14.6 to 90 kg) with HOCM and left ventricular outflow tract (LVOT) obstruction were treated. One patient had previously undergone surgical myectomy. All patients were on ß-blocker medication. The median subaortic pullback gradient ranged from 59 to 100 mmHg.
Method: RFCA in our patients was performed at the University Hospital of Cologne. Imaging of the LVOT was performed at the beginning of the procedure by left ventricular angiography. Subsequently, the most pronounced septum prominence was localized and, using 7F or 8F ablation catheters, radiofrequency was applied at a power setting of 60 Watts over 20 to 120 seconds. Transesophageal echocardiography was used to monitor acute pressure gradient changes during the procedure and to exclude pericardial effusion.
Results: RFCA was effective in all patients. No complications were reported. Pressure gradients measured invasively decreased by 25 to 55 mmHg. Follow-up visits showed further regression of pressure gradients by another 10 to 30 mmHg. Left bundle branch block did not occur in any patient.
Conclusion: RFCA is a novel therapeutic strategy in the management of HOCM and may be used as an alternative to surgical myectomy. In addition to acute gradient reduction, pressure relief of the left ventricle is observed progressively within 6 to 12 months following the procedure, which is assumed to be caused by myocardial involution of the treated region.

PW1-3
Intermediate and late outcome of Fontan patients after fenestration closure – single institutional study
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Introduction: The study was designed to determine changes in oxygen saturation (satO2) and hemodynamic parameters after closure, and character of complications during follow up. Secondary outcome was to evaluate potential risk factors for late complications.
Methods: Retrospective analysis of medical records approved by single institution. The study sample consists of all patients (pts) who accomplished inclusion criteria and underwent fenestration closure in the period of 1997 to 2011.
Results: In this period were performed 79 procedures in 73 pts. In 3 of the pts have been performed 2 following procedures because of the presence of residual leak or another fenestration. The most common diagnosis were hypoplastic left heart syndrome, tricuspid and pulmonary atresia (55.6%) with their variantions. Relation between systemic right and left/single ventricle (SV) was 28 to 44 (0.64). Median age of operation was 43 months, lateral tunnel represented 33.3% and extracardiac conduit 66.6%. The median of follow up postoperatively to fenestration closure was 33 months, of postclosure follow up 56 months. There was no significant difference in satO2 changes between the diagnoses, neither systemic RV and LV/indetermined ventricle. The immediate postoclussion mean baffle pressure increased significantly from 12.1 to 13.3 mmHg. Additional procedure was performed in 9 pts (12.5%). Hemodynamically trivial residual leakage was present in 16 pts (22.2%). Procedure-related complications were present in 34 pts (47.2%) - arrhythmias (11.1%) and transient SV dysfunction (13.9%). During follow up, in none pt was necessary to remove device or restore fenestration. Late complications occured in 19 pts (26.4%). The most frequent were impaired AV valve regurgitation and SV dysfunction (11.1%) and arrhythmia

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

(8.2%). The most serious result from plastic bronchitis. Character of SV did not accomplished statistical significance. Complication-free survival was 88.9%, 74.9% and 56.3% (independent from character of SV), freedom from exitus was 97.2%, 94.71% and 84.2% respectively at 1, 5 and 10 years. From risk factor end-diastolic SV pressure affects the onset of late complications.
Conclusions: The most frequent late complications after fenestration closure result from impaired SV function and arrhythmias (20%). Elevated end-diastolic SV pressure is associated with higher incidence of complications during follow up.

PW1-4
Successful off-label implantation of the 'aortic' Edwards SAPIEN XT valve (29 mm) in right heart valvular lesions
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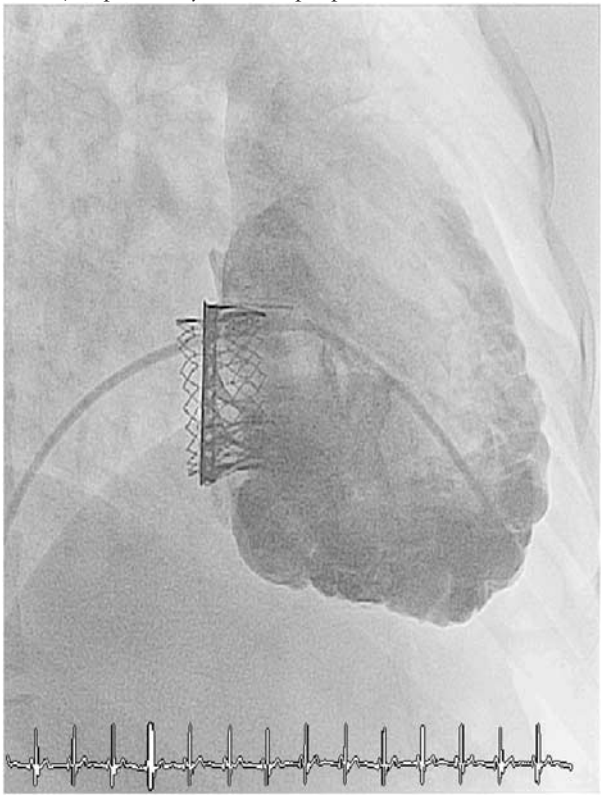
Introduction: Percutaneous implantation of the 'Melody' valve (Medtronic Inc., Minneapolis, MN, USA) and the Edwards valve (Edwards Lifesciences, Irvine, CA, USA) in pulmonary position has become a routine procedure in congenital heart diseases (CHD). The utilization of these valves is restricted to a maximum native valve diameter of 22 mm in the Melody valve and to 23 and 26 mm in the Edwards valves. Therefore their application in wider right ventricular outflow tracts as well as in alternative positions, such as the tricuspid ring, is substantially limited. To overcome this limitation, we describe the successful 'off-label' implantation of the new 29 mm 'aortic' Edwards SAPIEN XT transcatheter heart valve (Edwards Lifesciences) in pulmonary and tricuspid position.



Methods: Three patients (age 24–34 years) underwent a percutaneous valve procedure: 2 patients with valvular stenosis and insufficiency following tricuspid valve replacement by stented bioprosthesis (C-E 33 mm) received the Edwards SAPIEN XT valve (29 mm) in tricuspid position as a valve-in-valve procedure. In the third patient, who had pulmonary valve stenosis and insufficiency (s/p RVOT patch augmentation), the Edwards SAPIEN XT valve (29 mm) was implanted in pulmonary position after prestenting. All the implantations were performed using the Edwards Novaflex+ transfemoral system (20 F) with 'reverse' mounting of the valve.
Results: The transfemoral Edwards SAPIEN XT valve (29 mm) implantation was successful in all cases without any peri- or postprocedural complications. The implanted valves showed excellent functional and clinical results in all 3 cases, resulting in clinical and hemodynamic improvement in all patients during acute and short-term follow-up of a median of 1.2 months (0.7–4.2 months).
Conclusions: Off-label use of the 'aortic' transfemoral Edwards SAPIEN XT valve (29 mm) extends the application range of the percutaneous valve implantation technique in CHD patients beyond currently approved indications and is technically feasible and safe. It offers a good alternative to surgical valve repair in CHD patients with larger sized target regions up to a maximum diameter of 29 mm in either pulmonary or tricuspid position. These findings need to be confirmed by further cases and longer follow-up data.

PW1-5
Stent implantation as treatment for coarctation of the aorta with near total aortic occlusion
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Objective: To determine the safety and efficacy of endovascular treatment of coarctation in the setting of near total vessel occlusion.
Methods: From the total institutional database of 125 patients who had stent implantation for coarctation of the aorta, we identified 27 patients (15 male) over a 10 year period where the occlusion was near total as defined by a minimum aortic diameter of less than 4 mm on angiography. Median age was 18.4 years (range 12.4–61), median weight 65 kg (range 31.1–91 kg). All but 2 had native coarctation. Procedures were performed under general anaesthesia with intended final diameter equivalent to the transverse arch or isthmus, whichever was greater. Stents employed were Cheatham Platinum (CP) in 19 of which 5 were covered, self expanding in 3, Intrastent in 1, Palmaz in 1 and LD max in 2. Hand inflation only was employed for balloon mounted stents.
Results: Mean systolic pressure gradient fell from 34 ± 8 (1SD) to 5.5 ± 5 mmHg acutely and mean minimum diameter increased from 2.6 ± 1.2 to 11.9 ± 3 mm. The only procedure related complications were stent migration in two patients and a small dissection into the left subclavian in another which did not extend. One aneurysm was detected on post procedure CT scanning at 6 weeks which was successfully treated with a covered CP stent after a further 2.5 years. 5 patients have undergone successful stent re-dilations and one patient had a further stent implanted for stent fracture. At latest follow up median interval 2.6 range 1.1–9.6 years, none are hypertensive although 12 remain on treatment (monotherapy in 5).
Conclusions: In this age group, expansion to a 'normal diameter' of even near total vessel occlusion is readily achieved with a low complication rate. Bare metal stents appear to be safe, however in the absence of robust evidence and considering the consequences of vessel rupture, we would advocate the use of covered stents for near total occlusion.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

PW1-6

Early single stage hybrid approach for hypoplastic left heart syndrome: our experience

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Methods: From October 2011 to December 2012 at CCPM, 18 consecutive patients underwent hybrid approach procedure. After median sternotomy, the left and right pulmonary were banded by goretex, then stent was delivered in the arterial duct via a catheter positioned in the main pulmonary artery.
Results: All patients (median weight 3.04 Kg-range 2.2–4.2) received pre-operative continuous infusion of PGE2 and underwent procedure during the first 72 hours after birth. For arterial ductus stenting pre-mounted stents of diameter from 7 to 10 and lenght from 12 to 19 mm were used. Pulmonary arteries banding was performed by 3 mm custom goretex tube in 9 cases, and a 3.5 mm in the remaining cases. No patients required delayed sternal closure or ECMO. Post operative ICU management was characterized by continuous infusion of systemic vaso-dilatators, switched to oral therapy in all the cases. Enteral feeding was started in the first 48 hours; no cases of Necrotizing Enterocolitis (NEC) was reported. No major cerebro-vascular adverse events occurred during hospital stay. Mean hospital stay lenght was 20 days (range from 6 to 70 days), mean ICU stay was 11,8 days (range from 3 to 70 days). All patients survived to the procedure and were discharged successfully from the hospital. At the median follow up of 215 days no mortality was detected in patient while on Hybrid procedure. At a median age of 6,8 month, 9 patients underwent OMNIA procedure and one is listed for elective surgery; 8 patients are still on follow up after Hybrid procedure. 2 patient died after OMNIA procedure due to myocardial dysfunction and sepsis. Interstage follow up was conducted by twice visits per month in a dedicated Hybrid Clinic focused on medical therapy, echocardiography evaluation and growth pattern.
Conclusion: In our experience "Early" single stage hybrid approach for HLHS or complex, demonstrated to be safe, efficacy and free from major in-hospital or mid-term adverse events. According to avalaible literature freedom from peri-operative mortality, NEC, delayed sternal closure and high dose inotropic support make it a valid or superior approach when compaired with Norwood Stage I.

PW1-7

Results from an open-label, long-term safety and tolerability extension study using the pediatric FormUlation of bosenTan in pUlmonary arterial hypeRtEnsion (FUTURE-2)

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Objectives: This Phase 3, multicenter, open-label, non-comparative study (FUTURE-2) assessed the long-term safety and tolerability of bosentan pediatric formulation in children with idiopathic or familial pulmonary arterial hypertension (PAH). Exploratory evaluation of time to PAH worsening was also performed.
Methods: Children (≥2 and ≤12 years) with PAH who completed 12 weeks treatment in FUTURE-1, and for whom bosentan was considered beneficial, were eligible. Patients received the maintenance dose of 4 mg/kg bosentan bid which, if not tolerated, could be down titrated to 2 mg/kg bid. Safety and tolerability were assessed by adverse events (AEs), serious (S) AEs, deaths and laboratory abnormalities. Time to PAH worsening was defined as the first occurrence of death, lung transplantation or hospitalization for PAH worsening and presented using Kaplan–Meier estimates. Analyses were conducted in all patients who received ≥1 dose of bosentan from the start of FUTURE-1. Patients were grouped as 'all-patients' and by subgroups, depending on whether they had received bosentan prior to entry into FUTURE-1; 'previous bosentan treatment' or 'previously bosentan-naïve'.
Results: Out of 36 patients in FUTURE-1, 2 did not complete, 1 elected not to enrol in FUTURE-2 and therefore 33 patients continued into FUTURE-2. The overall median (range) time of monitored bosentan treatment was 119.9 (8.4–258.0) weeks, including exposure in FUTURE-1. 32 (88.9%) patients experienced ≥1 treatment-emergent AE and 15 (41.7%) patients experienced ≥1 treatment-emergent AE considered bosentan-related. The table shows treatment-emergent AEs occurring in >10% of all patients. 6 (16.7%) patients experienced ≥1 treatment emergent AE leading to premature treatment discontinuation. 18 (50%) patients experienced ≥1 treatment-emergent SAEs. Bosentan-related SAEs occurred in 3 patients. 6 deaths occurred during the study, 4 as a result of cardiac disorders and 2 due to infection. Autoimmune hepatitis was diagnosed in 1 patient who experienced treatment-emergent liver enzymes >3 x the upper limit of normal; this resolved following bosentan discontinuation. At 5 years, the Kaplan–Meier estimate of not having experienced worsening of PAH was 63.1% (95% CI 35.3–81.6%).
Conclusion: Bosentan pediatric formulation was well tolerated. There were no unexpected safety concerns associated with a longer bosentan duration in pediatric versus adult patients.

Treatment-emergent AE up to 1 day after permanent discontinuation of bosentan, n (%)	All patients (n = 36)	Previous bosentan treated (n = 15)	Previously bosentan-naïve (n = 21)
Total patients with ≥1 treatment-emergent AE	32 (88.9)	13 (86.7)	19 (90.5)
Abdominal pain	7 (19.4)	2 (13.3)	5 (23.8)
Nasopharyngitis	7 (19.4)	3 (20.0)	4 (19.0)
Pulmonary arterial hypertension (worsening)	6 (16.7)	4 (26.7)	2 (9.5)
Pulmonary hypertension (worsening)	6 (16.7)	1 (6.7)	5 (23.8)
Bronchitis	5 (13.9)	2 (13.3)	3 (14.3)
Upper respiratory tract infection	5 (13.9)	1 (6.7)	4 (19.0)
Chest pain	4 (11.1)	1 (6.7)	3 (14.3)
Fatigue	4 (11.1)	1 (6.7)	3 (14.3)
Flushing	4 (11.1)	1 (6.7)	3 (14.3)
Headache	4 (11.1)	1 (6.7)	3 (14.3)
Pneumonia	4 (11.1)	2 (13.3)	2 (9.5)
Syncope	4 (11.1)	3 (20.0)	1 (4.8)
Vomiting	4 (11.1)	–	4 (19.0)

PW1-8

Interventional closure of ventricular septal defects with the new Nit-Occlud Lê VSD device - 24 month of experience

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

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Introduction: Interventional closure of ventricular septal defects (VSD) has been aborted in many institutes because of bad risk of AV-blocs by using occluder devices. We report on our results during the past 24 month with the new Nit-Occlud Lê device. This retrospective study describes the effectiveness and complications implanting this occluder.
Methods: We report on 19 children with a body weight from 7.4 to 48.7 kg (median 18 kg) with perimembranous VSD (11 pts.) or muscular VSD (8 pts.). After adequate echocardiographic and angiographic documentation with measurements interventional VSD closure was performed with Nit-Occlud Lê devices in all cases.
Results: All devices were successfully implanted under sedation, without general anesthesia and without periprocedural complications, e.g. embolisation. A sufficient and immediate occlusion was documented by angiography in 12 cases, residual shunt in 7 cases, but disappeared 48h later. In two cases, hemolysis occured transiently for 48 hrs. In one case, a right bundle branch block appeared after releasing the occluder. This child developped a complete AV-block after one week, but was successfully treated with prednisolone and got back into sinus rhythm.
Discussion: Comparing the other currently available VSD occlusion systems the Nit-Occlud Lê device is built up of a flexible nitinol coil layer that adapts perfectly to the anatomy of the defect. Therefore, no permanent AV blocks were documented. Due to the polyester fibres in the distal part of the coil a rapid occlusion was expected, but transient hemolysis occured in two cases.

PW1-9

Technical aspects of hybrid procedure in hypoplastic left heart syndrome – the "true hybrid"

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Objectives: Hypoplastic left heart syndrome (HLHS) may be treated by hybrid procedure combining surgical bilateral pulmonary artery banding (PAB) with interventional stenting of patent arterial duct (PDA) and balloon dilatation resp. stenting of the atrial septum defect (ASD). The aim of the study was to analyze our results of hybrid procedure performed as a one step single procedure using median sternotomy for PAB and right atrial access for PDA and ASD interventions.
Methods: We analyzed hybrid procedures for HLHS and its variants regarding feasibility, safety and immediate periprocedural outcome.
Results: Between March 2006 and February 2012 hybrid procedure was performed in 31 infants (19 male) at median (range) age of 4 days (0–17) and body weight of 3 kg (1.9–3.8) within classical HLHS (n = 22) and its functional variants (n = 9). After median sternotomy and surgical bilateral PAB (3.5 mm, if body weight > 3.0 kg, otherwise 3.0 mm) we performed interventional PDA stenting resp. ASD enlargement using a right atrial (n = 15) or pulmonary artery access (n = 1) with surgical insertion of 5F sheath or in a separate second procedure using a femoral access (n = 15). The hybrid procedure could be performed safely in all patients without mortality. Due to maximal size of PDA with 7.9 mm (4.9–9.5) and minimal part with 6.9 mm (1.9–12.0) we used median 2 (1–4) self expanding stents (sinus Repo, OptiMed, Germany) with diameters ranging between 7 and 10 mm. For ASD interventions, we either used Tyshak Balloon (NuMed, USA) with diameters between 10–12 mm (n = 20), or transseptal perforation via radio frequency energy (n = 1). Additional interventions included balloon dilatation of coarctation of aortic arch (CoA) (n = 2) via a femoral arterial access. Periprocedural complications included rhythm disorders (n = 3), and dislocation of sheath (n = 1).
Conclusions: The one step single procedure ("true hybrid") with median sternotomy and right atrial access can be performed safely and offers the opportunity to treat ASD and PDA simultaneously in one session to spare infants from further procedures requiring longer hospital stay. Limitations of the "true hybrid" are restrictive ASD possibly requiring more stable catheter position, and stenotic PDA or additional interventions for CoA with a preferred retrograde approach.

PW1-10

Novel Method of Surgical Preparation for Transcatheter Completion of Fontan circulation: creation of an extracardiac pathway

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Objectives: We report creation of a novel surgical preparation for transcatheter completion of extracardiac Fontan circulation.
Methods: Nine lambs underwent surgical preparation; inferior vena cava (IVC) was cut with right atrium (RA) rim and anastomosed end-to-end with the inferior end of a Gore-tex conduit. A nitinol ring was placed around the IVC near the anastomosis. The SVC was cut and connected end-to-end with the RA auricle. Upper segment of Gore-tex tube was opened and connected to RA to allow free flow of IVC blood. The superior end was closed using PTFE membrane and a stented segment harvested from Contegra conduit was interposed between the superior end and the SVC for smooth transition. Fontan completion (opening of the SVC connection and closure of the fenestration) was attempted at 1–3 months following surgery. Animals were sacrificed just after (n = 7) and 3 months after completion (n = 2).
Results: All lambs were successfully preconditioned with one postoperative death due to SVC-RA occlusion, one elective sacrifice after 2-months to assess pathway integrity and seven successful completions. Autopsy findings showed widely patent IVC and SVC and covered stents well deployed to completely occlude fenestration.
Conclusions: A new method of surgical preparation and its subsequent transcatheter completion of extracardiac Fontan is created. This surgical preparation opens new frontiers for transcatheter and hybrid techniques for extracardiac Fontan completion.

PW1-11

Transcatheter Occlusion of Patent Ductus Arteriosus (PDA) in Low-Weight Pre-term Neonates (<2 kg) with Amplatzer Occluder II Additional Size (ADO-II-AS)

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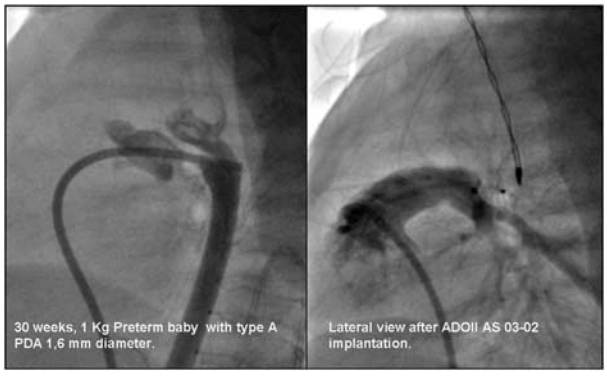
https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Background: Transcatheter treatment of PDA in very small infants is technically challenging and therefore often not considered as an alternative to surgery when medical treatment fails. However thoracotomy may cause pulmonary contusion and long-term sequelae as scoliosis
Objective: To describe our institutional experience with transcatheter PDA closure with ADO-II-AS in symptomatic low birth weight pre-term infants.
Methods: Retrospective review of all low birth weight pre-term infants who underwent device closure of PDA from January 2011-December 2012 with ADO-II-AS. Cases were selected based on the hemodynamic and respiratory repercussion of PDA, medical treatment failure (>2 cycles of intravenous Ibuprofen), and weight > 1000g. The procedure was done under anesthesia and tracheal intubation. Femoral vein access in all cases (4-F sheath in 8 patients, 5-F in 1 patient), arterial access was obtained in 5 patients inserting a microcatheter (2.7-F) into the femoral artery without sheath for aortic angiography. Immediate results were assessed by echocardiography before the device release.



Results: Nine infants. Median follow up was 3 months (range 1–17 months) Gestational age ranged from 24–32 weeks (27 ± 2.34). The median weight at the time of procedure was 1536 g (range 1000–1900g). Six patients were receiving mechanical ventilation before intervention. Type-A duct morphology was presented in seven patients and Type-C in two. The narrowest PDA angiographic diameter range was 1.23–2.94 mm (correlation coefficient with previous echo measurements of 0.95), descendant aorta size range was 2.65–3.60 mm., and PDA length range was 3.5–10.6 mm. The occluded device waist was 4 mm in all cases. Complete occlusion of the duct was instantly achieved in 7 patients, 2 patients had a small residual flow for 24 hours. Fluoroscopy time range; 5.6–27 min. One major procedure complication arose, device embolization in the left pulmonary artery, successfully removed. Two patients had moderate left pulmonary stenosis post-implantation, resolved during follow-up. Of the six patients who required mechanical ventilation, three were extubated in less than 10 days post-procedure.
Conclusions: The transcatheter closure of PDA with ADO-II-AS in carefully selected preterm infants is a safe and reliable alternative to surgical ligation.

PW1-12
Percutaneous Pulmonary Valve Implantation PPVI in post surgical conduit-free RVOT
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Introduction: to evaluate the safety and feasibility of PPVI in a non-stenotic conduit-free RVOT.
Methods: patients with important pulmonary regurgitation (PR > $^{3}/_{4}$) requiring revalvulation were screened with echo and/or magnetic resonance (landing zone <22 mm). Balloon-interrogation at low-pressure was performed using a flexible, non-compliant, mildly oversized balloon to delineate the potential zone of retention (careful observation for indentation of the balloon during submaximal inflation and deflation). Simultaneous coronary angiography was performed to exclude coronary compression. Pre-stenting was performed with a bare open cell stent (Andrastent XXL, Andramed, GE) to provide sufficient anchoring at the retention zone. Valve implantation was postponed for about 2 months to allow endothelial tissue ingrowth to fix the stent. Care was taken at the redo catheterization to cross the stent through the central lumen opening, by using a balloon-catheter.
Results: An acceptable landing zone was defined with balloon interrogation in 19 patients: after repair Fallot in 18 (infundibular resection or patch 4, transannular patch 14), and 1pt after neonatal balloon dilation of PS. Age 11.8 ± 4.2 years (6.5–20.8); weight 42.9 ± 19.1 kg (21–88). No coronary artery was at risk for compression. An Andrastent XXL (typically 39–43 mm, mean 41, range 30–57 mm) was implanted using a BIB (typically 24 mm, mean 23.1 ± 1.7 mm (18–24 mm). The only 30 mm short stent dislocated and was removed surgically. In 14 patients, PPVI was performed successfully 2 months later (Melody 13 (Medtronic,USA), Sapien 1 (Edwards, USA)); 4 pts are awaiting revalvulation. Typically the bare stent was expanded up to 24 mm; in these patients the Melody valve was implanted with a 22 mm Ensemble system. Echo evaluation day after PPVI showed a residual gradient of <10 mmHg in all, with none or trivial PR; RV dilation regressed significantly. Complications: 1 bare stent embolised to RV (cf above); 1 stent required additional flaring at PA bifurcation before PPVI; 2 Andrastents crumbled at re-entry during PPVI and required additional prestenting.
Conclusions: Percutaneous pulmonary valve implantation is safe and feasible in selected patients. Creating an adequate "landing zone" by pre-stenting makes the procedure safe and predictable. It needs to be determined whether this "window of opportunity" for "early" PPVI is beneficial in the long term management of these patients.

PW1-13
Transhepatic cardiac catheterisation in children: What have we learnt?
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Aim: To describe our experience in cardiac catheterisation in children via transhepatic approach where conventional venous access is impossible.
Patients and methods: Percutaneous transhepatic cardiac catheterization was performed on twelve occasions (2006–2012) in ten children aged 1 to 64 months (mean 12 months). Patient weight ranged from 3 to 16 kilogrammes. All children had documented bilateral femoral venous occlusion. Patient diagnosis were: univentricular heart (n = 5), pulmonary atresia with ventricular septal defect (n = 1), pulmonary vein stenosis (n = 2), atrial septal defect (n = 1) and cardiac transplant (n = 1). One patient was assisted with extracorporeal membrane oxygenation.
A 21 G and 4 cm long needle was used, introduced under fluoroscopy guidance. A 4F to 6F sheath was inserted into the hepatic vein using the Seldinger technique. To minimize risk of

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

bleeding the transhepatic tract was occluded with a vascular plug in three patients, with coils in one and with both a vascular plug and coils in two.
Results: Percutaneous transhepatic catheterisation was succesfully performed in all twelve attempts. The catetherization was diagnostic in seven children and interventional in five. A central venous catheter was placed in six patients: a *port a cath* was placed in one patient, a dialysis catheter (*Shaldon* catheter) in one patient, and an *Arrow* catheter was placed in the remaining (n = 4). Interventional procedures included stent implantation in pulmonary veins (n = 2), balloon angioplasty of pulmonary venous stenosis (n = 1), occlusion of atrial septal deffect (n = 1), and balloon angioplasty of right pulmonary artery and stent implantation in left pulmonary artery (n = 1). Ultrasound was performed 24 hours after transhepatic catheterization in seven patients. Peritoneal bleeding was encountered in one patient, resolved with conservative treatment; the coil implanted in the liver tract had embolized to the abdominal cavity.
Conclusions: Percutaneous transhepatic technique can provide a safe approach for cardiac catheterisation in children. In our experience, occluding the liver tract could minimize the risk of bleeding. This approach also allows for catheter implantation facilitating management of complex patients.

PW1-14

Lineage tracing of cells from the sino-atrial node area to determine the fate of the chicken cardiac conduction system

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Introduction: Knowledge about the developmental origin of the cardiac conduction system (CCS) is important to understand the pathophysiology of arrhythmias. During early development, the heart consists of two bilateral plates of splanchnic mesoderm, which fuse to form the primary heart tube (PHT) derived from the first heart field. A large number of cells at the venous pole of the PHT constitute a different cardiac progenitor population, called the second heart field (SHF). The contribution of this SHF population to the developing CCS remains the subject of discussion. We explored the feasibility of in ovo physical lineage tracing in chicken embryos, to perform long-term follow-up of SHF-derived cells contributing to the CCS.
Methods and results: Labeling was achieved by injecting a solution of two fluorescent dyes (DiI/5-TAMRA, Invitrogen) at HH stages 14–17, using a programmable microinjector (IM-300 Narishige, Japan). Right-sided labeling of the splanchnic mesoderm and the right cardinal vein was performed. Early embryos were sacrificed and analyzed 1–3 hours after injection (HH stages 14–17). Fluorescent labeling was found in the mesenchyme surrounding the sinus venosus and the right cardinal vein. In older stages of development (up to HH26), labeling of the developing sinus node and venous valves was found.
Conclusions: Our results show that it is feasible to label the SHF and elements of the CCS in ovo, which makes it possible to trace the fate of these cells over a longer period of time. Follow-up after HH stage 26 is needed to determine whether the contributions of the SHF are not only found in the sinus node but also in the atrioventricular node, common bundle and bundle branches Future plans include earlier labeling experiments as well as contribution of the left-sided SHF and determination of the differentiation capacities of these SHF derived cells.

PW1-15

Tenascin-C as a novel predictor of unresponsiveness to high-dose intravenous immunoglobulin and coronary artery lesions in patients with Kawasaki disease

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Introduction: Tenascin-C (TN-C) is an extracellular matrix protein specifically upregulated in response to tissue injury and inflammation. Recent studies have shown that serum TN-C could be useful biomarker for disease activity such as myocardial infarction or aortic aneurysm.
Around 10% of Kawasaki disease (KD) patients are unresponsive to high-dose intravenous immunoglobulin (IVIG) therapy, and these patients have increased risks of developing coronary artery lesions (CALs). We report the usefulness of TN-C as a novel biomarker to predict IVIG unresponsiveness and CAL formation in patients with KD.
Methods: Subjects consist of 108 patients with KD (6 patient with CAL) and 36 febrile child controls (FC group). We measured serum TN-C levels using ELISA and correlated with other laboratory data in KD patients. We compared the TN-C levels between KD and control as well as between the IVIG responder group (R group; n = 89) and the non-responder group (NR group; n = 19). In 51 KD patients, the chronological changes of the TN-C levels were evaluated; before IVIG treatment (pre TN-C), after IVIG at 9–15 days of the illness (post TN-C), and convalescent stages (late TN-C).
Results: In KD, pre TN-C levels positively correlated with serum AST ($r = 0.31$, $p = 0.01$), ALT ($r = 0.35$, $p = 0.004$), and CRP levels ($r = 0.25$, $p = 0.02$). Pre TN-C levels demonstrated no statistical significance between patients with KD and FC group (mean 86.2 vs. 76.9 ng/mL, $p = 0.57$, respectively). However, pre TN-C were significantly higher in NR group than those in R group or FC group (mean 106.1 vs. 69.6 and 76.9 ng/mL, $p = 0.029$). In addition, TN-C levels significantly decreased after IVIG administration (pre TN-C mean 69.5 vs. post TN-C 56.1 vs. late TN-C 38.8 ng/mL, $p < 0.01$). When pre TN-C was used to predict IVIG unresponsiveness, sensitivity, specificity and AUC were 63%, 76%, and 0.66 respectively. The increased TN-C levels were observed between 9–15 days of illness among CAL positive patients.
Conclusions: The TN-C level reflects not only inflammation but also tissue remodeling and may be a predictor of IVIG unresponsiveness and CAL formation.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

PW2-1

Plastic Bronchitis - Symptomatic improvement after pulmonary arterial stenting in four patients with Fontan circulation

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Introduction: Plastic bronchitis (PB) is a severe complication after a Fontan procedure with an estimated incidence around 1–2%. If not treated adequately the prognosis of PB in cardiac patients is very poor. We present four patients with PB after a total cavopulmonary connection with a stenosis of the left pulmonary artery that was stented successfully. In three patients symptoms of PB improved after the catheter intervention.
Methods: Four patients (cardiac diagnoses: hypoplastic left heart syndrome 2, tricuspid atresia 1, complex cyanotic heart disease with a functionally univentricular heart 1) with PB at a median age of 31 month (range 21.5–45 month) with a median weight of 12.7 Kg (range 9.5–14.8 Kg) presented at our clinic.
A complete separation of the pulmonary and systemic circulation by an extracardiac non fenestrated total cavopulmonary connection had been performed at a median of 11 months (18–21 month) preceding the hospital admission. All patients underwent a complete cardiological examination including cardiac catheterisation.
Results: All four patients had significant LPA stenosis, which was treated by stent implantation. The LPA-diameter was augmented from a median value of 4.8 mm (range 3.9–6 mm) to a median value of 8.7 mm (range 7.8–10.4 mm). The median transpulmonary gradient was 8 mmHg (range 6–9 mmHg) and the median central venous pressure was 15 mmHg (range 12–17 mmHg). Until today three children are free of symptoms. Solely one child, the one with the highest transpulmonary pressure, redeveloped casts. Median follow-up time is 13.5 months (range 8–25 month).
Conclusions: This series of cases shows significant improvement of PB after successful LPA-stenting in at least 3 patients. Our cases show that haemodynamic improvement is crucial in the management of PB. Any effort should be undertaken to relief any pulmonary artery obstruction in these patients before fenestration of the Fontan tunnel or cardiac transplantation are performed alternatively.

PW2-2

Values of immunoglobulin G are correlated with the responsiveness to initial

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Introduction: Low levels of serum immunoglobulin G (IgG) before intravenous immunoglobulin (IVIG) treatment in Kawasaki disease (KD) were reported as one of the risk factors for coronary artery abnormalities (CAAs). This risk factor has to be reevaluated, because the dosage of IVIG has been changed from 0.2–0.4 g/kg for 5 days to 2 g/kg/1–2 days and the incidence of CAAs has been decreased.
Patients and Methods: We reviewed the clinical records of the KD patients who were admitted to Kagoshima Medical Association Hospital in Japan between January 2001 and August 2011. The patients who were given IVIG (2 g/kg/1–2 days) within seven days of illness and were evaluated for their immunoglobulin values (IgG, IgA, and IgM) before treatment were assigned. The values of immunoglobulin and diameters of coronary arteries were evaluated using Z-scores for age or body surface area, respectively. A patient with CAA was defined as a patient with coronary Z-score beyond 3.0 at 1 month of illness.
Results: The subjects were 197 KD patients and contained 22 non-responders and 16 patients with CAAs. Of these, 150 patients (76%) showed IgG values below zero (median; −0.72, 25 percentile/75 percentile; −1.4/−0.0). Non-responders showed higher IgGz values than responders; −0.26 (−0.83/0.34) vs. −0.79 (−1.40/−0.03), $P = 0.020$. IgGz values were positively correlated with the duration of fever after initiation of IVIG treatment ($r = 0.095$, $P = 0.025$). Rogistic regression analysis revealed IgGz values were the significant independent factors for non-responders (Odds ratio; 1.36, 95%CI; 1.002/1.849, $P = 0.048$). ROC analysis showed that −1.0 of the IgGz value was the useful cut-off value. Its sensitivity was 77% and the specificity was 61%. Between the patients with CAAs and those without CAAs, IgGz values were not statistically different.
Conclusions: Low IgGz values were not a risk for CAAs in the present study. KD patients with higher IgGz values may have a risk of non-responsiveness for initial IVIG.

PW2-3

Brain volumetry in infants with congenital heart disease: pre- and postoperative assessments using cerebral MRI compared to healthy controls

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Objectives: MRI studies in neonates with congenital heart disease (CHD) have demonstrated delayed brain maturation and mostly focal brain injury. To better define the distribution of cerebral injuries and regional brain growth in neonates with CHD, we compared volumetric measures from pre- and postoperative MRI of patients to healthy neonates.
Methods: Cerebral MRIs of 32 term-born CHD patients, scanned before and after heart surgery (mean age: 6.8 days and 26.8 days, respectively), were manually segmented to measure volumes of total, grey and white matter and of selected brain regions. Results were compared with MRIs of 17 healthy term born neonates (mean age: 23.5 days).
Results: Between pre- and postoperative MRI, patients showed significant brain growth, especially in the cortical grey matter (0.25%/day), cerebellum (0.20%/day), and deep gray matter structures (0.10–0.15%/day, all $p < 0.004$). Volume increase of the white matter was 0.05–0.06%/day (left/right; $p = 0.017$/0.003); increase of total brain volume was 0.14%/day ($p < 0.001$). Compared to healthy controls, the size of all brain structures (except ventricles and right amygdala) was significantly reduced postoperatively. Largest differences were found in deep gray matter structures (13.8–16.8%, $p = 0.05 - < 0.001$), cortical grey (12.1%, $p = 0.01$) and white matter (11.8%, $p < 0.001$). Total brain volumes were reduced by 11.3% ($p < 0.001$).

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Conclusions: In neonates with CHD, significant differences of white and deep grey matter volumes were found postoperatively. Brain growth was high, with notable regional differences. Our results contribute to the knowledge on the timing of cerebral injury in neonates wth CHD.

PW2-4

Relation of quality of life, exercise capacity, and ventricular function in adults with repaired tetralogy of Fallot

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Introduction: Tetralogy of Fallot is the most common cyanotic congenital heart disease all over the world. Although early postoperative survival was excellent, patients with repaired tetralogy of Fallot (rTOF) were at risk for long-term complications related to heart failure and arrhythmia. In National Taiwan University Hospital, the long-term survival rates for rTOF was 95.8%, 92.7%, and 90.5% at 10-year, 20-year, and 30-year after surgical correction. Aside from cardiac mortality as the major cause of late deaths (52%), we found that unnatural deaths, including suicide, accounted for 26% of late deaths, and was significantly higher than that of the general Taiwanese population. This finding highlights the potential threat of psychosocial problems in this patient population.
Methods: From the database of National Taiwan University Hospital, quality of life (QoL) was assessed in 144 adults with rTOF, and 138 of them (age: 31.4 ± 10.1 years, male: 46%) had valid data for further analysis. Compared to the general population, rTOF had generally lowered QoL scores in physical and psychological domains ($P < 0.001$ and $P = 0.024$, respectively). In contrast, QoL in environmental domain was better in rTOF ($P = 0.003$).
Results: Among these patients, 92 received cardiopulmonary exercise test (CPX) immediately after the assessment of QoL. Patients' self-estimated exercise capacity before CPX correlated well with overall QoL satisfaction ($r = 0.659$, $P < 0.001$) as well as physical QoL Z score ($r = 0.518$, $P < 0.001$). However, all these measures failed to predict actual exercise capacity as evaluated by peak oxygen consumption. Furthermore, we found that QoL had no relationship with either right or left ventricular function (end-diastolic volume, end-systolic volume, and ejection fraction) as assessed by cardiac magnetic resonance imaging in a subset of patients ($n = 78$).
Conclusions: Therefore, self-reported QoL and physical functioning poorly predicts actual exercise performance or ventricular function in patients with rTOF. Although QoL should be incorporated in the evaluation of patients' functional performance late after TOF repair, objective measurements of exercise and ventricular function remain essential and mandatory for decision making in the clinical follow-up of rTOF patients.

PW2-5

Pulmonary Arterial Hypertension in Children with Congenital Portosystemic Venous Shunt

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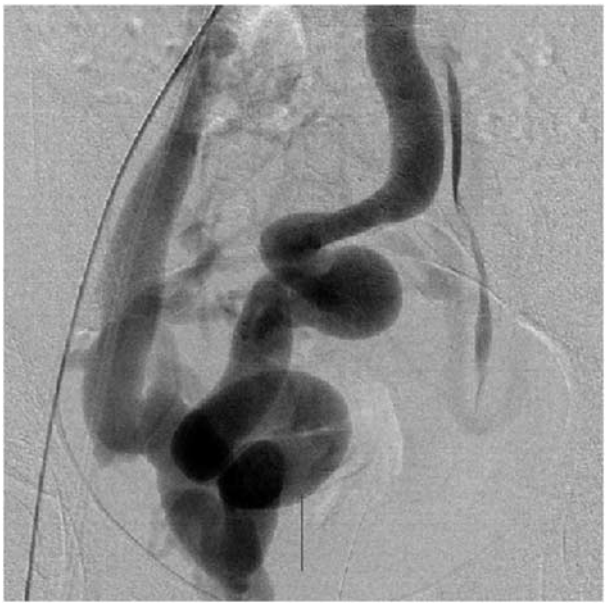
Objectives: Congenital portosystemic venous shunt (CPSVS) is a rare disease with a variety of connections. Some patients of CPSVS presented with pulmonary arterial hypertension (PAH) which is a life-threatening disorder. The clinical perspective of PAH secondary to CPSVS is unknown. The purpose of this research is to bring out the clinical, laboratory and physiological characteristics of this disorder.
Methods: Nineteen patients with CPSVS were studied about clinical, laboratory and our institution.
Results: The median age at the diagnosis of CPSVS is 3 years. Congenital hypoplasia or absence of portal vein was detected in 11 patients. Eight of the 19 patients were identified to have mild to severe PAH (mean pulmonary artery pressure: 50 ± 17 mmHg, pulmonary artery resistances: 15.1 ± 12.9 U). The serum total bile acid (TBA) and aspartate aminotransferase (AST) levels of the patients with PAH were significantly higher than those of the patients without PAH (TBA: with PAH 150 ± 62, without PAH 64 ± 45 μmol/L, $p = 0.0099$ AST: with PAH 52 ± 19, without PAH 35 ± 19, $p = 0.0468$). There was no difference in the blood ammonia levels. As treatment for PAH, bosentan or sildenafil: prostacyclin ($n = 2$) and prostacyclin ($n = 4$) were administered. Catheter embolization of the shunt was conducted in 2 patients with PAH. Three of 8 patients with PAH died because of right heart failure. In only one patient with PAH, pulmonary artery pressure was decreased to almost normal range by catheter embolization.
Conclusions: The CPSVS patients with PAH had high levels of TBA. This suggested that the amounts of shunt flow might be related to the onset of PAH. Appropriate catheter intervention may be an effective treatment for the CPSVS patients with PAH whose portal vein pressure was not elevated by balloon test occlusion of the shunt vessels.



PW2-6

Effectiveness of Ambrisentan Therapy in Patients of Protein-Losing Enteropathy after Fontan Operation

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Aim: The purpose of our study is to investigate the efficacy of ambrisentan in patients with steroid-dependent PLE after

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Fontan procedure who has been treated by steroids, immunosuppressant, and pulmonary vasodilators (PDE5-inhibitors, bosentan, and beraprost).
Method: The study subjects were 4 patients with PLE after Fontan (4 Males and age ranged 4–19y, the interval of PLE from Fontan 1.3–8 months). All were steroid-dependent with immunosuppressant and pulmonary vasodilators (bosentan, and sildenafil) and needed repetitive infusion of albumin and γ-globulin.
Treatment protocol: ambrisentan was given at the dose from 0.09 to 0.19 mg/kg per day, replacing bosentan without changing the dose of the rest medications. Then the efficacy of ambrisentan was assessed (1) by the supplement dosage of albumin and γ-globulin and (2) by the dosage of steroids.
Results: No severe adverse effect but transient puffy face (2 pts) was found. Ambrisentan could reduce the daily dose of steroid from 15 mg (median) to 9 mg, and could prolong the supplement cycles of albumin/γ-globulin from 2 days to 25 days as a median for 3.3 months, comparing to bosentan. The serum albumin also increased from 3.7 to 4.3g/dl.
Conclusion: Ambrisentan is a potent therapeutic option for steroid-dependent PLE after Fontan procedure.

PW2-7
Normal Blood Pressure and Maximum Rate Pressure Product Responses to Treadmill Exercise Test in Healthy British Children

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Background: Normal cardiovascular responses to exercise in childhood with regards to maximum blood pressure response, rate pressure product and recovery time are not well documented. In addition, maximum normal blood pressure response to exercise in childhood is extrapolated from adult studies which make assessment of hypertensive response in disease situation such as coarctation rather difficult. We therefore aimed to define normal cardiovascular responses to exercise in healthy British children.
Method: We retrospectively reviewed our experience in all children who underwent exercise testing (Bruce treadmill protocol) at University Hospital of Wales between 2003 and 2010. One hundred and thirty-seven children without any structural heart disease were included in the study.

Age No of subjects	9 to 10 years All (n = 22)	11 to 12 years All (n = 36)	13 to 14 years All (n = 46)	15 to 16 years All (n = 33)
Exercise duration (min)	13.1 ± 0.67	13.3 ± 0.50	13.1 ± 0.33	14.3 ± 0.49
Exercise capacity (METS)	14.7 ± 0.61	15.5 ± 0.45	15.4 ± 0.41	16.5 ± 0.51
VO_2 max ml/kg/min	51.6 ± 2.1	54.4 ± 1.6	53.7 ± 1.4	57.8 ± 1.8
Recovery time (min)	4.8 ± 0.30	6.3 ± 0.34	6.2 ± 0.24	5.7 ± 0.35
HR max (bpm)	194.3 ± 2.8	195.2 ± 2.0	196.4 ± 2.4	198.6 ± 2.2
% HR max reached (%)	92.3 ± 1.3	93.6 ± 0.99	95.1 ± 1.2	97.1 ± 1.1
SBP max (mmHg)	136.1 ± 3.5	139.8 ± 2.0	146.5 ± 2.4	151.5 ± 2.9
DBP max (mmHg)	70.5 ± 2.1	70.9 ± 1.3	72.8 ± 1.3	73.5 ± 1.6
Rate pressure product (HR max*SBP max)	26567.1 ± 910.1	27275.8 ± 483.4	28824.9 ± 634.2	30125.7 ± 725.9

Results: There were 80 males and 57 females and their age ranged from 9 to 16 years. All children achieved minimum exercise duration of 12 minutes regardless of age or gender. Lower VO2max values were observed in females compared to male subjects in younger age but not so much in adolescents. All subjects achieved over 90% of maximum predicted heart rate for any given age. Younger subjects showed quicker heart rate recovery compared to older individuals. Maximum blood pressure did not exceed above 153.6 ± 3.6 mmHg in any age group. Maximum rate pressure product was similar both gender before puberty but higher in males in 15–16 years group.
Conclusions: Exercise duration in healthy children is minimum 11.7 ± 0.39 minutes. 90% of maximum heart rate response is always achievable standard. Maximum blood pressure response is much lower than historically quoted which may necessitate redefining hypertensive response to exercise in childhood. Maximum rate pressure product becomes progressively higher with age but is not influenced by gender. Recovery time is quicker in children than adults.

PW2-8
Reference values of the right ventricular outflow tract systolic excursion (RVOT SE) in 711 healthy children and calculation of z-score values

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Objective: Determination of right ventricular (RV) function has gained more interest in the last years. The RV outflow tract systolic excursion (RVOT SE) has been recently introduced as an echocardiographic tool to assess RV systolic function in adults. We aimed to determine growth related changes of RVOT SE in children to establish references values.
Methods: A prospective study was conducted in a group of 711 healthy pediatric patients (age: day 1 to 18 years), (BSA: 0.14 to 2.26 m^2). We determined the effects of age and body surface area (BSA) on RVOT SE values. RVOT SE values were also correlated with established RV systolic function parameters tricuspid annular plane systolic excursion (TAPSE) and tricuspid annular peak systolic velocity (S').
Results: The RVOT SE ranged from a mean of 3.9 mm (±3 SD: 1.5–6.4 mm) in neonates to 9.5 mm (±3 SD: 5.7–13.3 mm) in 18 year old adolescents. The RVOT SE values showed a positive correlation with age ($r = 0.90$, $p < 0.001$) and BSA ($r = 0.91$, $p < 0.001$). No significant difference in RVOT SE values between females or males was found ($p = 0.707$). A positive correlation was seen between RVOT SE and TAPSE ($r = 0.83$, $p < 0.001$) and between RVOT SE and S' ($r = 0.86$, $p < 0.001$)
Conclusions: Z-scores of RVOT SE values were calculated and percentile charts were established in the pediatric age group. RVOT SE provides a simple measure and, in combination with RV long-axis excursion parameters TAPSE and S', provides comprehensive assessment of RV systolic function.

PW2-9
Functional analysis of the anatomical right ventricular components: should assessment of right ventricular function after repair of tetralogy of Fallot be refined?

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Objective: Follow-up after ToF correction is directed to detect timely RV dysfunction by following the pulmonary regurgitation and global RV function and size, with little attention for the effective contribution of regional RV dysfunction. This study aims to determine the contribution of regional RV dysfunction on functional capacity after repair of ToF
Methods: 42 ToF patients were investigated with CMRI for regional RV dysfunction in relation to global RV function by functional quantification of sinus and outflow part of the RV. Impact of regional and global RV dysfunction on clinical status was studied by exercise testing.
Results: Global RV function was lower than RVsinus function ((EF $52 \pm 12\%$ versus $57 \pm 10\%$, $p < 0.001$), attributable to the adverse influence of RVOT dysfunction (EF $34 \pm 17\%$). Percent predicted peak VO2 correlated better with RVsinus EF compared to global RVEF ($r = 0.51$, $p = 0.001$ versus $r = 0.44$, $p = 0.004$). Multivariate analysis revealed RVsinus EF ($\beta = 0.34$, 95% CI 0.07–0.61, $p = 0.013$) and extent of RVOT akinesia ($\beta = -0.28$, 95% CI -0.50; -0.06, $p = 0.015$) as significant determinants of exercise capacity. Impaired exercise performance, defined as % predicted peak VO2 < 85%, occurred in 43% of the patients and was independently determined by type of repair (transventricular versus transatrial : OR. 6.0, 95%CI 1.31–17.3, $p = 0.02$) due to greater sinus and RVOT dysfunction.
Conclusion: Functional analysis of the RV components shows that exercise capacity after ToF repair is better predicted by systolic function of the RVsinus as the extent of RVOT dysfunction commonly leads to underestimation of global RV function. Further validation in larger scale studies is needed to postulate whether this method of RV analysis is more appropriate than assessment of global RV volumes for timely detection of early RV dysfunction in order to initiate subsequent pulmonary valve implantation with eventual associated surgical RVOT remodeling.

PW2-10

Assessment of early onset chronic progressive anthracycline cardiotoxicity by tissue Doppler imaging in children

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Introduction: Most of the clinical studies concerning anthracycline cardiotoxicity have investigated "late onset cardiotoxicity" and focused on particularly the left ventricular systolic functions. We aimed in this study to assess "early onset chronic progressive anthracycline cardiotoxicity" in the left and right ventricular segments using tissue Doppler imaging (TDI) with increasing cumulative anthracycline doses.
Methods: The patient group included the patients who had been received doxorubicin and/or daunorubicin within a time period of between one week and one year when they were examined (mean 2.6 ± 2.9 months, range: 0.3–11.5 months). During TDI studies, apical 4-chamber views were obtained and diastolic and systolic parameters were measured at four different segments [lateral annulus of the mitral valve (MV-lat), middle part of left ventricular lateral wall (LVLW), lateral annulus of the tricuspid valve (TV-lat) and right ventricular lateral wall (RVLW)]. The echocardiographic data on all patients were classified into three groups according to their cumulative anthracycline doses: treatment group (TG)-I ($\leq 120\,mg/m^2$; $n = 26$), TG-II ($120–240\,mg/m^2$; $n = 39$), TG-III ($\geq 240\,mg/m^2$; $n = 40$). Standard echocardiographic and TDI parameters of the patients were compared with healthy controls.
Results: 72 patients (38 girls, 34 boys) and 31 controls (18 girls, 13 boys) were enrolled in this study. The mean age was 8.2 ± 4.5 years in patient group and 9.6 ± 4.2 years in the control group ($p > 0.05$). The comparison of control and TG-1 groups revealed significant decreases in E' velocities in TV-lat and RVLW segments ($p < 0.001$ and $p < 0.05$, respectively). While MPI was significantly increased in all four segments, a significant decrease in S' velocity was seen only in MV-lat and LVLW segments ($p < 0.05$ and $p = 0.001$, respectively). Conventional echocardiographic parameters including EF did not change significantly in TG-I. These changes in TDI were more prominent in TG-II and TG-II. Moreover, increase in IRT in all segments except MV-lat segment and decrease in EF were statistically significant with a cumulative anthracycline dose of $>120\,mg/m^2$.
Conclusions: Abnormalities in diastolic functions by TDI were observed in the RV earlier than LV with $\leq 120\,mg/m^2$ cumulative anthracycline doses, whereas systolic dysfunction findings were observed first in LV with increasing drug doses.

PW2-11

Progression of aortic valve dysfunction and aortic root dilatation in paediatric patients with isolated bicuspid aortic valve

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Background: Few data exist regarding the rate of progression of aortic valve dysfunction and aortic root dilatation in paediatric patients with isolated bicuspid aortic valve (BAV).
Methods: A total of 179 paediatric patients (median [95% IQ] age at enrolment 7.8 [2.7–12.0] years, 76.5% male) with isolated BAV were prospectively followed. Patients with severe valve stenosis or regurgitation at baseline were excluded. Progression of aortic stenosis (AS) or regurgitation (AR) from baseline to follow-up was defined as an increase by ?1 grade according to a three-level scale severity. Progression of aortic dimensions at different levels of the aortic root was estimated by calculating corresponding changes in the Z-scores. A Z-score >2 was considered significant.
Results: At baseline, AS was present in 25 (14.0%) patients (mild in 23, moderate in 2), whereas AR was present in 87 (48.6%) patients (mild in 79, moderate in 8). Mean diameters at the level of annulus, Valsalva sinuses (VS), sino-tubular junction (STJ), and proximal ascending aorta (AA) were 17.4 ± 4.5 mm, 22.2 ± 5.6 mm, 19.3 ± 5.1 mm, and 21.7 ± 6.2 mm, respectively. The corresponding proportions of subjects with significant enlargement (Z-score > 2) at baseline of 24.6%, 15.1%, 22.4% and 39.1%.
After a median follow-up of 5.4 [2.3–9.2] years, AS was present in 28 (15.6%) patients (mild in 21, moderate in 6, and severe in one), whereas AR was present in 102 (57.0%) patients (mild in 75, moderate in 27). A progression ?1 grade in AS and AR was observed in 9 (5.0%) and 29 (16.2%) patients, respectively.
Mean diameters at the level of annulus, VS, STJ, and AA were 20.4 ± 5.0 mm, 26.7 ± 6.2 mm, 21.9 ± 5.9 mm, and 26.4 ± 6.8 mm, respectively. The corresponding proportions of subjects with Z-score > 2 were 18.4%, 11.2%, 17.3%, and 49.2%. A progression from normal dimensions (Z-score ?;2) to significant enlargement (Z-score > 2) was observed in a minority of patients (10.6%, 5.6%, 9.5%, and 19.0% of patients, respectively).
Conclusions: The rate of progression of aortic valve dysfunction and aortic root dilatation in paediatric patients with isolated BAV is

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

relatively slow. These findings may be taken into account to better guide risk assessment and clinical follow-up in these patients.

PW2-12

Comparison of echocardiography and ECG for detection of abnormal phenotype in childhood mutation carriers for familial hypertrophic cardiomyopathy

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Background: Studies from tertiary centres specialised in hypertrophic cardiomyopathy (HCM) have claimed that frequency of abnormal ECG in affected adults range from 75–97% but data from geographically based cohorts and childhood patients are lacking. We have therefore studied a geographical cohort of childhood patients positive for a mutation causing familial HCM.
Methods: 36 mutation carriers, age 0.2–18 years (median age 10 yrs), identified from a systematic screening project in the West Götaland region in Sweden, were compared with age and gender-matched normal controls. The parameters compared were measures that have been suggested useful for screening for early detection of abnormal phenotype. Firstly, M-mode indices of relative wall thickness and contractility from long-axis M-mode: septum-to-cavity ratio (sepcavr), left ventricular wall-to-cavity ratio (lvcavr), posterior wall systolic wall-to-cavity ratio (syscavr) and fractional shortening (FS). Secondly, electrocardiographic measures such as QRS-amplitude sum in limb-leads (LLQRSS), twelve-lead QRS-amplitude sum (TwQRSS), twelve-lead amplitude-duration product (TwLProd). In addition the ECG-risk score was quantified. Data were compared with Mann-Witney U-test.
Results: Comparing mutation carriers with controls neither LLQRSS (median values 6.5 and 6.7mV respectively), TwQRSS (20.7 versus 19.3mV) or TwLProd (1.64 versus 1.68mV.s) were significantly different. The most sensitive measure cut-off was TwQRSS >2.4mV where 28% of mutations carriers were positive, but there were also 16% false positives among normals. Pathological Q-waves were present in 43% of mutation carriers but only in 3% of normals so useful. ECG-risk score was significantly higher in mutation carriers ($p = 0.0002$), and 19% of mutation carriers had a high-risk score of 6 or above, versus none among controls, where the highest value was 3. Sepcav, lvcavr, syscavr and FS are all significantly increased in mutation carriers ($p < 0.00001$ in all). A cut-off of ≥ 0.26 has a sensitivity of mutation carriage of 75%, as has a syscavr ≥ 0.55, lvcavr $\geq 0{,}22$ has a sensitivity of 47%, all with no false positives in normals. FS $\geq 42\%$ has a sensitivity of 64% and no false positives.
Conclusions: 57% of childhood mutation carriers for familial HCM have normal ECGs, whereas M-mode indices of relative septal thickness, and radial systolic contractility, are abnormal in 75% of mutation carriers already in childhood.

PW2-13

Anomalous left coronary artery from the pulmonary artery associated with other cardiac defects: a difficult joint diagnosis

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Background: Anomalous left coronary artery connected to the pulmonary artery (ALCAPA) is a coronary abnormality which can be associated to other congenital heart defects which complicates the positive diagnosis of the coronary abnormality, especially before surgery.
Objective: Here we report a series of 13 patients with ALCAPA with a focus on the type of associated heart defect, the moment of diagnosis of the coronary abnormality related to surgery and their outcome.
Methods: Retrospective assessment of medical files of all patients with ALCAPA and other congenital heart defects in two important French Departments of Pediatric Cardiac Surgery from 1987 to 2012.
Results: Thirteen patients with ALCAPA and other cardiac defects were identified. Five patients had had a prenatal diagnosis of congenital heart disease concerning the associated cardiac defect. The heart defect most frequently encountered in association to ALCAPA was aortic coarctation ($n = 4$) followed by tetralogy of Fallot with or without pulmonary atresia ($n = 3$). There was one case of hypoplastic left heart syndrome, one right aortic arch, one congenital mitral malformation and one infant with divided left atrium and anomalous venous return. Only three patients had a complete diagnosis of the cardiac defect and the left coronary abnormality before surgery. In four cases the coronary anomaly was discovered during surgery conducted for another cardiac defect and treated at the same time by coronary reimplantation. The six remaining patients were diagnosed after cardiac repair. Three of these patients only had a post-mortem diagnosis. Eight of 13 patients died after surgery. Half of them deceased within the first 30 days after repair. The remaining patients are in good health with a median follow-up of 5.3 years (range: 2.1–8.5 years).
Discussion: This series confirms that ALCAPA associated with other cardiac defects is often misdiagnosed before surgery. Pulmonary hypertension due to shunt or coarctation can maintain an anterograde flow in the anomalous coronary artery until cardiac repair. Myocardial ischemia will only become apparent once the defect has been repaired when pulmonary pressure lowers. In this series postoperative survival was compromised especially if the coronary anomaly had not been diagnosed preoperatively.

PW2-14

Outpatient Clinical Follow Up of Children With Implantable Continuous Flow Ventricular Assist Devices: The Preliminary Experience from Ege University Hospital

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Introduction and Objectives: Shortage of donor hearts leads to increased waiting times on the transplant list. There is an urgent need for VAD systems that allow long-term support with low morbidity and minimal restrictions of the daily activities of Patients. With the development of implantable continuous flow VADs of the third generation, survival and quality of life have improved. In this study, we report our experience with outpatient follow up three children with continuous flow VAD (HeartWare). Our first case is the youngest patient among outpatient followed patients in Europe.
Methods: In this retrospective study, we report our preliminary experience with continuous flow VAD. All of the patients were followed in outpatient clinics.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Results: Between August 2012 and January 2013 three patients (2 girls, 1 boy; aged 7 to 13 years; wieghted 18 to 44 kg) recieved continuous flow HeartWare VAD. All of the three patients had end stage heart failure due to dilated cardiomyopathy and under high dose positive inotropic support. All of the patients were extubated on postoperative first day. The duration of intensive care stay varied between 6 to 10 days. Patients were discharged from hospital 35 to 60 days after VAD implantation. All of the three patients are still under VAD support and waiting for Heart transplantation. On follow up our three patients, we make INR measurement twice in a week; physical examination including growth monitorization and echocardiographic and psychological evalution every month. Early mobilization and discharge from the hospital decreased the risk of hospital infection. They can easily perform daily social activities and attend school. Their school performance are very good. Living with their families at home certainly decreases patients's and their parents's anxiety; the affect of the patients totaly changed positively after discharge from the hospital. All of the patients gained weight.
Conclusion: Implantable continuous flow device support offers a safe and comfortable alternative to paracorporeal systems for larger children with end stage heart failure. HeartWare system provide early mobilization of these patients and continue to perform their normal daily social activities.

PW2-15
Effects of carvedilol therapy on cardiac autonomic control, QT dispersion and ventricular arrhythmias in children with dilated cardiomyopathy
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Introduction: The purpose of this study was to examine the effects of carvedilol therapy on autonomic control of the heart and QT-interval dispersion (QTd) as an arrhythmia marker among children with idiopathic dilated cardiomyopathy (DCM) whose symptoms were not adequately controlled with standard congestive heart failure (CHF) therapy.
Methods: A total 34 patients with idiopathic DCM were analyzed in the study.All patients had undergone carvedilol in addition to standard therapy for at least 6 months. Clinical, echocardiographic, electrocardiographic parameters, and 24-h Holter records of patients were evaluated before and after carvedilol treatment.
Results: After the six months of carvedilol therapy, the Ross clinical scales significantly decreased from, left ventricular ejection fraction (LVEF) and left ventricular fractional shortening (LVFS) significantly increased, left ventricle end diastolic dimensions (LVEDd) and left ventricle endsystolic dimensions significantly decreased. There were statistically significant increases in mean SDNN, SDANN, rMSSD, and pNN50 after carvedilol therapy. Baseline SDNN was significantly correlated with baseline HR and total premature ventricular contraction (PVCs). After carvedilol, SDNN was correlated with the clinical score of CHF, heart rate, LVEF, LVSF and total PVCs. In addition, rMSSD and pNN50 were correlated with heart rate, LVEF and LVSF after carvedilol therapy. A significant reduction was observed in the terms of QTc min, QTc max and QTd values after carvedilol treatment. QTd was slightly higher in patients with a lower clinical score than in those with a higher clinical score, however, the difference was not statistically significant after carvedilol treatment. QTd was significantly related to total PVCs. Although eight patients had PVCs before treatment, they disappeared in four patients and PVC decreased in two patient after treatment. Sustained ventricular tachycardia was not observed in any patients before and after treatment. Before carvedilol therapy, five patients had ventricular couplets and two patients had nonsustained ventricular tachycardia. A trend toward a decrease in ventricular couplets and nonsustained ventricular tachycardia did not reach statistical significance after six months.
Conclusions: We concluded that the addition of carvedilol to standard medical regimens can improve clinical symptoms, heart rate variability in association with improved left ventricular function and reduce in arrhythmia markers in children with DCM.

PW3-1
Effects of surgical truncus block rotation on the conduction system in children with transposition of the great arteries and left ventricular outflow obstruction
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Introduction: The standard surgical management of patients with transposition of the great arteries (TGA) and left ventricular outflow obstruction (LVOTO) has been the Rastelli operation. More recently, truncus block rotation (TBR), by cutting out aortic and pulmonary root in one block and by rotating it 180 degrees, has been introduced as a new option for anatomical repair.
Aim: To evaluate the effects of this surgical method on the conduction system.
Methods: 16 consecutive patients (Median age at surgery 244 days, range: 4–2360) with TGA and LVOT were treated in our institution by TBR. Preoperative, postoperative and follow-up ECGs were reviewed for QRS duration, QRS-pattern, repolarisation and QTc. The 1st ECG was obtained at median 3,5 (1–30) days before surgery, the first ECG after surgery at median 12 (3–27) days later. The median follow up time was 585,5 (11–2572) days.
Results: Except one complete AV-block, no major arrhythmias were observed during the study period. One patient had a transient atrial rhythm right after surgery, which changed to sinus rhythm during the follow up period. All patients without typical bundle branch block (BBB) pattern had a median QRS duration of 65 ms (54–112 ms) before surgery, 62 ms (54–122 ms) after surgery and 84 ms (66–128 ms) at the last follow up visit. None of the patients had a typical BBB pattern before surgery, but 8/16 pts (50%) had a right BBB after surgery, which persisted during follow up. This compares well to a comparable Rastelli cohort, where a right BBB prevalence of 77% was reported. In the BBB group the median QRS-duration was 100 ms (86–116 ms) right after surgery and 100 ms (92–128 ms) at last follow up. In 12 of 16 patients there were unspecific ST changes and negative T-waves, which persisted in the follow-up with unknown significance for the future.
Conclusion: Our data suggests, that negative effects on the conduction system and arrhythmias do not play a major role in TBR. Their prevalence is comparable if not less than in patients after a Rastelli procedure. More patients and longer follow-up are needed to confirm the results of this study.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

PW3-2

Microdialysis – a new diagnostic tool in research of congenital heart surgery

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Objective: Capillary leak syndrome (CLS) affecting especially neonates and young children after cardiac surgery requiring cardiopulmonary bypass (CPB), contributes to higher morbidity. The mechanism involving the related inflammatory processes is poorly understood. We investigated whether different cytokines, measured with microdialysis, can monitor local inflammation in adipose tissue one of the target organ of CLS and predict the development of CLS on cytokine level, before severe clinical signs appear.
Methods: We performed a prospective feasibility study with serial measurements of inflammatory response in subcutaneous adipose tissue up to 24 hours after surgery. The cohort consists of 23 neonates and infants (median age 155, range 6–352 days; median body weight 5.4, range 2.6–9.2 kg) underwent congenital heart surgery with CPB. Microdialysis catheter were introduced in one lateral thigh subcutaneously using a velocity of 1.0 μl/min. Serial microdialysis analysis for cytokines (interleukin [IL]-6, IL-8, IL-10) and anaphylatoxin (C3a) were performed. CLS was quantified by X-ray subcutaneous-thoracic ratios (S/T). We studied age-related differences of inflammatory response.
Result: Median bypass time was 150 min (range 42–432 min) and aortic crossclamp was 76 min (range 0–188 min). In all patients pro- and anti-inflammatory and complement activation were verifiable. After onset of surgery the C3a levels distinguished rose (167 ng/ml), followed by a release of IL-10 at the end of CPB. The highest levels of IL-6 (55 pg/ml) and IL-8 (66 pg/ml) were detected two hours after CPB. Six of 23 infants developed postoperative CLS. These patients disclosed a gentle but significant second rise 8 to 10 hours postoperatively (CLS 64 ng/ml vs. non-CLS 24 ng/ml; $p < 0.01$). We could show an aged-related difference in the release of IL-6 and C3a. Younger age ($p = 0.02$), longer bypass time ($r = 0.48$; $p = 0.021$), higher inotropic demand ($r = 0.67$; $p = 0.001$) and longer intubation time ($r = 0.63$; $p = 0.001$) correlated closely with the development of CLS.
Conclusion: With diagnostic microdialysis it is feasible to give valid data about local inflammatory response subcutaneously in paediatrics during and after CPB. We are able to disclose age-related differences in the inflammatory response. Our results implicate the possibility to predict CLS early before severe clinical signs appear.

PW3-3

A comparative histopathological study of heparin coated and uncoated polytetrafluorethyle grafts in children with congenital heart defects

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Introduction: Polytetrafluorethylene (PTFE) grafts are used to ensure pulmonary blood flow in children presenting with complex congenital heart defects. Recently, heparin coated grafts are available and are believed to improve inherent graft problems such as thrombosis and excessive and incomplete neointima formation or occlusion. We aimed at comparing the potential histopathological differences of the neointima in uncoated (UCG), and heparin coated (HCG) PTFE grafts.
Methods: 16 grafts (8 UCG, 8 HCG) were explanted. 3 grafts with stents in the distal anastomosis (2 UCG, 1 HCG) were excluded from further histopathological comparison. The specimens were fixed in formalin and embedded in paraffin or in methylmethacrylate. Tissues were characterized by standard and immunohistochemical staining. The thickness of pseudointima proliferation was graded as follows: 0 = no cell layers, 1 = few layers < 100μm, 2 = partial layers > 100μm, 3 = complete layers <300μm, 4 = complete layers >300μm, 5 = occlusion.
Results: Mean shunt size was 3.4 ± 0.2 mm in UCG, and 3.1 ± 0.2 mm in HCG ($p = 0.053$). Mean time of implantation was 163 ± 75 days in UCG, and 97 ± 52 days in HCG ($p = 0.091$). There were no significant differences in the proportion of patients with functionally single ventricle, body surface area, age at implantation, or implantation type, between both groups. Graft occlusion did not occur. Unplanned graft explantation due to cyanosis was performed in 1 patient in each group. Partial thrombus formation was observed in 1 UCG ($p = 0.462$). There was complete endothelialization in 67% of UCG, and 86% of HCG ($p = 0.559$). The grade of pseudointima proliferation was 1.8 ± 0.4 in UCG, and 1.7 ± 0.5 in HCG ($p = 0.646$).
Conclusions: The histopathological work-up of PTFE grafts revealed equally partial endothelialization and discrete pseudointima proliferation in both groups. The process of endothelialization may be faster in HCG. However, this small series could not demonstrate a superiority of HCG over UCG.

PW3-4

Early Ross-Konno surgery for treatment of critical aortic stenosis

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Background: newborns with critical aortic stenosis (CAS) suffer from severe left ventricular (LV) hypertrophy and dysfunction. Interventional aortic valve dilation usually leaves significant increased LV myocardial stress. Early aortic valve replacement may allow improved LV recovery and long-term outcome.
Aim: to review our experience with neonatal RK surgery.
Patients: Between 10/2000 and 1/2013 22 children with CAS, median age: 17d (7–547d); median weight: 3,03 kg (2,18–9,2 kg) underwent early RK surgery in our institution. Hypoplastic or interrupted aortic arch was additionally corrected in 9 patients, 5 of them had a VSD closure. Severe endocardial fibroelastosis (EFE) was present in 8 neonates. Seven had a fetal aortic valve dilation, 16/22 a postnatal aortic valve dilation. A 12 mm Contegra valve was used as a right-sided conduit in 6 patients, homografts from 8–14 mm in 16 children. Median follow-up was 5,5 years (1,0–12,3 years)

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Results: early mortality was 3/22 (14%), there was no late mortality. 1 patient required a pacemaker due to complete heart block, 1 patient had a mitral valve replacement. Conduit replacement was necessary in 9 patients after a median period of 2,2 years (0,6–11 years), 3 had a second replacement median 3,3 years (2,5–4,1 years) later. Neo-aortic valves showed excellent function without gradient, no aortic regurgitation in 8, grade I in 15 patients. Aortic valves showed good growth (median z-score after 3 years: 1,92; range: −0,6–2,94), valve function remained stable, however z-scores of the aortic sinuses were significantly larger (median 2,77; range: 0,79–4,51; $p < 0.0001$). Out of 8 patients with severe EFE and LV Dysfunction there were 2 neonatal deaths, LV SF improved significantly from median 11,8% (5–23%) to 30,7% (10–41%). PA pressures were normal in all.
Conclusion: in neonates and young children with CAS, early RK seems to be a safe and effective treatment to unload the LV and to allow recovery of LV function. Early conduit replacement may be necessary. Neo-aortic valve showed good growth and function but dilation of neo-aortic sinuses may occur and may become a concern in the future.

PW3-5
An old surgical approach renewed: Pulmonary Artery Banding for treatment of Left Ventricular Dilated Cardiomyopathy
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Background Dilated cardiomyopathy (DCM) in childhood has a considerable morbidity and mortality. Based on the experience of Pulmonary artery banding (PAB) in patients with sub-aortic right ventricle, we adopted this old surgical approach of PAB in young patients with left ventricular dilated cardiomyopathy (LV-DCM) with preserved right ventricular function.
Methods: A retrospective single center observational study; evaluation of transcatheter dilatable surgical PAB in infants and young children with LV-DCM.
Results: Since April 2006 17 infants and two toddlers with LV-DCM who were referred to our centre for Heart Transplant received a PAB. Five of the patients underwent additional mitral valve repair or replacement, repair of a left-sided Anomalous Pulmonary Vein Return, re-implantation of an ALCAPA. All patients had been on catecholamines. There was no hospital mortality. All patients showed clinical improvement.
In the 12 patients without additional operation the pressure gradient across the PAB increased significantly within 3–6 months. The LV ejection fraction increased from 15% (median) pre-PAB to 43% at discharge home, and 47%, 3–6 months later. The median LVEDD z-score decreased ($p > 0.001$) from +7.3 to +3 and +1.3, respectively. Plasma B-type natriuretic peptide levels decreased, corresponding to the functional class improvement ($P < 0.001$). Eight children were subsequently (partially) de-banded by trans-catheter technique and are currently functional class 1. Two patients with non-compaction DCM deteriorated 5 and 6 months after PAB-de-banding and died. One of the five patient with additional cardisc surgery died during the follow-up.
Conclusion: In young children with LV-DCM and preserved right ventricular function, PAB can lead to an improvement of left ventricular and mitral valve function by ventricular interaction and might offer a way to delay or even prevent transplant.

PW3-6
Early results and long-term follow-up after mechanical circulatory support (MCS) with a variety of devices in children
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Introduction: MCS is increasingly used in children when conventional medical treatment fails. New pediatric sized devices have been developed in the last several years. Nevertheless, poor overall survival and long-term outcome have been observed. We report our early and late outcome as well as the health-related quality of life (HRQoL).
Methods: Between 2001 and 7/2012, a total of 51 children, median age 3 months (range 4d–17y), underwent 53 MCS courses with a median supporting time of 3 days (range 1–248). MCS (9 different systems) included Extracorporal Membrane Oxygenation (v-v-ECMO, n = 2), Extracorporal Life Support (v-a-ECLS n = 33), and the use of Ventricular Assist Devices (VAD n = 9). In some patients, we took a switch from one system to the other: v-v-ECMO/v-a-ECLS (n = 6) and v-a-ECLS/VAD (n = 3).
Diagnosis: Myocarditis n = 5, Cardiomyopathy n = 5, in n = 33 weaning from bypass failed after surgery, 6 others.
For HRQoL, all 21 long-term survivors answered standardized questionnaires (Kiddy-Kindl®, Kid-Kindl®, Kiddo-Kindl® or SF36®) and were graded according to age into 5 groups: 0–3 years (n = 6), 4–7 years (n = 6), 8–12 years (n = 2) and patients older than 13 years (n = 6).
Results: 51 children underwent 53 MCS courses. 64% survived (55% weaned, 9% transplanted). Two weaned patients underwent later transplantation (after 4 and 10 months respectively). Despite successful weaning, 9 died over the course of hospitalization and 25 (47%) were discharged home, 4 dying later.
Despite individual cases with motoric problems or cognitive delay, HRQoL showed a favorable outcome for selected patients regarding their social and psychomotoric development and their morbidity in the view of their parents (patients younger than 12 years), and a most favorable outcome in their own sensation (patients older than 13 years).
Conclusion: Overall, long-term survival and QoL is satisfactory. Nevertheless, a long-term follow-up program of these severely ill children should be structured to detect early disorders to offer individual support.

PW3-7
Extracorporeal Life Support for Children with Late (post discharge) Rejection after Heart Transplantation: 10 year single center experience
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Objectives: To describe the management, course and outcome of late (following initial hospital discharge) rejection with acute haemodynamic compromise supported on extracorporeal life support (ECLS) in children with orthotopic heart transplantation (OHT).
Methods: Review of all children with acute haemodyamically relevant rejection requiring ECLS following OHT between 2/2002 and 10/2012.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Results: Of 197 consecutive children undergoing OHT (84 male; mean age 8.3+5.7 (range 0.1–18.8 years), 187 children survived and were discharged from hospital. Seven patients presented with severe haemodynamic compromise after initial hospital discharge following their transplantation (of whom one patient had been transplanted elsewhere). Mean follow-up was 5.0+3.1 (range 0.1–10.6) years.
All 7 children required ECLS, two were placed on to ECLS following in-hospital cardiac arrest. Median duration of ECLS was 8 (range 5–15) days. All children survived to decannulation with one death after ECLS from sepsis 20 days after presentation. The median (range) duration of inotropic requirement post ECLS was 11 (5–27) days, the median ventilation time was 8 (7–30) days, median ICU length of stay was 14 (10–54) days and median hospitalization was 24 (19–118) days.
In all patients, ventricular function normalized (FS > 28%) within 10 (7–22) days. There was significant short-term morbidity; however, all survivors have a good functional status with no significant apparent neurological sequelae.
Conclusion: ECLS appears to be a good rescue therapy for children with severe acute rejection post OHT, refractory to conventional treatment, leading to good medium-term outcome.

PW3-8

Left coronary artery Doppler systolic flow reversal is associated with adverse myocardial events post arterial switch operation for Transposition of the Great Arteries

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Introduction: Transposition of the great arteries (TGA) is generally repaired using the arterial switch operation (ASO). Early complications are mainly following coronary artery related events. Intra-operative coronary flow patterns may predict clinical outcome.
Objective: The objectives of this prospective study were 1) to determine whether coronary Doppler patterns intra-operatively predicted an adverse outcome, 2) to compare transesophageal (TEE) vs. epicardial echocardiographic assessment of coronary arteries post ASO.
Methods: Patients with TGA undergoing the ASO were eligible. All patients (when technically feasible) underwent a TEE plus an epicardial echo intra-operatively. All predischarge transthoracic echocardiograms were reviewed for ventricular function. The primary clinical endpoint was a composite myocardial ischemic event (any of: post-operative ST changes, ventricular tachycardia, need for ECMO support). Correlation and Kappa statistics were used to assess agreement between the imaging modalities.
Results: From May 2009 – Dec 2012, 36 patients (26 male, birth weight 3260+590 grams) were recruited. Of those, 11 had a ventricular septal defect, 27 had usual coronaries (1LCx2R), 6 had 1L2RCx, and 2 were intramural. The median age at the time of the ASO was 10 days, IQR: 6–21 days, N = 28 had TEE + epicardial, 4 TEE only, 4 epicardial only. N = 7/36 (19%) patients had an adverse myocardial event (5 ST changes, 2 ventricular tachycardia (1 also ECMO), 1 death). Systolic flow reversal in the left coronary artery was associated with the primary endpoint (86% vs. 10%, $p < 0.001$). By TEE, the median VTI for the right coronary artery was 0.112, IQR 0.084–0.159 and the left coronary was 0.117, IQR 0.094–0.150. There was excellent agreement for global assessment of function between TEE and epicardial (K = 0.925) and visualization of the left coronary (91% visualized with both), and strong agreement (92% visualized with both) of the right coronary.
Conclusion: Systolic flow reversal in the left coronary artery is associated with myocardial ischemic events post ASO. Intra-operative evaluation of coronary artery flow patterns is a potentially useful tool and should be considered for all ASO procedures.

PW3-9

Changes of left ventricular rotation from Infancy to Adulthood

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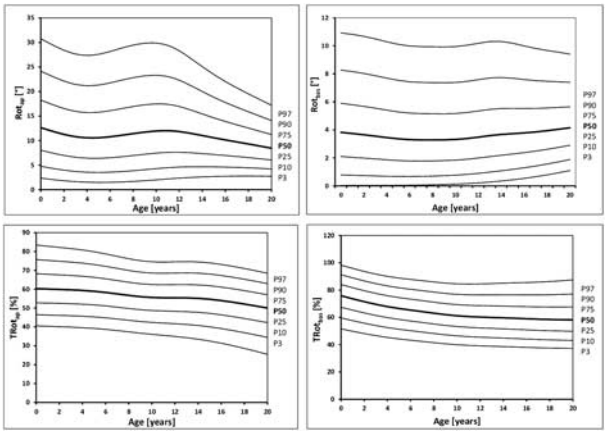
Introduction: Rotation is an important component of left ventricular deformation. We investigated the impact of heart rate and age on the amount and timing of rotation in healthy subjects.
Methods: 158 healthy children (78f, 0–20y, median 9.6y) were investigated with Speckle tracking echocardiography (STE) using apical and basal short axis loops. Data were recorded using a Vivid 7 machine (GE), calculation by dedicated Speckle-tracking software (Echopac). Peak systolic apical (Rot_{ap}) and basal rotation (Rot_{bas}) as well as timing ($TRot_{ap}$, $TRot_{bas}$) of these events were assessed. Statistical analysis was performed using Pearson's correlation and LMS-method to create percentiles.
Results: There was a strong correlation between $TRot_{ap}$ and $TRot_{bas}$ in comparison to heart rate ($TRot_{ap}$:r = −0.7, Trotbas: r = −0.6, $p < 0.001$) and age ($TRot_{ap}$:r = 0.6, $Trot_{bas}$:r = 0.5, $p < 0.001$). Throughout all ages counterclockwise Rot_{ap} appears before clockwise Rot_{bas} and with higher magnitude in infancy. Normalization to percentage of cardiac cycle results in less decrease of $TRot_{ap}$ than $TRot_{bas}$ during maturation. Quantitative Rot_{ap} has two peaks in infancy and at the age of 10–12 ys, whereas $TRot_{bas}$ remains relatively constant.
Conclusions: Maturation of the myocardium during childhood strongly correlates with heart rate and age. The delay of timing between apical and basal rotation is higher in infancy whereas timings of all rotational parameters shorten if getting normalized to cardiac cycle length with aging. Creation of percentiles is a valuable tool to follow changes in individual patients especially in case of dyssynchrony.



https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

PW3-10

Impact of age and gender on longitudinal indexes of left ventricular systolic function in a normal paediatric population

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Background: Age and gender may have a relevant impact on left ventricular (LV) longitudinal function in normal adults, but these effects in paediatric patients have not been fully explored. We aimed at investigating these associations in a population with relatively wide age range.
Methods: Fifty-six patients aged 1–16 years (age 7.5 ± 3.8 years, 51.8% female), with unremarkable history and no evidence of cardiovascular or systemic disease, were studied by echocardiography. Tissue Doppler was used to assess LV peak longitudinal velocity at systole (S'), early diastole (E'), and late diastole (A'). Values measured at the septal and lateral site of the mitral annulus were averaged. The ratio of peak early diastolic LV filling velocity to E', an established index of LV filling pressure, was also calculated.
Results: Average S', E' and A' were 8.3 ± 1.6 cm/s, 15.2 ± 2.6 cm/s, 6.4 ± 1.4 cm/s. Higher velocities were found at the lateral site than the septum for both S' and E' ($p < 0.0001$), but not for A' ($p = 0.80$). Average E/E' was 5.9 ± 1.5, with lower values at the lateral site than the septum (5.3 ± 1.3 vs 7.2 ± 1.7, $p < 0.0001$). Age showed positive relationships with S' ($R = 0.49$, $p < 0.0001$) and E' ($R = 0.44$, $p < 0.0001$), and negative relationships with A' ($R = -0.28$, $p = 0.034$) and E/E' ($R = -0.40$, $p < 0.0001$). The association with age was stronger for lateral S' ($R = 0.53$, $p < 0.0001$) than septal S' ($R = 0.29$, $p = 0.030$; $p < 0.0001$ by comparison of correlation coefficients) whereas those with E', A', and E/E' were not significantly different between the two annular sites. Male and female subjects showed similar S' (8.4 ± 1.7 vs 8.2 ± 1.5 cm/s, $p = 0.77$), E' (15.8 ± 2.7 vs 14.7 ± 2.4 cm/s, $p = 0.09$), A' (6.4 ± 1.5 vs 6.4 ± 1.3 cm/s, $p = 0.94$), and E/E' (6.0 ± 1.1 vs 6.1 ± 1.4 cm/s, $p = 0.75$). No gender differences were found in septal or lateral velocities as well. In multivariable analysis adjusting for confounding factors, age remained positively associated with S' ($\beta = 0.488$, $p < 0.001$) and E' ($\beta = 0.342$, $p < 0.001$), and negatively with E/E' ($\beta = -0.492$, $p < 0.001$), whereas the association with A' was no longer evident ($\beta = 0.185$, $p = 0.55$). Gender was not associated with any longitudinal index.
Conclusions: In a population of paediatric patients aged 1–16 years, age, but not gender, had a considerable impact on LV longitudinal dynamics.

PW3-11

Surgery impacts right atrial function in tetralogy of Fallot

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Objective: To analyse the impact of surgery and pericardial integrity on right atrial function and total heart volume variation in the setting of pulmonary valve insufficiency.
Methods: Right atrial function and total heart volume variation were analysed in two subgroups of patients with pulmonary valve insufficiency in comparison to healthy controls: group I with surgically repaired tetralogy of Fallot (n = 20 patients) and group II after balloon angioplasty of pulmonary valve stenosis in atients with isolated valve disease without surgery (n = 7 patients). Volumetric analysis of MRI data revealed parameters of atrial function (reservoir-, conduit- and pump-function and cyclic volume change) and of total heart volume (enddiastolic and endsystolic total heart volume and the variation). Statistical analysis included uncorrected and corrected pairwise comparisons and the calculation of groupwise Pearson correlation coefficients.
Results: In group I with a pulmonary regurgitation fraction of 31.0 ± 14.9%, right atrial function was clearly impaired, with reduced reservoir and elevated conduit function, and total heart volume variation was elevated to 13.9 ± 3.4%. In group II, with a pulmonary regurgitation fraction of 22.8 ± 6.9%, the values were close to normal, with unaffected atrial function and a total heart volume variation of 9.9 ± 3.3%.
Conclusion: The hydrodynamic effect of pulmonary valve insufficiency alone is likely not the only reason for impaired right atrial function and elevated total heart volume variation in Fallot patients, it is rather the scar in the right atrium, the injured pericardium and the disease itself that are responsible for the energetically unfavorable alterations.

PW3-12

Effect of chronic right ventricular volume overload on ventricular interaction in patients after Tetralogy of Fallot repair

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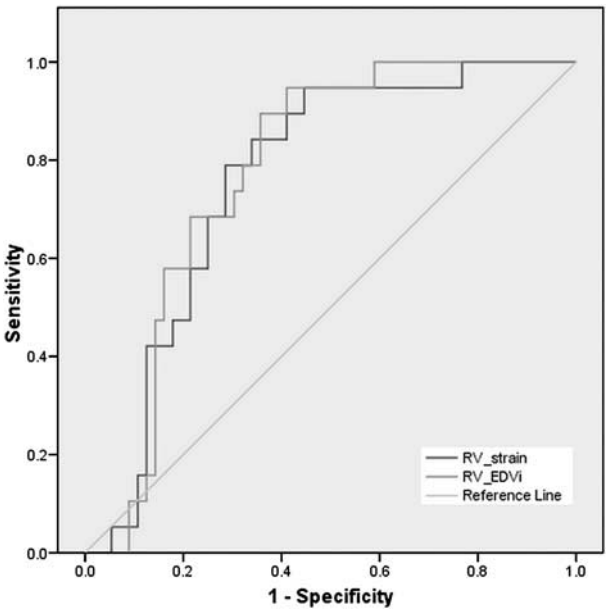
Background: Right ventricular (RV) volume overload results in RV dilatation in patients with pulmonary regurgitation after tetralogy of Fallot (ToF) repair. Due to ventricular interaction, RV dilatation influences left ventricular (LV) function but the LV myocardial mechanics have not been well studied in relationship with RV functional parameters.
Aim: To study the effect of chronic RV volume loading on LV myocardial mechanics in patients after ToF repair.
Methods: We studied 75 subjects: 50 patients after ToF repair, and 25 age-matched controls. All ToF patients and controls underwent an echocardiography at the time of a clinically indicated cardiac magnetic resonance imaging (cMRI). Myocardial deformation, including LV torsion was analyzed using speckle-tracking echocardiography.
Results: There was no difference between the two groups in LV EF measured by cMRI. LV end-systolic ε values were significantly reduced in ToF patients compared with controls : longitudinal (−18.4 ± 8.6 vs −22.1 ± 2.9%, $p = 0.007$), radial (44.3 ± 16.1 vs 65.2 ± 15.6%, $p < 0.001$ and circumferential (−19.1 ± 2.7 vs 21.6 ± 2.2%, $p < 0.001$). LV rotational mechanics are also significantly different with reduced basal and apical rotation and decreased LV torsion (8.7 ± 4.6° vs 14.7 ± 5.7°, $p < 0.001$). Especially basal rotation was very abnormal with 38% of ToF patients having counterclockwise basal rotation. Apical rotation was reduced but not reversed. Both RV end diastolic volume index (EDVi) and RV ε were good predictors of counterclockwise basal rotation, with area under the ROC curve of 0.77 (95% CI: 0.67–0.88) and 0.76 (95% CI: 0.65–0.87) respectively, $p < 0.001$, see figure. Cutoff values of

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

160 ml/m^2 for RV EDVi and −24.5% for RV ε had the best sensitivity and specificity for predicting counterclockwise basal rotation, OR = 7.3 (95% CI: 1.9–30.6, p = 0.001) and 9.4 (95% CI: 2.4–40, p < 0.001), respectively.
Conclusion: All parameters of LV deformation appear to be affected in children and adolescents after ToF repair despite preserved LV EF. Especially LV basal rotation is very abnormal and often reversed, related either to RV dilatation or decreased deformation.



PW3-13
Longitudinal right ventricular function quantified by speckle tracking predicts exercise capacity in adults with transposition of the great arteries
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Background: Systemic right ventricle (sRV) dysfunction is common in transposition of the great arteries (TGA) and is associated with increased mortality. Assessment of sRV function is a key point in the follow-up, but is challenging and its relation with exercise capacity is controversial. We hypothesized that a quantitative measurement of longitudinal ventricular function may be more sensitive in detecting early myocardial damage compared with conventional measures such as ejection fraction, and more related to exercise capacity.
Objectives: 1/To characterize sRV function with standard and speckle-tracking echocardiography, comparing with normal RV and LV function in healthy subjects, and with the gold standard, CMR. 2/To validate the imaging parameters by quantifying their relation to objective measurements of exercise capacity.
Methods and results: The study population consisted in 54 patients with a sRV (47 D-TGA and 7L-TGA, 32 ± 5 years old). Conventional echography and bidimentional strain, cardiac magnetic resonance (CMR) imaging and cardiopulmonary exercise (CPET) were performed on consecutive patients on the same day. Twenty five healthy subjects were matched for age and sex with 25 TGA patients in NYHA I and with the highest peak oxygen uptake at CPET, defining the asymptomatic TGA group. Sub-aortic RV longitudinal peak systolic 2D-strain was significantly reduced in the "asymptomatic" TGA patients compared to LV and RV of the control group. A base to apex gradient was observed regardless RV load conditions. RV transverse 2D-strain was not different between the asymptomatic patients group and control group, and between symptomatic and asymptomatic TGA group. In multivariate analysis, longitudinal 2D strain of the inter-ventricular septum was significantly correlated with RVEF (r = 0.3047, p = 0.0398), while only longitudinal 2D strain of the RV free wall was significantly correlated with percentage of the predicted peak oxygen uptake (standard coefficient = 0.621, p = 0.0297), independently of strain of the inter-ventricular septum, and CMR RVEF.
Conclusion: in sRV longitudinal shortening remains predominant. Longitudinal 2D-strain of the systemic RV free wall is predictive of exercise capacity, and seems to be more sensitive than RVEF to detect early myocardial damage.

PW3-14
Isolated left ventricular non compaction: Relationships between MRI criterias for non compaction and clinical events
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Background: Isolated ventricular non compaction is a congenital cardiomyopathy, based on an arrest of normal embryonic myocardium development and characterized by the presence of a two-layered myocardial structure with a compacted epicardial band and a non compacted endocardial layer of prominent trabeculations with deep intertrabecular recesses. Ventricular non compaction is sometimes complicated by ventricular dysfunction and heart failure, malignant arrhythmias or cardioembolism. The aim of our study is to look for a potential relashionship between MRI's non compaction criteria and these clinical events.
Methods and results: Among all cardiovascular MRI realized at the University Hospital of Nantes between 2004 and 2012, 120 patients presented MRI's non compaction criteria, but 45 of these patients had another associated cardiomyopathy and so were excluded. 75 patients fulfilled the diagnosis of isolated ventricular non compaction and were included in the study (63% male, mean age 43 ± 15 years).
LV ejection fraction, LV volumes, global LV mass, compacted LV mass, number of non compacted segments and non compaction score were measured. Non compaction score was the sum of the ratio of the thickness of non compacted to compacted myocardial layers superior to 2.3, measured in the diastolic phase.
We tried to establish some associations between clinical events and MRI data. Mean LVEF was 53 ± 11%, negatively correlated with non compaction score (p = 0.04). Mean number of non compacted segments and mean non compaction score were significantly higher in patients with stroke (respectively p = 0.056 and p = 0.014). Nevertheless, there were no statistical association between ventricular arrhythmias and LVEF, number of non compacted segments or non compaction score.
Conclusion: Our study, which is, so far, the largest prognostic MRI study of isolated ventricular non compaction, shows a clear association between non compaction extension and LVEF degradation or stroke incidence but no evident relationship with ventricular arrhythmias risk. Thus, according to our results, MRI evaluation of non compacted extension did not seem to be a good predictor for the stratification of the risk of ventricular arrhythmias.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

PW3-15

Measurement of Flow mediated vasodilatation in Patients with a History of Kawasaki disease

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Background: It is well known that some patients with a history of Kawasaki disease (KD) with persistent coronary artery lesions (CAL) develop post-inflammatory arteriosclerosis, wherein the lesions consist mainly of hyalinized fibrous tissue, and that calcification is usually observed in some of these patients with massive aneurysms. The mechanism of calcification in the coronary arteries of KD patients is not well understood. In terms of the temporal sequence, however, the basic cause is believed to be functional damage to vascular endothelial cells, which undergo organic changes resulting in the formation of sclerotic lesions through the involvement of inflammatory mechanisms or oxidative stress; eventually, calcium deposition occurs at these sites. In order to prevent the development of calcification, which is regarded as a risk factor for coronary vessel events, it would be more appropriate to initiate treatment at the stage of functional damage. Therefore, the focus on detecting functional damage to endothelial cells has increased. The flow-mediated vasodilatation (FMD) method, which utilizes reactive hyperemia induced by the release of vascular endothelial nitric oxide, has received increasing attention in recent years due to its sensitivity. In the present study, by using FMD, we investigated the vascular functional damage in patients with KD.
Methods: We included 31 patients with a history of KD. Eleven patients were CAL(–) and 20 were CAL(+); of the 20 CAL(+) patients, 7 were negative and 13 were positive for calcification on multi-detector computed tomography (MDCT). We performed FMD by using a UNEX testing device (UNEX Corporation, Japan).
Results: FMD values were significantly lower in CAL(+) patients compared with CAL(–) patients ($p = 0.0001$). Among the CAL(+) patients, FMD values were significantly lower in those who were positive compared with those who were negative for calcification ($p = 0.0027$). No medication that may have affected the results of FMD was identified.
Conclusion: Our study suggested that FMD measurements may predict the development of arteriosclerosis and the appearance of coronary artery calcification in patients with a history of KD. Thus, FMD may provide an important means of assessing changes in coronary artery damage over time in KD.

PW4-1

In search of an improved QT correction formula in children

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Introduction: The Bazett and Fridericia formulae are most frequently used to correct QT interval for heart rate in children. At higher heart rates they over- or under-correct the QT interval. We set out to compute the best formula in our children population.
Methods: we enrolled a cohort of 650 healthy children. In quiet state a digital 12 lead electrocardiogram (50 mm/second) was recorded and stored. The QT and RR intervals were digitally measured in lead 2. The QT/RR curve was fitted with 2 different regression analysis, a linear regression for constant α, whereby $QTc = QT + \alpha \times (1-RR)$, and a log-linear regression analysis for constant α whereby $QTc = QT/RR\alpha$. Furthermore, linear regression analysis of QTc/RR for the two formulae were performed (least squares method), obtaining slope and R2. A slope and R2 close to zero were judged to eliminate the effect of heart rate on QT interval. Same analysis was performed using Bazett and Fridericia as comparison.
Results: Mean age: 2,2 years, SD + 4.0 (range 0–18 years). Mean QT 301+35 ms, RR mean 530 + 137 ms; QTc Bazett 414+17 ms; QTc Fridericia 370+16 ms. Computed linear regression formula was $QTc = QT+0.2308*(1-RR)$, QTc linear = 409+16 ms. Computed log-linear regression formula was $QTc = QT/RR0.44$, QTc log –linear = 402+21 ms. Linear Regression plots of QTc against RR intervals: QTc linear slope 0.000005, R2 10-7; QTc log-linear slope 0.0001, R2 5*10−5; QTc Bazett slope – 0.06, R2 0.16; QTc Fridericia slope 0.053, R2 0.15.
Conclusion: The new linear and log – linear regression formulae corrected well, linear regression being marginally better. Both these new derived formulae look promising, best corrected the effect of RR on QT interval, even in infants, and showed superior dissociation of the QTc interval from RR interval (least slope and lowest R2). Both formulae are superior to the Bazett and Fridericia formulae. Further evaluation is in progress.

PW4-2

Early recognition and treatment of critical arrhythmias in adults after the atrial switch operation for transposition of the great arteries: remote monitoring vs standard follow-up

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Background: Adults with transposition of the great arteries (TGA) after atrial switch repair have an increased risk for arrhythmias and sudden cardiac death. We analysed whether a remote monitoring (RM) system as part of an implantable cardiac device contributes to timely recognition and improved treatment of critical arrhythmias in this patient population.
Methods: All consecutive TGA patients requiring a pacemaker or cardiac resynchronization therapy with or without implantable cardioverter defibrillator at the Medical University Graz, Austria and the German Pediatric Heart Centre St. Augustin, Germany between 2008 and 2011 were included. RM-detected arrhythmias, abnormalities of the cardiac device integrity and reaction times from event transmission until acknowledgement via email and clinical decision making were analysed and compared to standard follow-up.
Results: In 11 adult patients 17 arrhythmias were detected in 10 (91%) patients of whom 8 patients (80%) indicated no symptoms. Mean time interval from transmission to acknowledgement was 2.4 (0–4.5) days. Clinical decision making was advanced by a mean of 77.5 (10–197) days compared with conventional follow up and implied adaption of antiarrhythmic medication in 8 patients, electrical cardioversion in 2, overdrive pacing in 1 and radiofrequency ablation in 2 patients. A coronary sinus lead fracture was identified in one patient followed by successful replacement.
Conclusions: RM enables early detection of tachyarrhythmias followed by optimization of medical treatment and potentially life-saving antitachycardic interventions in adults after atrial repair of TGA.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

PW4-3

Result of implantatable cardioverter-defibrillator in children with hypertrophic cardiomyopathy: an european experience

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Background: Hypertrophic cardiomyopathy (HCM) is an important cause of life-threatening arrhythmias. Demonstrated risk factors (RF) guide primary prevention indication of cardiac defibrillator implantation in adult. Attitude toward pediatric patient is less consensual.
Objective: Describe result of primary and secondary prevention cardiac defibrillator implantation in pediatric patients.
Materiel and method: Multicenter retrospective study from 5 centers in Europe. 45 patients under 20 years old were gathered, 16 from Paris, 9 from Zurich, 7 from Madrid, 7 from Roma and 6 from Ljubljana.
Results: Among the 45 patient 31 are male. Median age at implantation is 12.4 years old (3–20). Family history of HCM was described for 20 patients (44%). Most of the patient, 35 (78%), were implanted for primary prevention of sudden death, 10 (22%) for secondary prevention. The average number of risk factor per patient is 2: 11 patients were implanted for only 1 RF, 14 patients had 2 RF and 10 patients had 3 or more RF. A great majority of them (26/45) were implanted transvenously. Appropriated shocks were experimented by 12 patients (27%): 5 in the primary prevention group (14%) and 7 in the secondary prevention group (70%). Thirteen patients had an ICD-related complication: 8 inappropriate therapies, 4 device or lead failures and 1 delayed therapy. Two patients died and one had major neurological complication after electric storm.
Conclusion: The rate of appropriated shock in the secondary prevention group is significantly higher than in the primary prevention group. The annual rate of appropriated shock in the primary prevention group is very low but the complications are frequent. This result suggests that the demonstrated risk factor for adult may not be accurate at the pediatric age.

PW4-4

Catheter ablation of focal atrial tachycardia in pediatric patients: A ten year single centre experience using modern mapping systems

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Introduction: Experience of catheter ablation of focal atrial tachycardia (FAT) in pediatric patients is still limited and data have mainly been gathered prior to the introduction of modern 3D mapping and navigation systems into clinical routine. In these studies, conventional catheter ablation of FAT was associated with relatively low success rates and long fluoroscopy times in comparison to ablation of accessory pathways and atrioventricular nodal reentrant tachycardia.
Patients and methods: We retrospectively reviewed 17 consecutive pediatric patients and young adults with a median age of 15 [4–30] years and a mean body weight of 47.5 [16–73] kg who underwent electrophysiological study (EPS) for FAT. Indications included recurent supraventricular tachycardia (n = 9), permanent tachycardia (n = 4), tachycardia-induced cardiomyopathy (n = 3), and side effects of antiarrhythmic medication (n = 1). For EPS the Navx® system (n = 7), the non-contact mapping system (n = 6) or the LocaLisa® system (n = 4) were used, respectively. Cryoablation was performed in selected patients with foci close to the AV node, in all other patients radiofrequency (RF) was the primary energy source for catheter ablation.
Results: In 16 patients a total of number of 19 atrial foci (13 right-sided and 6 left-sided) could be targeted during EPS. In the remaining patient FAT was not present/inducible. Using modern mapping systems acute success was achieved in 14/16 patients (87.5%) with a median number of 10 [1–31] ablation lesions. Ablation was unsuccessful due to an epicardial location of a right atrial focus in one patient and due to the proximity of a focus to the HIS bundle in the remaining patient with cryoablation resulting in transient second-degree AV block. Mean procedure time was 210.4 ± 78.1 min, mean fluoroscopy time was 12.6 ± 4.9 min.
Conclusions: Compared to standard mapping, 3D non-fluoroscopic approach for pediatric FAT resulted in improved success rates and significantly reduced fluoroscopy times. 3D mapping and RF ablation provided improved clinical quality of care, therefore, this approach should be considered early in the course of treatment of this tachyarrhythmia.

PW4-5

JT and other parameters of ventricular repolarization in healthy children

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Introduction: The rate-corrected (Bazett's formula) QT interval (QTc) is the conventional measurement of ventricular repolarization. Prolongation of the QTc interval increases risk of ventricular arrhythmia and sudden cardiac death. Intraventricular conduction abnormalities complicate evaluation of the QTc interval. In these patients JT rate-corrected (JTc) interval may be more accurate and sensitive measurement of ventricular repolarization by eliminating of QRS complex duration. Very little is known about JT interval and its derivatives in children and adolescents.
Methods: We studied 131 healthy children (64 girls and 67 boys), at age from 2.3 to 18.5 years (mean 9.07 ± 3.89 years). In the course of the study we analyzed standard, 12-lead electrocardiograms (ECG) at a paper speed of 50 mm/s. We manually measured: RR, JT, JTp (from J point to the peak of T wave) and TpTe (from the peak to the end of T wave) intervals in all 12 leads and mean results were calculated from three consecutive cardiac cycles.
Results: Due to technical reasons not in all leads were possible to obtain measurements (flattened or small amplitudes of T waves), the majority of such cases were observed in leads III (25.19%) and AVL (19.08%). JT interval in all leads but AVL were similar ($p > 0.05$) and ranged from 180 to 365 ms. In lead II mean intervals were: JT = 246.18 ± 29.23 ms, JTc = 295.1 ± 23.37 ms, JTp = 178.17 ± 28.84 ms, TpTe = 63.4 ± 9.16. Calculated JT dispersion was 34.20 ± 18.89 ms and JTc 41.14 ± 23.22 ms. The JT, JTp, TpTe intervals increased with age and was longer in teenagers than in younger children. In JT, JTp, JTc and dispersion there were no gender differences ($p > 0.05$).

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

TpTe was the longest in lead V3 and longer in boys. The JTc interval did not change with age.
Conclusions: In measurement of JT interval and its derivatives leads III and AVL should be avoided. In healthy children, JT in all leads but AVL were similar, but the TpTe interval was the longest in lead V3. In older children measured intervals were longer, rate-corrected interval did not change with patients age.

PW4-6
600 percutaneous catheter ablations in children and adolescents – one center experience
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Introduction: Percutaneous catheter ablation (PCA) in pediatric patients was first described in early 1990s. High efficacy of the method and low complication rate allowed these procedures to be offered as first-line therapy to many children with tachyarrhythmia. In our Institute the ablation therapy has been performed since 2003. We present our data.
Method and results: Six hundred ablation procedures were performed in 523 children and adolescents (259 girls and 264 boys). Age ranged from 3 months to 19.2 years (mean 14.9 ± 2.9 years), 470 of them had normal heart anatomy. The procedure targets were accessory pathways (AP): left – 174, right – 164, left and right – 4, atrio-ventricular reentry tachycardia (AVNRT) – 154, ectopic atrial tachycardia (EAT) – 27, long R-P' tachycardia - 11, junctional ectopic tachycardia (JET) – 5, multifocal atrial tachycardia/atrial fibrillation – 6, ventricular arrhythmia – 55 procedures. In the most of the procedures we used only two catheters diagnostic and ablation, which were introduced by puncture of femoral vessels. Radiofrequency (RF) catheter ablation was performed in 593 pts including CARTO XP system in 69 pts, crioablation in 7. Time of fluoroscopy ranged from 4.0 to 84.3 minutes (mean 26.7 ± 15.9 minutes), RF application time was from 0.7 to 30 minutes (mean 5.9 ± 4.99 minutes). The longest application time were in pts with Ebstein's anomaly (mean 14,5 minutes). The major complication was atrioventricular block follow by pacemaker implantation in 2 pts:1 boy with combine fascicular and AVNRT tachycardia and 1 infant with congenital JET (0,3%). In 4 pts - false aneurysm occurred (closed by thrombin injection), in 2 atriovenous fistula which closed spontaneously. In 66 (11%) children ablation has to be repeated, most often in patients with right AP (24/164, 14,6%), left AP (17/174, 9,8%) and ventricular arrhythmia (7/55, 12,7%).
Conclusion: In children and adolescents with tachycardia percutaneous catheter ablation is effective and save method of treatment, but recurrence of arrhythmia is possible. In patients with abnormal heart anatomy the procedure may be difficult and RF applications lasted longer.

PW4-7
The impact of an absent ductus arteriosus on clinical outcomes in fetuses diagnosed with Tetralogy of Fallot
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Objectives: Fetuses with Tetralogy of Fallot (TOF) almost universally have a patent ductus arteriosus (PDA). Two recent fetal cases of TOF had an absent PDA, requiring emergent intervention at birth. The objective of this study was to determine whether fetuses diagnosed with Tetralogy of Fallot (TOF) without a PDA have worse clinical outcomes compared with fetuses diagnosed with TOF+PDA.
Methods: All fetal cases of TOF between January 2000 and January 2012 were retrospectively identified from The Hospital of Sick Children (Toronto, Canada) database. Concomitant diagnoses of atrioventricular septal defect, pulmonary atresia, or absent pulmonary valve were excluded. Cases (TOF + no PDA confirmed at first postnatal echo) and controls (TOF + PDA, matched for gestational age (GA)) were reviewed. Optimal outcome was defined as valve sparing repair with no residual lesions. Student's t-tests and Fisher's exact chi-square were used to compare groups.
Results: A total of 115 fetuses were diagnosed with TOF: 11 (9%) had no PDA and 22 were matched controls (mean GA at diagnosis 32.1 ± 6.5 weeks, 30.8 ± 6.6 weeks, respectively). Cases had a higher proportion of right aortic arches (64% vs. 14%, $P < 0.001$). At birth, mean right outflow gradients were 31 mmHg (25–52) vs. 27 mmHg (21–42), $P = 0.30$. Fetal and postnatal echocardiographic data did not reveal significant differences in branch pulmonary artery sizes, pulmonary valve sizes, or ventricular function. No differences were identified for cyanosis at birth (2/10 vs. 7/10, $P = 0.67$), or early catheter intervention (5/10 vs. 4/22, $P = 0.12$). Overall survival was 9/11 in cases vs. 18/22 controls, $P = 1.00$. Optimal outcome rates were similar between cases and controls (4/11 (36%) vs. 5/21 (24%), $P = 0.68$).
Conclusions: The PDA does not appear to be relevant in fetuses with TOF to ultimate clinical outcome. The TOF physiology may allow for redistribution of pulmonary blood flow, enabling normal growth of the pulmonary vascular bed.

PW4-8
Long term follow-up of fetal cases with tricuspid valve anomalies
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Objectives of the study: Retrospective-prospective study of the characteristics and long-term outcomes of fetal cases with tricuspid valve anomalies, aiming to assess negative prognostic factors.
Material and methods: The echocardiographic data and long term outcome (6m–27 yrs) of 41 fetuses diagnosed between 1986 – June 2012 as Ebstein (Ebst) or non-Ebstein anomaly (NE), at 21–37 week's gestation (wg), median 29, were analysed. Nineteen cases had Ebst and 22 NE, one with mitral dysplasia and one with coarctation. Two fetuses had extracardiac anomalies, 3 Ebst had familial history of congenital heart disease, 1 mother was taking lithium and 1 gardenal. Following variables were compared in cases who died and in survivors: grade of tricuspid regurgitation (TR), Celermajer index (CInd), cardiothoracic ratio (CR), fetal hydrops (FH), pulmonary stenosis/atresia (PS, PAtr).
Results: Echocardiographic features: 13/19 fetuses with Ebst had a moderate-severe displacement of the TV and moderate-severe TR at presentation, 3 had pulmonary stenosis (PS) and 5 pulmonary atresia (Patr), 13/22 cases with NE had severe TR, 3 had PS and 5 PAtr. Seven had fetal hydrops (FH)–5 Ebst, 2 NE.
Outcome: Two Ebst and 1 NE opted for the termination of pregnancy, 3 Ebst died in utero (2 FH, 1 supraventricular

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

tachycardia). Thirty six cases were delivered at 31–39 wg. Six neonates died spontaneously at 1–7 days (3 Ebst, 3 NE). Five neonates with Ebst and 6 with NE were operated: 7 died, 4 NE survived. One Ebst died late at 3 yrs for resistant complex arrhythmias and severe worsening.
Total mortality was 17/38 cases (44,7%), 12/17 Ebst (70,6%), 5/21 NE (23,8%). Five of cases that died had FH, 2 severe arrhythmias, all a higher grade of TR, CInd > 1, CR > 0.65 and 9 had PAtr; the variables TR, CInd and PAtr were highly significantly different with respect to the survivors (p = 0.002–0.006). Twenty one cases with milder forms are alive at 6 m–27 yrs, stable or improved (5 Ebst, 16 NE).
Conclusions: Our data confirm a relevant mortality of severe tricuspid valve anomalies diagnosed in utero, main negative prognostic factors being the grade of TR, CInd and PAtr. Milder forms of both variants stabilized after birth.

PW4-9
The Bicuspid Aortic Valve in Turner Syndrome: a Fetal Morphology Study
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Background: Bicuspid aortic valve (BAV) is the most common congenital heart defect. Recent animal studies have suggested BAVs with different leaflet orientations to be of distinct etiologic origin. BAV leaflet orientation may have prognostic significance in terms of valve disease, aortic dilatation and necessity for intervention. BAV is particularly common in Turner syndrome (TS) and 95% of BAVs in adult patients with TS were shown to have anterior-posterior (AP) leaflet orientation, due to fusion of right- and left coronary leaflets. Data in fetal stages are scarce. Available date indicate that a larger proportion TS fetuses with BAV has a latero-lateral (LA) orientation of the leaflets (fusion of the right or left coronary leaflets with the non-coronary leaflet). This might indicate a worse prognosis with a higher risk of fetal demise in BAVs with LA orientation, possibly due to associated congenital heart disease. The aim of our study was to gain insight in the morphology of the aortic valve and its associated cardiovascular malformations in prenatal hearts of TS patients.
Methods and Results: We studied post-mortem heart specimens of 36 TS fetuses and 1 TS newborn. Abnormal aortic valve morphology was observed in 32 of 37 (86%) hearts: BAV was observed in 28 (76%) hearts, unicommissural aortic valve in 2 (5%) and aortic atresia in 2 (5%) hearts. In hearts with BAV that could be reinvestigated (n = 18), the leaflets showed AP orientation in 61% and latero-lateral (LA) orientation in 39%. There were no significant differences in the overall occurrence of additional cardiovascular malformations between hearts with AP- or LA orientation of the BAV leaflets. However, all hearts with LA orientation showed ascending aorta hypoplasia and tubular hypoplasia of the B-segment, as opposed to 55% and 64% of hearts with AP orientation respectively.
Conclusion: A large proportion of TS fetuses have abnormal aortic valve morphology. The proportion of BAV with LA orientation is higher in fetuses than in adults with TS. Fetal TS hearts with LA leaflet orientation were all associated with severe pathology of the aorta, which may contribute to a worse prognosis of LA oriented BAV in TS.

PW4-10
Fetuses with Tetralogy of Fallot – important information from prenatal diagnosis
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Objective: We analyzed prenatal history and perinatal outcome of fetuses with tetralogy of Fallot (ToF). The type of ToF, associated anomalies, karyotype, prenatal evolution and pregnancy outcome were analyzed.
Methods: Prospective echocardiographic examinations were conducted in the referral center for fetal cardiology since 2002. We performed retrospective review of database and recorded exams for the period 2002–2012.
Results: There were 99 fetuses (52 males, 45 females, 2 unknown) examined at average 28 weeks (median 28, 36% before 24 weeks). 68 fetuses had classic ToF, 24–ToF with pulmonary atresia (ToF-PA) and 4–ToF with absent pulmonary valve (ToF-APV). In 3 cases a transition from ToF to ToF-PA was observed. Karyotype was checked in 66 cases and was normal in 36 fetuses. Abnormalities included: T21–8 cases, T18–3, T13–3, Klinefelter-1,with 22q21 microdeletion, other – 3, 22q21 microdeletion – 12 fetuses. In all DiGeorge cases thymus was small or invisible. This syndrome was detected in 2 out of 4 fetuses with TOF-APV and in 2 out of 3 transitional cases.
Associated anomalies within cardiovascular system included: 4 AVSD (2 in T21 fetuses), 4 lSVC, 13 right aortic arches, 6 aberrant origins of RSA, 19 MAPCAs. We did not find significant correlation between additional cardiovascular lesions and abnormal karyotype. DA was absent in all cases of ToF-APV. Extracardiac malformations (ECM) were in 32 cases, IUGR – 28. Correlation between IUGR, karyotype and ECM was not significant.
Perinatal outcome was known in 93 cases (94%). There were 81 live births (41-VD, 37–CS), 6 stillbirths and 6 terminations (6.5% of all pregnancies, 20% of diagnosed before 24 week). 14 (17.9%) children were born pre-term. 15 (19.2%) had low body weight. Majority were born in good condition: Apgar score 8–10:56 (74%); 5–7: 14 (19%); had 1–4:5 (7%).
Conclusions: ToF was diagnosed after 24 weeks in 63% cases. ToF is an indication for detailed USG scan and karyotyping with 22q21 microdeleton exam, especially when thymus is absent. Right aortic arch, aberrant right subclavian artery or lSVC were not markers of abnormal karyotype. Termination rate was low, even in early recognized cases, and occurred more commonly in fetuses with genetic anomalies. Neonatal condition was good in 93% cases.

PW4-11
Maternal Serum Antiarrhythmic Drug Levels Do Not Predict Fetal Supraventricular Tachycardia Response Time
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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Background: Fetal tachycardia requires rapid diagnosis of fetal rhythm and timely initiation of antiarrhythmic medication. Fetal mortality and neurological morbidity has been unacceptably high when recommended treatment protocols have been followed. However not only maternal antiarrhythmic drug levels but also fetal response time may play an important role in unfavourable outcomes.
Methods: We reviewed all fetuses presenting with tachyarrhythmia to University Hospital of Wales Fetal Cardiology. Flecainide and digoxin combination was treatment of choice. Maternal antiarrhythmic drug levels and fetal response time to tachycardia have been evaluated.
Results: There were 51 patients and 37 fetuses were given flecainide and digoxin combination treatment. The sinus rhythm was established in mean of 4.31 ± 3.21 days (range, 1–14 days) in fetuses with supraventricular tachycardia (96%). The response time in atrial flutter was longer with a mean of 9.0 ± 6.95 days (range, 1–18 days). Hydrops resolved completely in all fetuses but it took as long as 2 weeks after normalization or reduction of fetal heart rate below 160 bpm. There was no correlation between maternal serum peak drug levels and fetal response time to tachycardia (digoxin $r = -0.17$ and flecainide $r = -0.06$).
Conclusion: The two drug combination treatment with flecainide and digoxin has been effective in improving fetal haemodynamics in most cases with fetal supraventricular tachycardia and atrial flutter but tachycardia response time had no significant correlation with maternal serum peak drug levels. Tachycardia response time is highly likely to play an important role in fetal outcomes therefore efforts should concentrate on rapid restoration of sinus rhythm or reducing fetal heart rate as close to tolerable levels as possible rather than following escalation protocols or applying wait and see policy.

PW4-12

Determining the best treatment for fetal SVT and Atrial Flutter (AFl): a comparison of two common drug treatment protocols

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Background: The best treatment for sustained fetal SVT with 1:1 AV relationship or sustained fetal AFl is not known.
Methods: 155 consecutive fetuses with supraventricular tachyarrhythmia presented 2000–2012. 127 had SVT with 1:1 conduction and 28 had AFl. 86/127 with SVT were treated: first-line maternal intravenous digoxin (n = 52, centre 2), or maternal oral flecainide (n = 34, centre 1). 25/28 of those AFl received drug treatment: digoxin ± sotalol/other drug (n = 16, centre 2), or digoxin ± flecainide (n = 9, centre 1). Treatment success was defined as conversion to sinus rhythm, or >15% rate reduction
Results: SVT subgroup analysis
Short ventriculo-atrial (VA) interval occurred in 69 and long VA in 17. Hydrops was present in 30/86 (35%). Digoxin was successful in 23/28 (82%) and flecainide in 26/27 (96%, p 0.19) of non-hydropic fetuses, compared to 8/21 (38%) and 6/7 (86%, p 0.07) with hydrops.
For short VA SVT, conversion to sinus rhythm and rate control was 31/44 (70%) and 0/44 for digoxin, and 23/25 (92%) and 1/25 (cumulative 96%, p 0.01) for flecainide.
For long VA SVT, conversion to sinus rhythm and rate control was 4/8 (50%) and 0/8 for digoxin, and 5/9 (55%) and 2/9 (cumulative 78%, p 0.3) for flecainide
IUD or NND occurred in 9/21 hydropic fetuses treated with digoxin compared to 0/9 (p 0.03) treated with flecainide.
AFl subgroup analysis
In non-hydropic fetuses, sinus rhythm occurred with digoxin monotherapy in 7/17 (41%), with digoxin+additional agent in a further 2/17 (cumulative 53%). In hydropic fetuses, sinus rhythm occurred with digoxin monotherapy in 3/8 (38%) and with digoxin+additional agent in a further 4/8 (cumulative 88%). Hydrops was present in 11% of fetuses with sustained AFI compared to 44% of those converting to sinus rhythm (p 0.18). IUD did not occur (0/8 hydropic fetuses, 95% CI 0–0.37).
Conclusions: Flecainide was more effective than digoxin in short VA SVT, especially when hydrops was present. No adverse fetal outcomes were attributed to flecainide.
For those in AFl, surprisingly, hydrops did not reduce the likelihood of conversion to sinus rhythm.

PW4-13

Cardiac output measurement during maximal exercise test in patients with tetralogy of fallot (TOF) demonstrates chronotropic incompetence as a major cause of limited exercise capacity

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Introduction: After repair of Tetralogy of Fallot a decreased exercise performance has been described. Cardiac output (CO) measurement during exercise testing is possible via a CO2-rebreathing technique. The aim of our study is to evaluate CO at rest and at maximal exercise in patients with TOF.
Methods: Chlidren with corrected TOF (6–16 yrs) were invited to participate in a prospective study including a maximal cycle exercise test with spirometry combined with measurement of CO via the Oxycon Pro (Jaeger) system. Our institutional protocol (continuous ramping at 4W/kg) was applied. CO was measured at start and 20 seconds after maximal exercise performance. Data were compared with children referred for innocent complaints (chest pain) who performed a normal exercise test during the investigation period.
Results: 34 patients (18 boys) with TOF were included and compared to 36 normal exercise tests (19 boys). Groups were matched for gender, age, weight and length; BSA was equal. The duration of the exercise test was significantly shorter in the TOF-group (9.4 ± 2.3 min vs 11.7 ± 3.3 min, $P < 0.05$). Maximal heart rate (HRmax) (177 ± 17 bpm vs 188 ± 10 bpm), maximal load (95 ± 36 W vs 118 ± 51 W) and V02 max/kg (38.9 ± 7.8 ml/kg/min vs 47.7 ± 10.2 ml/kg/min) was significantly lower in the TOF group (all $P < 0.05$). 57.6% of patients reached anaerobic treshold (AT), which is not different from the control group (55.6%). HR at AT was significantly lower in the TOF group (164 ± 18 bpm vs 175 ± 15 bpm, $P < 0.05$). VE/VCO2 slope was equal in both groups. At rest, investigation of cardiac output showed no difference in Cardiac Index (CI) and stroke volume (SV). At maximal exercise, CI was signficantly lower in TOF-patients (10.2 ± 1.6 vs 11.1 ± 1.7, $P < 0.05$) whereas SV stayed equal (61 ± 11 ml versus 66 ± 10 ml) in both groups.
Conclusion: Patients with TOF have significantly lower oxygen consumption, HRmax, duration and load of maximal exercise test. This is consistent with earlier published data. CO measurement via the CO2-rebreathing technique, demonstrates significant difference in CI at maximal exercise, SV however

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

remains equal. This finding, in association with significant lower HR at AT, is in support for chronotropic incompetence being a major factor of exercise intolerance in TOF patients.

PW4-14

Intellectual Function in Prematurely born School-aged Children with Congenital Heart Disease

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Objectives: Few studies investigated neuro-developmental outcome in very low birth weight (VLBW) infants with congenital heart disease (CHD). The purpose of this study was to define the intellectual function of prematurely born school-aged children with CHD.
Methods: We reviewed 456 patients who admitted in our neonatal intensive care unit because of VLBW infants from January 2000 to December 2006. The isolated patent duct arteriosus and patent foramen ovale were excluded from CHD. A standardized test of intelligence (Wechsler Intelligence Scale for Children, 3rd edition, Japanese version) performed at 6 years old was use to evaluate intellectual function. The data were compared between the CHD and no-CHD groups.
Results: The CHD was detected in 27 infants. The most common lesions were ventricular septal defect (VSD) ($n = 9$, [33.0%]) coarctation of aorta (CoA) ($n = 3$, [25.0%]). At 6 years old, 18 patients were survived in the CHD group and 371 in the no-CHD group, respectively. The mortality was higher in the CHD group than that in the no-CHD group (33.3% vs. 13.5%, $p < 0.05$). The CHD group consisted of 8 patients (VSD 3, VSD after surgery 1, Double outlet of right ventricle and pulmonary stenosis after surgery 1, CoA after repair 2, Congenital aortic regurgitation 1 patient), excluding 2 of cerebral palsy, 4 of lost follow-up and 4 patients due to other test. Non CHD group consisted of 238 patients, excluding 6 of cerebral palsy, 84 of lost follow-up, and 43 pts due to other test. Full Scale IQ in prematurely born school-aged children ($n = 246$) was slightly lower than published data in normal children. However, compared with non CHD-group, there was no significant difference in Full Scale IQ (86.1 ± 14.0), verbal IQ (90.4 ± 14.8) and performance IQ (85.1 ± 13.8) in the CHD group. Also, no difference for verbal comprehension, perceptual organization, freedom from distractibility and processing speed score were found between the CHD and no-CHD group.
Conclusions: CHD is associated with increased mortality in VLBW infants. When VLBI infants with CHD are survived, similar intelligence capacity to prematurely born school-aged children without CHD may be expected.

PW4-15

Depression in Congenital Heart Disease; a forgotten issue

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Introduction: Treatment of childhood congenital heart disease (CHD) may be extremely stressful, resulting in psychological sequelae that may be present years later during adult life.
Purpose of this study was to examine the presence of depressive symptoms in adolescents and adults with CHD and the association of functional status and disease severity with the presence of depression.
Methods: Sixty patients (mean age 28.89 ± 11.41 years, 53.3% men), with CHD were recruited for the study as outpatients (stable patients) of a territory center between July 2007 and December 2010. All patients were asked to complete scales screening depressive symptoms (BDI, Zung SDS). Functional status was assessed using the New York Heart Association (NYHA) classification.
Results: Twenty-nine patients (48%) of the population study had severe forms of CHD (repaired Tetralogy of Fallot, univentricular anatomy (Fontan circulation), repaired transposition of great arteries), twenty-four (40%) had Eisenmenger syndrome and seven (12%) had moderate severity defects (aortic valve disease, atrioventricular canal defects). Fifteen percent (15%) received scores indicative of moderate to severe depressive symptomatology (scores of 20 and above) according to BDI and sixteen (16%) according to Zung SDS. The vast majority was classified as NYHA II and III (86.7%). The effect of NYHA functional status on BDI and ZUNG scores was proved significant. Both BDI score and ZUNG score were positively associated with NYHA ($b = 0.311$, $p < 0.001$, CI95% 0.143–0.478 and $b = 7.054$, $p < 0.001$, CI95% 3.252–10.856 respectively). Cardiac defect severity did not emerge as significant predictor of depression ($p > 0.05$).
Conclusions: This study confirms that patients with CHD and limited functional status, even stable ones, are depressed and underlines the need for the development and evaluation of psychosocial interventions targeted specifically to this group. Altering the psychological burden of these patients may have a positive impact in their functional status and quality of life.

P-1

Flecainide failure and proarrhythmia in pediatric supraventricular tachycardia: effectiveness and safety of different treatments

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Background: In our institution flecainide is the first choice drug in the chronic prophylaxis of supraventricular tachycardia (SVT). Among class I-C drugs, Flecainide is favorable due to its dosability and pharmacokinetics. In this study, from a cohort of 154 patients we selected those non-responder to Flecainide, in order to investigate efficacy and safety of different antiarrhythmic drugs.
Methods and results: We analyzed 154 patients ($4,21 \pm 3,20$ years) with supraventricular reentrant tachycardia (with the exclusion of atrial flutter) treated with flecainide alone or flecainide and beta blocker at maximal dose (propranolol or atenolol). Beta blockers were added exclusively in case of recurrence of SVT despite therapeutic range of serum Flecainide levels. Among these 154 patients, 16 ($4,65 \pm 2,13$ years) showed therapeutic failure, defined by recurrences of arrhythmias (more frequently than once per month) and/or incessant tachycardia. The supraventricular arrhythmias in non-responders showed different mechanisms: 7 concealed accessory pathway atrioventricular reentrant tachycardias (cAVRT), 3 Wolff-Parkinson-White (WPW), 4 atrioventricular nodal reentrant tachycardia (AVNRT), 2 persistent junctional reciprocating tachycardia (PJRT). Pharmacological failure was eligible for alternative therapy with sotalol (8 patients) 7 mg/kg/die, diltiazem (5 patients) 4 mg/kg/die, sotalol and diltiazem together at a dose respectively of 5 mg/kg/die and 4 mg/kg/die (3 patients). The mean time of follow up was $1,90 \pm 1,47$ years (range 0,27–3,23). Therapeutic success was achieved in 7/8 (87,5%) patients with sotalol, 4/5 (80%) patients with diltiazem and all the 3 patients (100%) treated with sotalol together with diltiazem.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Totally, the alternative therapy based on Sotalol and/or Diltiazem showed a lower incidence of recurrence of SVT in a year (2/16 patients, that is 12,5%) with no incessant tachycardia (0%), whereas the standard treatment based on flecainide and beta-blockers registered greater rate of recurrence (6/16 that is 37,5%) and a larger onset of incessant tachycardia (3/16 patients, 18,7%).
Conclusions: In pediatric patients unsuccessfully treated with flecainide, different antiarrhythmic drugs were effective and safe. Despite high success rate of flecainide (138/154; 89,61%) it's mandatory to be aware about recurrences of SVT and proarrhythmic related risks. Sotalol and diltiazem may be considered alternative and effective antiarrhythmic drugs.

P-2

Syncope in pediatric age: psychopatological profile in children with neurocardiogenic syncope

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Introduction: To evaluate a large cohort of pediatric patients with syncope and to assess the psychopathological profile in patients with neurocardiogenic syncope (NCS), compared with healthy sample.
Methods: A total number of 242 patients with syncope or pre-syncope (median age 12,3 years) were evaluated with our two-step diagnostic protocol. The Head-Up Tilt Test (HUTT) and a psychological assessment were performed in patients with suspected NCS with the aim to reveal a possible correlation between syncopal events and psychopathological characteristics. Of the 126 patients underwent HUTT and evaluated by our psychologist, 92 completed the psychological tests and the results were compared with a normative group.
Results: Compared with normative group, patients with syncope reported worse ($p < .001$) scores to syndrome scales of anxiety problems, of the depressive withdrawal problems and of thought problems. Also two of three broad problems scales, the internalizing and the total problem scales had significantly higher scores compared with sample. During a medium-term follow-up, no syncope recurrences were observed; this was associated with somatic and thought problems improvement, along with marked improvement in family functioning.
Conclusions: Patients with NCS had 2.0-fold higher rate of clinically significant somatic complaints and thought problems, identifying as a sort of psychopathological profile in these patients. Our findings call for additional investigation on the possible pathophysiologic association between somatic complaints, thought problems and the reflex mechanism that produces syncope. Furthermore, future clinical studies are necessary to plan an interventional strategy and optimize management.

P-3

Successful Catheter Ablation of a Concealed Superio-paraseptal Accessory Pathway along the Tricuspid Valve in a Fontan patient with extra-cardiac Conduit after Creation of a Trans-Catheter Communication

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Introduction: An extracardiac PTFE Fontan conduit may prevent antegrade access to the pulmonary venous atrium. However, such an antegrade approach may be necessary for successful catheter ablation of a "left-sided" arrythmia substrate.
Objective: We describe the technique of creating a trans-catheter communication through an extracardiac Fontan conduit for catheter ablation of a "left-sided" arrhythmia.
Case Report: A 23-year-old male patient with double outlet right ventricle, mitral atresia, severe left ventricular hypoplasia, subpulmonary ventricular septal defect, valvular pulmonary stenosis and D-transposition of the great arteries after modified Fontan anastomosis with an extracardiac PTFE conduit presented with recurrent supraventricular tachycardias despite antiarrhythmic medication. Invasive electrophysiological testing showed a "left-sided" concealed accessory pathway that could not be reached by a retrograde approach from the aorta through the tricuspid valve. Based on our experience with creation of trans-catheter fenestrations in Fontan patients, we decided to cross the extra-cardiac conduit by Brockenbrough technique utilizing a trans-septal needle, followed by gradual balloon dilation of the conduit- atrial-wall, which allowed placement of a 8F long sheath within the pulmonary venous atrium. The accessory pathway could be uneventfully mapped and ablated at the superoparaseptal aspect of the tricuspid valve.
Conclusion: In Fontan-circulation with extra-cardiac-conduit catheter ablation of "left-sided" arrythmias is feasible by creation of a trans-catheter fenestration.

P-4

Course of the Defibrillation Threshold After ICD-Implantation Using the Extracardiac Technique: Midterm Follow-up

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Introduction: There is still no standard concept for ICD implantation in infants and smaller children. The previously described extracardiac technique offers an effective and safe concept of an ICD implantation in this special patient group. However, data of midterm follow-up are lacking, especially the course of the DFT facing further growth of the patients.
Patients and methods: An extracardiac ICD-system was implanted in 28 patients (mean age: 5.1 [0.2–11.5] years; mean body lenght: 107 [61–147] cm) as previously described. Under fluoroscopic guidance a defibrillator lead was tunneled subcutaneously ($n = 8$) or subpleurally ($n = 23$, including 3 patients with a former subcutaneous lead) along the course of the 6th rib until almost reaching the vertebral column and bipolar steroid-eluting sensing and pacing leads were sutured to the ventricle. The ICD device was implanted as "active can" in the right upper abdomen or inserted in a horizontal position between the diaphragm and the pericardium. Sensing, pacing, and defibrillation thresholds (DFT) as well as impedances were verified intraoperatively, 3 months later and every 12 months, respectively.
Results: In 30/31 ICD implantations using the extracardiac technique an intraoperative DFT < 15 J (median DFT 10 [5–15] J) between the extracardiac lead and device could be achieved. However, rise of the median DFT was noted from 10 J intraoperatively to 15 J after 1 year. There was a significant correlation between the DFT and body length but not between

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

DFT and body weight. After a mean follow-up of 2.5 years, revision was required in 5/7 patients with a subcutaenous defibrillation lead and in 5/23 with a subpleural lead. Reason for revision were lead problems (n = 3, all with subcutaneous leads), device infection (n = 1) and inadaequate ventricular sensing (n = 1), respectively. In the remaining 5 patients, increase of the DFT > 20 J was noted during regular DFT testing without change of the impedance of the shock electrode or any signs of dislocation or fracture of the defibrillation coil.
Conclusions: The extracardiac technique offers a safe and effective approach for ICD implantation in infants and small children. However, regular DFT testing is mandatory to recognize failure of the system.

P-5
Preparticipation Screening in Europe in Competitive Athlets
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Introduction: Sudden cardiac death (SCD) in young competitive athletes is a rare event. The main causes of SCD are cardiovascular diseases (55–85%) occurring in about 80% during exercise. According to the literature about 90% may be detected by preparticipation screening including ECG. At the moment preparticipation screening in Europe is not standardized and ECG is not included in the most parts of Europe. Although we did an accurate literature search we could not collect data of the actual practiced preparticipation screening guidelines in most European countries. The aim of this study was to do an online survey contacting pediatric cardiologists all over Europe to evaluate the actual standard of preparticipation screening.
Methods: An online survey including six questions was send to pediatric cardiologist and was then analyzed.
Results: Preparticipation screening in European countries is different. There is no standard what examinations are included in the preparticipation screening in each country and who is performing the examination. In most countries preparticipation screening is not obligatory or it depends on the national sports organizations if it is required to participate in competitive sport events.
Conclusion: There is no standard in preparticipation screening in Europe, although the Italian model- preparticipation screening including ECG in every competitive athlete- showed a dramatic reduction of SCD. According to the literature we strongly recommend standardized guidelines for preparticpation screening including an ECG.

P-6
Junctional Ectopic Tachycardia after Pediatric Cardiac Surgery: Single-Center Experience from Turkey
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Background and Objectives: Junctional ectopic tachycardia (JET) that significantly increase morbidity in postoperative period is serious complication that can be seen after surgery for congenital heart diseases. The aim of this study was to determine the incidence and risk factors for JET after pediatric cardiac surgery and to assess the effect of JET on patients' outcome.
Patients and Methods: From January 2010 to August 2012, 623 pediatric patients who underwent cardiac surgery were enrolled in this study. Data were collected retrospectively. Patients who have arrhythmias preoperatively, who cannot have available data and who were referred to another clinic postoperatively were excluded. JET was defined as a narrow complex tachycardia with atrioventricular (AV) dissociation and that ventricular rate is higher than atrial rate. AV dissociation was confirmed in most of the patients using atrial pacing wires.
Results: The mean age at procedure was 32 ± 50 months, the mean weight was 11.7 ± 12.9 kg. We identified 33 patients with JET (33/623, 5.2%). Compared to patients have no arrhythmias, patients who have developed JET were significantly young (14.6 ± 29.5 vs. 33.5 ± 51 months), have had lower body weight (7 ± 5.2 vs. 11.8 ± 13.1 kg), have a higher inotropic score (14.8 ± 11.4 vs. 8.7 ± 10.9). JET developed most frequently after atrioventricular septal defect (AVSD) repair (9/43, 20%) and tetralogy of Fallot repair (11/81, 13.5%) in our series. JET is not associated with electrolyte levels, surgery close to the AV node or Risk Adjustment for Congenital Heart Surgery (RACHS) score. Multivariate logistic regression analysis showed that the duration of cardiopulmonary bypass (CPB) is significant predictor of postoperative JET [odds ratio, 1.007 (confidence interval, 1.002–1.011): p = 0.02]. Furthermore the use of dopamine is another risk factor for JET [odds ratio, 8 (confidence interval, 1.01–64.5): p = 0.049)]. JET is associated with prolonged mechanic ventilation time, prolonged PCICU and hospital stay.
Conclusion: JET is most frequently seen after AVSD and tetralogy of Fallot repair. The highest risk factors for development of JET are the use of CPB, the duration of CPB and the use of dopamine in postoperative period.

P-7
Left Posterior Fascicular Ventricular Tachycardia Unresponsive To Verapamil
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Introduction: Ventricular tachycardia (VT) originates from right ventricular outflow tract or left ventricular septum in children with normal heart. VTs originating from the left ventricular septum are often re-enterant and responsive to verapamil. Left fascicular VT from an ectopic focus are less frequent. We present a patient with left posterior fascicular VT unresponsive to verapamil and treated with ablation.
Case: Fourteen-year-old male patient presented with a wide QRS tachycardia. His only symptom was palpitation, no history of syncope or signs of cardiac failure was present. Heart rate was 170 bpm, and blood pressure was 120/60 mmHg. Echocardiography revealed an anatomically normal heart, with mildly decreased systolic function (FS = 27%, EF = 55%). The case was unresponsive to adenosine or metoprolol, and amiodarone infusion was started. Heart rate decreased to 130/min, but sinus rhythm could not be obtained. Atrioventricular dissociation became visible with an atrial rate of 75 bpm and ventricular rate of 137 bpm. Right bundle branch block morphology and left axis deviation suggested left posterior fascicular VT and diltiazem was given. However, tachycardia did not resolve. Therefore; radiofrequency ablation was planned. Tachycardia mapping was performed by a quadripolar catheter in His position. First, right bundle crioablation was attempted considering bundle branch re-enterant VT because of verapamil resistance. Since the

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

tachycardia persisted, mapping at left ventricular septum was performed, purkinje cell action potential was detected at the left posterobasal area, close to the apex. Radiofrequency waves at 50 watt, 60°C were applied to this region for 4 minutes. Tachycardia was terminated, and could not be induced with single or double ventricular extrastimuli. The patient is being followed-up without any anti-arrhythmic medication.
Conclusion: Left posterior fascicular VT causes right bundle branch block morphology and left axis deviation on the surface ECG. Verapamil responsiveness is typical. This case was interesting for its verapamil unresponsiveness.

P-8
A family with Myotonic Dystrophy (DM1) associated with Sudden Death, Long QT and a Brugada-like ECG pattern in different affected relatives
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Myotonic dystrophy is a multisystem condition inherited in an autosomal dominant manner. In addition to myotonia and muscular dystrophy, affected individuals are also at risk of cardiac dysrhythmias and conduction defects. A 17 year old male died suddenly whilst riding a bicycle having been previously well. Post mortem examination was normal and cause of death was considered to be due to a primary underlying arrhythmia. His parents and 13 year old brother were referred for cardiac screening. His father's resting ECG showed evidence of R waves starting from V1. His echo showed some septal hypokinesia with a normal MRI. Following an ajmaline test, there was asymptomatic sustained VT. His mother had a normal echo, MRI and ajmaline test but had a corrected QT interval of 450 ms associated with hypoparathyroidism which resolved with treatment. Initially, his brother had a normal resting ECG and echo but subsequently was found to have a corrected QT interval of 500 ms. At the time of cardiac screening his mother and brother were well with no other medical history. However, his father was concurrently being assessed for dysphagia. On further examination, he had frontal balding, facial muscle weakness and myotonia. On direct questioning there was also a history of myotonia in the deceased. Gene testing identified a DM1 gene expansion from leucocyte DNA in the father and brother and in DNA from pathological blocks from the deceased. Long QT and Brugada gene testing (KCNQ1, KCNH2, KCNE1, KCNE2, SCN5A) in the father and brother was normal. This family highlights the variability of cardiac dysrhythmias associated with Myotonic Dystrophy and that gene testing for this condition should be considered in all patients being investigated for Long QT syndrome or with abnormal ajmaline tests.

P-9
Implantable Cardioverter-Defibrillator therapy in children: a single-centre 10 year experience
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Background: The use of implantable cardioverter defibrillator (ICD) therapy is increasing in paediatric patients. Whilst ICD therapy prevents sudden cardiac death (SCD) in children, this occurs at the expense of a high rate of complications. This study represents the largest single centre series of ICD implantation in children.
Methods and Results: 52 consecutive patients (31 male [60%]) underwent ICD implantation between November 2002 and March 2012. Median age at implantation was 14.7 years (range 2.9–18.4 years). Diagnosis was hypertrophic cardiomyopathy in 36 (69%), long QT syndrome in 6 (11%), arrhythmogenic right ventricular cardiomyopathy in 3 (6%) and repaired congenital heart disease in 3 (6%). One patient each (2%) had dilated cardiomyopathy, catecholaminergic polymorphic ventricular tachycardia, idiopathic ventricular fibrillation and coronary artery disease secondary to Kawasaki disease. 39 patients (75%) underwent ICD implantation for primary prophylaxis whereas 13 patients (25%) underwent ICD implantation for sustained ventricular arrhythmia or following cardiac arrest (secondary prophylaxis). Over a median follow-up of 3.2 years (range 0.2–7.8 years), 13 patients (25%) had at least one appropriate discharge; median time to first appropriate shock was 0.42 years (0.4–6.9 years). Appropriate shocks occurred more commonly in the secondary prevention group (46% vs. 18%; annual rate 44% vs. 4%; $p < 0.05$). Complications included inappropriate shocks in 7 patients (14%), infective endocarditis ($n = 3$, 6%) and psychological effects ($n = 11$, 22%). 1 patient died from incessant ventricular arrhythmia and 2 underwent cardiac transplantation.
Conclusion: ICD therapy is effective at reducing the rate of SCD in children. Appropriate shock rates are substantially higher in patients with previous cardiac arrest, but complication rates are high. This study highlights the need for psychological support and vigilance against infection in children undergoing ICD implantation.

P-10
ECG monitoring of treatment response in the rat pulmonary arterial hypertension model
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Introduction: Although, recent studies emphasize the relationship between experimental pulmonary artery hypertension (PAH) and arryhythmias, the issue is not so clear. The aim of this study is to evaluate the effects of bosentan, sildenafil, and combined treatment on the surface ECG in rats with monocrotaline (MCT) induced PAH.
Methods: Sixty three-month-old male Wistar rats were included in this study and randomized into five groups. All the rats were given monocrotaline (MCT) subcutaneously except the control group. After 4 weeks, following the development of PAH, bosentan sildenafil and combined therapy was given to the groups for 3 weeks. Another group that developed PAH did not receive any medication. ECHO and surface ECG studies were performed in all the rats at the baseline, after PAH development (4th week), and after treatment.
Results: Right ventricular pressure, calculated from the tricuspid regurgitation, was increased in all MCT groups. After the treatment, only in sildenafil group right ventricular pressure was significantly decreased ($p < 0.001$).

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

ECG Studies: P amplitudes increased after developing PAH comparing to the saline group ($p = 0.011$) and this effect continued after treatment ($p = 0.014$). There was no significant change related with other P and QRS parameters. Although, there was no significant change on T amplitude and QTc values after PAH, the values were significantly increased in all groups after treatment ($p < 0.001$).
In all treatment groups after the treatment period, T repolarization duration was longer comparing to saline group ($p < 0.001$); however, T depolarization duration was longer only in bosentan and combined therapy groups ($p = 0.002$). By means of T wave parameters, combined treatment group had the longest amplitude and duration whereas sildenafil group had the shortest.
Conclusions: It has been found out that the development of PAH in rats especially influenced T-wave parameters. The continuum of the influence for both treatment and non-treatment groups has supposed to increase based on the longer duration of PAH. That the most considerable T-wave changes occurred in combined group whereas the least in sildenafil group thought to be the effect of medication on ECG changes as well as PAH should also be taken into consideration.

P-11
Left atrial inexcitability in pediatric patients with congenital lupus induced complete atrioventricular block
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Introduction: Atrial standstill and atrioventricular (AV) conduction blocks have been described in systemic lupus erythematosus adult patients and older children. In newborns from women with anti-Ro/SSA antibodies, AV block and ventricular cardiomyopathy are well known immunologic complications; to our knowledge, no cases of atrial inexcitability or atrial standstill have been described in this setting.
Methods: We recently encountered four pediatric patients with maternal lupus induced complete AV block meeting the criteria for cardiac pacing, who demonstrated left atrial inexcitability and/or interatrial conduction block. Clinical and pacemaker data, electrocardiograms and echocardiograms were reviewed.
Results: Diagnosis of maternal lupus induced AV block was made prenatally ($n = 2$), at birth ($n = 1$) and at 34 months ($n = 1$). All 4 female patients underwent epicardial dual chamber pacemaker implantation (mean age 28.5 ± 25.4 months) using steroid eluting leads, through a left thoracotomy. In 3 patients, no appropriate left atrial appendage (LAA) or left atrial capture could be achieved despite high outputs. The atrial lead was thus fixed on the right atrial appendage (RAA), along with a left ventricular (LV) lead. LAA biopsy performed in one patient was unremarkable. One patient developed cardiomyopathy soon after pacing was initiated. Upgrade to a biventricular pacemaker system did not improve function. This patient is awaiting heart transplant.
The fourth patient underwent LAA and LV lead placement. Follow up demonstrated early LV dysfunction and increased delay between surface P wave and intracardiac atrial depolarisation raising suspicion for interatrial conduction delay. The pacemaker was upgraded to a biventricular system, the LAA lead moved to the RAA, resulting in LV function improvement.
Conclusion: In patients with congenital lupus induced complete AV block, pacemaker implantation can be challenging due to left atrial inexcitability and/or interatrial conduction delay, suggesting an associated atrial myopathy, yet to be characterized. Longer follow up will be needed to better understand the true significance of this finding in the setting of congenital lupus induced complete AV block. Further preoperative delineation should be done, possibly using echocardiography, to guide surgical approach.

P-12
10 Years Experience of Interventional Treatment of Tachyarrhythmias in Newborns and Infants
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Basics: To perform single centre experience of managing tachyarrythmias in infants 1 year of life.
Methods: From 2001 to 2012 entered 95 infants with tachyrrhythmias with av.age 5.7 months (range 1 day–12 m) and av. weight 6,5 kg. (min 2,5 kg). 27,4% ($n = 26$) were under 3 months of age, and 13,7% ($n = 13$) were newborns. In 50% of infants arrhythmias appeared during neonatal and in 13,7% in antenatal period. Arrhythmias were represented by the WPW syndrome in 59%, atrial flutter in 20%, ectopic atrial tachycardia in 16% and ventricular premature beats in 5% of cases. Combination of arrhythmia and CHD were detected in 32%. Most frequent CHDs were VSD, ASD, PDA and coarctation of the aorta. 78% of the total amount of infants were operated. The rest were discharged after selection of the efficient antiarrhythmic therapy. The majority of infants (62%) had concealed form of the WPW syndrome, 25% - manifesting and 13% - intermittent forms. WPW syndrome was clinically apparent by paroxysmal or incessant life-threatening tachycardia. Clinical signs of NYHA class 1 heart failure had 60% of infants, NYHA class 2–24%. Arrhythmogenic cardiopathy was detected in 22% of infants.
Results: All infants with tachycardia underwent electrophysiological examination. Ablation (RFA) performed in 40 infants. In some patients we made transesofageal electrostimulation, Sili operation, single-stage correction of CHD and arrhythmia. 64,5% of patients with WPW syndrome had left-sided accessory pathways, 27% right, 8,5% septal. The RFA was efficient in 97.5%. In 1 infant we repeated RFA because of recurrent tachycardia returned in a day after 1st intervention. There were no complications after RFA. Infants were discharged on the 3–5 day after RFA. By the moment of discharge infants presented regression of heart failure signs and as well as signs of arrhythmogenic cardiopathy. Control long-term examination showed no tachycardia attacks.
Conclusion: Tachyarrhythmias can present early in infants and in paroxysmal or incessant life-threatening character they cause decompensation of heart failure soon after birth. The leading cause of medication-refractory tachyarrhythmias in infants under 1 year is WPW syndrome. Interventional treatment of tachyarrhythmias in infants is efficient but should be performed by absolute indications.

P-13
Corrected QT interval in children with congenital deafness from high consanguinity population
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Background: Long QT syndrome is characterized by prolongation of the QT interval. Patients are prone to syncope and sudden death due to ventricular arrhythmias. The disease can be familial, with or without deafness. In some of the patients the prolongation

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

of corrected QT interval appears during rhythm acceleration. In this study we aim to check the corrected QT interval in congenitally deaf children from high consanguinity population.
Methods: We performed a rest and exercise electrocardiogram for 97 children with congenital deafness (group1) and 96 healthy children (group2). Gender distribution was 54:43 and 47:49 Males/females in group 1 and 2 respectively. Average age of the children was 11.5 ± 6.5 years. Family history of syncope, arrhythmia, deafness, convulsions and sudden death were included. The corrected QT interval (QTc) was calculated by Bazett's formula.
Results: In group 1, we found 3 children with QTc > 0.440. Range 0.440–0.457 mesc in rest ECG. During exercise QTc was > 0.440 mesc in another 8 children. Range 0.442–0.569 mesc. Positive family history of syncope (2) convulsion (2), sudden death (13), arrhythmia (3) and, deafness (5) was found in 25.7% of congenitally deaf children. In group 2 we found 2 children with QTc interval of > 0.440 in rest electrocardiogram and 3 during exercise. Range 0.444–0.495 mesc.
Conclusion: Long QT is more frequent in children with congenital deafness from high consanguinity population. Exercise test in these children may uncover those with normal QT interval at rest. High consanguinity population and family history of syncope, sudden cardiac death or convulsions warrants further investigation of the disease.

P-14
Resynchronisation pacing in patients with a congenital heart disease

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Introduction: Biventricular pacing (CRT) has become an option in young patients with a congenital heart disease. The mechanisms of ventricular dis-synchrony in these patients cannot be deduced from the adult patient. So alternative strategies had to be invented and are still under evaluation. We retrospectively analyse our patients in regards of safety and positive effects for the cardiac function.
Methods: We report on 21 patients with a mean age at time of device implantation of 16.3 years [1.9 to 33.2 years]. The observation period ranges from 3 months up to 5 years (mean 2.1 years) and summarizes up to 44 patient years. A transvenous approach was chosen in 8 patients (age from 14.8 up to 33.2, mean 25.7 years). Biventricular defibrillator (CRT-D) systems were used in 2 patients. Indications were set primarily according to the guidelines for conventional pacing systems. Decision making for a CRT system based on inter- or intra-ventricular contractile delays in echocardiography (echo) with a reduced overall myocardial function. Resynchronisation was programmed under echo guidance with optimisation of the AV-period at rest and an interventricular delay to best achieve a simultaneous contraction of the anterior and posterior ventricular wall.
Results: Five system revisions were performed because of lead failure (early $n = 1$, late $n = 3$) and one because a lead removal had become necessary causing tricuspid regurgitation. One patient with a Mustard palliation died 2.5 years after the implantation of a CRT-D system while waiting for heart transplantation. Significant improvement with restoration of normal cardiac function was found in two of the youngest patients. Significant improvement in echo could be followed in 10 patients and unchanged function in 7 patients.
Conclusions: CRT in congenital heart disease is a multivariate therapeutical approach in optimizing reduced cardiac function and has to follow the individual requirements of the patients. The underlying heart disease and arrhythmia, the age of the patients, the location for lead placement, the history of the myocardial dysfunction and the results of surgical procedures seem to play an important role in achieving improvement. Exchange of all experience available has to stimulate further refinement and optimisation of this therapeutical approach.

P-15
Percutaneous RF ablation procedures in children with Ebstein's anomalny

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Introduction: In Ebstein's anomaly (Ea) the septal (and often posterior) leaflets of the tricuspid valve are displaced into the right ventricle (RV). The anomaly is often associated with ventricular pre-excitation and Wolff-Parkinson-White (WPW) syndrome or conduction abnormalities (delayed intra-atrial conduction, right bundle branch block -RBBB). In children with Ea tachyarrhythmias are frequently encountered. In this study we present our children with Ea treated by percutaneous radiofrequency (RF) ablation because of tachycardia.
Methods: From 2004 to 2012 we performed 598 percutaneous ablation procedures in children, in 8 of them (7 boys, mean age $13.1{,}8 \pm 3{,}6$ years) Ebstein's anomaly was diagnosed. Seven children were on long lasting antiarrhythmic therapy because of tachycardia attacks. One 14 year old boy was admitted to the hospital with heart failure because of his first attack of tachycardia which was ventricular. RF ablation procedures were done under general anesthesia in 5 children, we used elecroanatomical system Carto XP.
Results: In the 4 children wide right accessory pathway (AP), in 1 intra-atrial reentry tachycardia (IART), in 1multifocal atrial tachycardia (MAT) and in 1 atrioventricular nodal reentry tachycardia (AVNRT) and in 1 RV outflow tract ectopic tachycardia were diagnosed. During the ablation procedure RF applications time ranged from 6,5 to 23,2 minutes (mean 14,5 minutes), it was much longer then mean application time in our laboratory (mean 5,9 minutes). We had no complications. Follow-up period ranged from 3 months to 4,3 years. Two boys required the second procedure: one with WPW and another one with AVNRT because of the arrhythmia returned two months after the first ablation. To this time 7 patients are arrhythmia free, the boy with MAT improved but he still is on pharmacotherapy, in one girl on last holter monitoring EKG we observed some WPW QRS compexes.
Conclusion: In children with Ebstein's anomaly the most frequently arrhythmia is related to right-sided accessory pathways but other types of supraventricular and ventricular arrhythmia are possible. Catheter ablation procedure may be effective in most arrhythmias related to Ebstein's anomaly however in children the procedure is difficult and the long time of RF energy application may be necessary.

P-16
The effect of Metoprolol treatment for ventricular arrhythmias in Andersen-Tawil Syndrom

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Andersen Tawil syndrome (ATS) is a channelophatic and sporadic disorder. Its present with cardiac arrhythmias, periodic paralysis and dysmorphic features. Although the burden of ventricular ectopy is often high in patients with ATS, degeneration into life-threatening arrhythmias is relatively low. Because the QT and QTc measured with U wave is prolonged it has been proposed as LQTS7. The standard treatment for LQTS is betablocker therapy, so it is often use in ATS patients. The mechanism leading to arrhythmia in ATS patients is much more complicated and it probably depend not only of impairment function of Ik1 channel.

The aim of the study was to estimate the effect of Metoprolol therapy in ATS children.

We compare the Holter track preformed without any medication and on Metoprolol therapy in our ATS patients.

The study group consist of 6 ATS children: 4 girls and 2 boys aged 9 yrs 2 months to 17 yrs 10 months (mean 14 yrs). The diagnosis of ATS was based on clinical symptoms (cardiac, neurological and facial dysmorphic) and confirmed by genetic test in 3 of them. Another 3 children are waiting for result of genetic test. Three children had cardiological symptoms of arrhythmia: syncope, palpitations, shortness of breath. An standard ECG track mean QTc measured in lead II was 430 ms (ranged 390–440 ms), mean QUc measured in V3 was 580 ms (ranged 500–620 ms). Measurements were done in all but one patient due to frequent ventricular arrhythmia. Table (M means Holter on Metoprolol) show comparison of frequency of ventricular arrhythmia with and without Metoprolol treatment. In 2 children Metoprolol had no influence on arrhythmia, in 2 slightly reduce the number of ventricular arrhythmia and in other 2 increase. Because the low number of patients statistical analysis was not done,

No pts	HR min	HR min M	HR max	HR max M	HR mean	HR mean M
1	57	41	132	132	85	68
2	48	46	161	132	91	74
3	57	47	189	154	91	83
4	48	48	161	125	84	73
5	58	49	147	175	85	84
6	48	43	163	147	90	77
	% of VeEx	% of VeEx M	No VT	No VT M	VeEx Day/night	VeEx Day/night M
1	5,9	6	6	65	d = n	d = n
2	1	33	2	58	d = n	d = n
3	33	56	253	3831	d = n	d = n
4	2	2	19	13	d < n	d < n
5	4	1	0	0	d < n	n > d
6	12.8	5	20	42	d > n	d > n

Conclusion: Metoprolol does not reduce ventricular arrhythmia in ATS patients.

P-17

Accurate, highly time resolved flow rate and volume quantification with multiview 3D Colour Doppler echo

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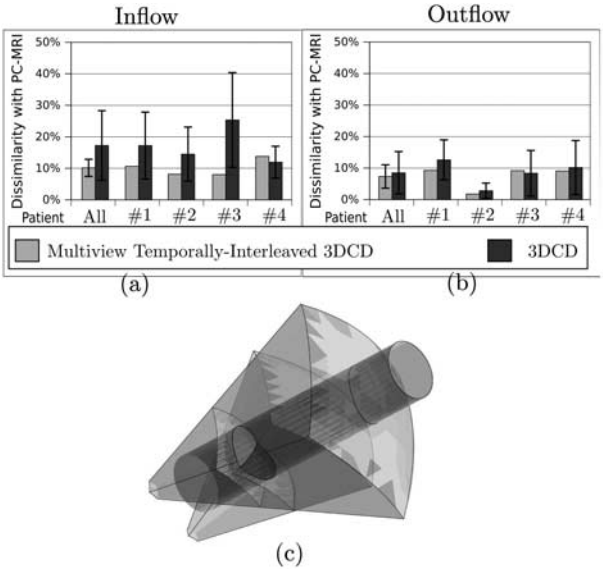
Introduction: Accurate flow quantification with echocardiography is an unmet clinical requirement especially in congenital heart disease. Pulsed-Wave Doppler (PWD) is widespread but has low accuracy and repeatability. 3D colour Doppler Images (3DCD) can be used to calculate angle-independent flow rate but lacks spatial and temporal resolution. The current gold-standard modality, phase-contrast Magnetic Resonance Imaging (PC-MRI), is expensive, requires general anaesthesia in infants. MRI has a long scan time for 3D imaging and is prone to motion artefacts. We propose an angle-independent, highly time resolved method for flow rate and volume quantification from multiview temporally interleaved 3DCD.



Methods: Echo data was acquired on 4 patients with Hypoplastic Left Heart Syndrome post total cavopulmonary connection, using a Philips iE33 (X3-1) 3D probe. Angle-independent flow rate was obtained by integrating velocity over composite spherical surfaces computed from multiple 3DCD images (Fig. c). The surfaces are made of spherical patches which belong to different 3D views, allowing increased coverage and approximation to user defined surfaces. Increased frame rate was achieved by interleaving trigger-delayed acquisitions. Cardiac inflow and outflow were measured. For the inflow, 2 apical views were acquired covering the tricuspid valve. For the outflow, coverage was achieved with 1 view. In all cases, 3 delays were used in the echo acquisition: 0, 20 and 40 ms. 3DCD images were acquired in 7 beats. The total duration of the echo exam was less than 10 min. Results were validated against flows from PC-MRI.

Results: Results are in the tables (a-b) which show the relative error in inflow and outflow volumes with respect to PC-MRI, using single view, non interleaved 3DCD and the proposed multiview, temporally interleaved 3DCD. The table shows that the proposed method outperforms 3DCD with good correlation in volume quantification and an average error in estimation of flow volume below 10%.

Conclusions: Standard approaches for flow quantification with Doppler lack accuracy due to angle dependency, temporal resolution and spatial coverage. We have proposed a method which compounds velocity values from multiple interleaved 3DCD images to calculate accurate, highly time resolved flow rate and flow volumes through user-defined surfaces.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

P-18

To fenestrate or not: a computational model as decision support for Fontan palliation

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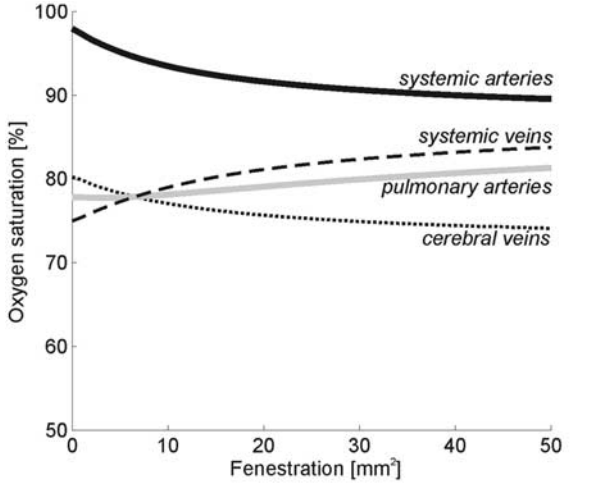
Objectives: Fenestration of the Fontan circuit was introduced to decrease mortality rates in patients undergoing completion of the total cavopulmonary connection (TCPC). Fenestration allows venous blood flow from the vena cava inferior directly into the atrium, preventing a rise in central venous pressure (CVP) and preserving cardiac output when pulmonary flow is restricted, but at the cost of a decrease of arterial oxygen saturation. It is likely that the hemodynamic effects of fenestration depend on patient-specific characteristics, such as the ability of the ventricle to cope with the acute decreased volume load. Quantitative insight in hemodynamic consequences of fenestration is limited. Accordingly, we use a computational model of the cardiovascular system to evaluate the effect of a fenestration on postoperative hemodynamic parameters, i.e., CVP, ventricular function, cardiac output, and oxygen transport in rest and during raise of cardiac output.



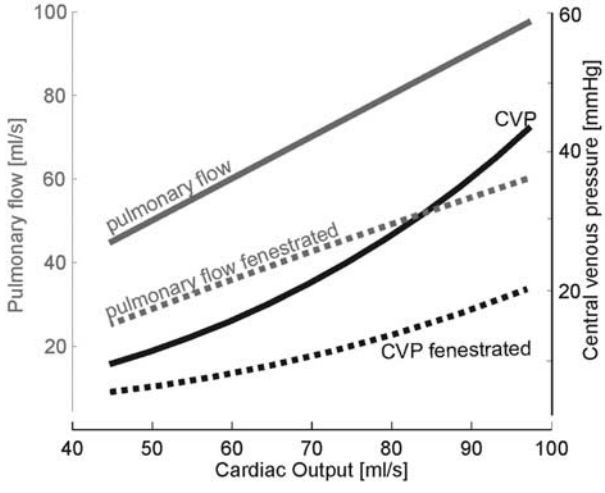


Fig. 1 Effect of fenestration size on oxygen saturation in a completed Fontan circulation during rest
Fig. 2 Effect of a fenestration of 20 mm^2 on the relation between CVP and CO (black) and pulmonary flow and CO (grey) in a completed Fontan circulation.

Methods: The multi-scale computer model used (CircAdapt) simulates beat-to-beat dynamics of the two cardiac cavities, the valves, and the systemic, cerebral and pulmonary circulation. The regulating effects of renal fluid retention and venoconstriction on mean arterial pressure have also been incorporated. The cerebral circulation includes arterial oxygen saturation induced auto-regulation of cerebral blood flow (CBF). Rest and increased cardiac output (CO), induced by increased stroke volume mimicking exercise, were simulated to evaluate the effect of fenestration in both situations.
Results: Fenestration of the TCPC decreased arterial oxygen saturation and thereby caused an increase of CBF due to cerebral autoregulation (Fig 1). The raise in CVP with increasing CO was less in a TCPC with than without a fenestration (Fig 2). The fenestration acts as a pop off valve for systemic venous return when increase of pulmonary flow is restricted. During exercise, fenestration preserves ventricular filling and prevents increase of CVP, but effective pulmonary flow (oxygen delivery) increases less than systemic cardiac output (Fig 2).
Conclusions: Fenestration of the TCPC is beneficial for patients with limited capacity to increase pulmonary flow or with high CVP in rest. It preserves ventricular filling and prevents increase of CVP during exercise. Ultimately, we aim to reveal preoperative patient-specific parameters that may guide the decision to fenestrate or not.

P-19

Changes of potassium channel gene expressions after human umbilical cord blood derived mesenchymal stem cells transfusion in pulmonary hypertension rat models

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Objectives: Pulmonary arterial hypertension (PAH) is difficult to treat and is characterized by increased pulmonary arterial pressure, right heart failure and death. PAH has been shown to be refractory to most of the conventional pharmacological therapies. Human umbilical cord blood-derived mesenchymal stem cells (hUCB-MSCs) are regarded as an alternative source of bone marrow-derived mesenchymal stem cells. hUCB-MSCs have recently been studied for evaluation of their potential as a source of cell therapy. The purposes of this study were to investigate changes of pulmonary pathology, haemodynamics and gene expressions of K+ channels, especially Kv1.7, Kir6.2.
Methods: The rats were grouped as follows: control group (C group), subcutaneous injection of saline; M group, subcutaneous injection of MCT (60 mg/kg); hUCB-MSCs transfusion (U group). hUCB-MSCs (3×106/mL/cm^2) were transfused by intraperitoneal injection 1 week after MCT injection.
Results: The mean right ventricular pressure (RVP) significantly decreased in the U group compared with the M group in weeks 2 and 4. RV weight and the ratio of RH/LH+septum significantly decreased in the U group compared to the M group. The number of muscular pulmonary arteriole significantly decreased in the U group compared with the M group in weeks 2 and 4. Medial wall thickness of the pulmonary arteriole significantly decreased in the U group compared to the M group in week 2. Gene expressions of Kv1.7 was significantly decreased in M group compared to C group and increased in U group compared to M group in week 4. And Gene expression of Kir6.2 is significantly increased in M group and significantly decreased in U group in week 4. Protein expression of

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Kv1.7 was significantly decreased in M group and increased in U group compared to U group.
Conclusions: After hUCB-MSCs transfusion, there was improvement of RVH, mean RV pressure and survival rate. Kv1.7 gene expressions were decreased in M group and increased in U group in week 4. Additional research on the dose and frequency of hUCB-MSCs infusion is needed to determine the optimal parameters for PAH treatment.

P-20
The increased serum levels of Interleukin-21 in Kawasaki disease
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Purpose: It has been reported that serum level of immunoglobulin E (IgE) is increased in patients with Kawasaki disease (KD) after acute phase. However the exact mechanism of increasing IgE is yet to be revealed. We investigated whether the interleukin-21 (IL-21) could be related with the high IgE in KD. Instead of IL-4, IL-21 was focused in this study because it has been reported that its level is increased in various autoimmune vasculitis.
Methods: From June 2008 to June 2010, 49 patients with KD admitted in Wonju Christian Hospital and 13 controls with high fever due to unknown infection who had no history of KD were included in this study. The sera from patients and controls were collected and checked in terms of immunoglobulin E (Chemiluminescent method, Siemens, Munich, Germany) and IL-21 (ELISA, eBioscience, San Diego, USA).
Results: The median age of patients with KD was 3 years of age (range: 0.4–10) and that of controls was 7 years of age (range: 1–12). The group of patients with KD was composed of 39 complete KD and 10 incomplete KD. Among patients with KD, 10 patients had coronary arterial dilatation (CAD) and 39 patients had no coronary complications. The median value of IL-21 in patients with KD was significantly increased as 466 pg/mL (range: 0–1544) while that value in controls was <62.5 pg/mL (range: 0–825 pg/mL) ($P < 0.01$). We could not find the significant correlation between the serum level of IgE and that of IL-21 in patients with KD (Spearman $R = 0.2$, $P = 0.08$) though 30% of patients with KD showed increased IgE more than 100 IU/mL. In addition, our data showed no significant difference between CAD group and non CAD group in terms of serum IL-21.
Conclusion: Our data showed firstly that IL-21 is increased in patients with KD. There was no significant correlation between high IgE and the level of IL-21.

P-21
Safety and early outcomes using a different induction therapy protocol in low and high-risk recipients in paediatric hearts transplantation
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Background and objectives: Advances in immunosuppressive therapy over the past two decades have contributed to improve the outcome of paediatric cardiac transplantation (HTx). Use of induction therapy, basiliximab (Ba) or antithymocyte globulin (ATG), has increased significantly in last years.
The aim of this study is to determine whether pre-operative administration of a different induction therapy is beneficial in preventing early heart allograft rejection.
Methods: The notes of 17 children undergoing HTx at our center from Dec-08 to Dec-12 were retrospectively reviewed. Of the 17 patients, 8 received Ba and 9 ATG as an induction therapy. From Dec-08 to Oct-10 Ba was the unique treatment used during induction. Since Oct-10 ATG was used in children with ventricular assistance pre-transplantation or/ and a number of cardiac surgeries greater than one (considered high-risk recipients). Maintenance immunosuppression included tacrolimus, mycophenolate mofetil, and steroids.
Results: The median age at transplant was 21 months (range 4 months to 16 years) and median follow-up was 18 months (range 4 to 49 months). Five were children who had prior palliation for congenital heart disease, 12 affected by cardiomyopathy and 1 re-HTx. Ventricular assistance pre-transplantation was used in 10 patients (58%). Of the 7 patients without ventricular assistance Ba induction was used in 6. Rejection during follow-up was diagnosed in 8 patients (Grade 2R cellular rejection in 5 and antibody-mediated rejection in 3). There were 2 deaths post-HTx (11 days, 1,5 months) due to antibody-mediated rejection, both with Ba induction. Freedom from Grade 2R or greater rejection or antibody-mediated rejection in the first 2 years was slightly greater in ATG group than in the Ba group. Post-transplant survival was 94%, 88% and 82% at 1 month, 12 and 24 months respectively.
Conclusions: An immunosuppression protocol consisting of ATG induction appears to achieve acceptable rejection rates during the first 2 years post-transplant in high-risks paediatric heart transplant recipients.

P-22
Effect of bosentan therapy on ventricular and atrial function in adults with congenital heart disease and Eisenmenger syndrome. A prospective, multi-center study using conventional and Speckle tracking echocardiography
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Introduction: The effect of bosentan on the ventricular and atrial performance in patients with Eisenmenger syndrome is unclear. We aimed to evaluate the midterm effect of bosentan in Eisenmenger patients on physical exercise, ventricular and atrial function, and pulmonary hemodynamics.
Methods: Forty adult patients before and after 24 weeks bosentan therapy underwent six minute walk test, two-dimensional

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

speckle tracking echocardiography, plasma NT-proBNP measurement and cardiac catheterization.
Results: After 24 weeks bosentan therapy an improvement was observed regarding the six minute walk distance from a median (quartile 1 - quartile 3) of 382.5 (312–430) to 450 (390–510) m ($p = 0.0001$), NT-proBNP from 527.5 (201–1691.25) to 369 (179–1246) pg/ml ($p = 0.021$), right ventricular mean longitudinal systolic strain from 18 (13–22) to 19 (14.5–25)% ($p = 0.004$), left ventricular mean longitudinal systolic strain from 16 (12–21) to 17 (16–22)% ($p = 0.001$), right atrial mean peak longitudinal strain from 26 (18–34) to 28 (22–34)% ($p = 0.01$) and right atrial mean peak contraction strain from 11 (8–16) to 13 (11–16)% ($p = 0.005$). The invasively obtained Qp:Qs and Rp:Rs did not significantly change under bosentan therapy.
Conclusions: In adult patients with Eisenmenger syndrome, bosentan therapy improves ventricular and atrial functions possibly by ameliorating cardiac remodeling, resulting in enhancement of physical exercise and reduction of the NT-proBNP level while the pulmonary vascular resistance does not change substantially.

P-23
Infections in Children with Left Ventricular Assist Device (LVAD): The Experience of Ege University Hospital

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Introduction and Objectives: Despite improvements in treatment of heart diseases with ventricular assist devices (VAD), infections are still associated with significant morbidity. Infections in patients with VAD described in three groups: left ventricular assist device (LVAD) related, non-LVAD related and sepsis. LVAD related infections includes driveline, pump pocket infections and defined as those that required treatment with antimicrobial therapy, when there is clinical evidence of infection such as pain, fever,drainage, and or leukocytosis. We report our three years of experiences about infections in 12 pediatric cases with LVAD.
Methods: In these retrospective study, we evaluated our twelve children with LVAD, between 2009 and 2012. Outcomes of this study includes postimplantation infection, types of infection and microbiologic profile of microorganisms.
Results and Conclusion: Eight of the patients had Berlin-Heart Excore LVAD and three of the patients had Heart Ware LVAD. All patients had the diagnosis of dilated cardiomyopathy. The mean age of the patients was 8,33 years (range, 17 months to 15 years). Five of the nine patients (55.5%) with Berlin Heart Excore LVAD had at least one episode of infection; None of the patients with HeartWare LVAD had infection. Fever and drainage from the exit site were the most common symptoms. The most common type of LVAD related infection site was the exit of the drive line. The most common type of non-LVAD infections were urinary and upper respiratory tract infections. Most of the driveline infections remained superficial and were managed with local wound care and antibiotics until transplantation. Staphylococcus aureus was the most commonly detected microorganism in LVAD related infections. Patients who had LVAD-related infection had a significantly prolonged hospital stay than the other patients with non-LVAD related infection. Only two of twelve patients died before transplantation, one of the patients died because of sepsis; coagulase negative Staphylococcus and Candida pelliculosa were detected in the blood cultures. Other one died due to intracranial hemorrhagia.
Although LVAD support is associated with improved survival and quality of life, infectious complications remain a major limitation. Novel evidence-based approaches to infection prevention remain critical.

P-24
Cardiomyopathy in a pediatric population: Key results from 10 years

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Introduction and objectives: Cardiomyopathy is a common cause of heart failure and the main cause of heart transplantation in children.
Objectives: (1) To elaborate a pediatric cardiomyopathy registry, (2) to describe the incidence, presentation and outcome of cardiomyopathy in a pediatric population, (3) to examine the medium and long-term course of functional status related to clinically important events.
Methods: Medical records of the patients under 15 years old affected by cardiomyopathy according to echocardiographic patterns, from January 2002 to December 2012, were reviewed. Patients with congenital heart defects and infants of diabetic mothers were excluded.
Results: 57 patients met inclusion criteria, 35 (61.4%) boys and 22 (38.5%) girls. Median age at diagnosis was 12 months. 30 (52.6%) patients were less than one year old and 3 patients were diagnosed during fetal life. The type of cardiomyopathy was: hypertrophy in 30 (52.6%), dilated in 18 (31.5%), left ventricular noncompaction in 7 (12.2%), restrictive in 1 and ventricular dysplasia in 1. In the hypertrophic cardiomyopathy group, 23 (76.6%) had a known cause: 7 with familial cardiomyopathy, 5 with malformation syndrome, 4 with neuromuscular disorder, 4 with arterial hypertension, 1 with mitochondrial disease, 1 with errors of metabolism, 1 with hyperinsulinism. In the dilated cardiomyopathy group, 12 (66.6%) had a known cause: 6 with familial cardiomyopathy, 2 with myocarditis, 1 with syndrome, 1 with anthracycline toxicity, 1 with arterial hypertension, 1 with mitochondrial disease. Mean follow up time was 3.7 years. 25 (43.8%) were admitted due to heart failure decompensation, 7 (12.2%) had an arrhythmogenic event and 2 (3.5%) had an aborted sudden death event. Surgical intervention was undertaken in 4 (7%), heart transplantation in 11 (19.3%), and implanted defibrillation deviced in 5 (8.8%). 6 (10%) patients dead, all of them were less than 1 year old.
Conclusions: More than 50% of children affected by cardiomyopathy have a known cause. The most frequent type of presentation is hypertrophic cardiomyopathy. Cardiac decompensation occurred in 44% of children affected by cardiomyopathy and heart transplant was undertaken in 19% of them. Younger age at presentation are associated with poor outcomes.

P-25
Genetic Origin Cardiomyopathies in a Pediatric Cohort

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

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Introduction: The continuously increasing knowledge on the genetic basis of myocardial disease allows understanding more and more the behavior of these diseases in individuals and families. It is our believe that promoting the knowledge of this basis in the pediatric population, where the genetic cause is not so established, is important to understand the disease from its origin, so that is our objective.
Methods: Prospective observational cohort study from January 2010 to December 2012 of children diagnosed with a cardiomyopathy of unknown origin: Hypertrophic Cardiomyopathy (HCM), Dilated Cardiomyopathy (DCM) or Left Ventricular Non-Compaction (LVNC). Demographics, family tree and genetic test for sarcomere protein genes (MYH7, MYBPC3, TNNI3, TNNT2, TPM1, MYL2, MYL3, ACTC, TNNC1 and TAZ) were performed.
Results: 56 patients (50% male) were included, 36 (64,3%) under 9 years of age. 24 patients (42,9%) showed mutations related to cardiomyopathies (1 had a double mutation). Mutation rate by age group: 33,3% (3/9) under 1 year of age, 36,8% (7/19) from 1–5 years and 50% (4/8, 4/8, 6/12) in the other age groups (5–9, 9–13 and >13 years).

	Num patients	Gender	Age (years) Mean(range)	Positive results
DCM	20	9M/11F	2,8(0-14,4)	5/20(25%)
HCM	16	11M/5F	10,1(2,3-17,5	10/16(62,5%)
LVNC	20	8M/12F	8,6(1,6-18,1)	9/20(45/%)
Total	56	28M/28F	7,0(0-18,1)	24/56(42,9%)

Mutations were found in 8 of the 9 genes studied: 11 (44%) in MYH7 (3HCM, 3DCM, 5LVNC) and 6 (24%) in MyBPC3 (5HCM, 1LVNC). The other 8 in TNNT, ACTC, MyL3, TPM1, TNNI3, TAZ. 12 (48%) were novel mutations, not previously described in literature (3HCM, 4DCM, 5LVNC). From the 21 families studied with positive results, 6 were de novo mutations, 13 inherited mutations and 2 have an incomplete study. During the study period 4 patients died, 2 had a heart transplant and 3 had an ICD implantation.
Conclusions: Mutations in sarcomere proteins are a common cause of cardiomyopathy in children, even in the youngest groups and more frequently in those with HCM and LVNC. Mutations must be searched not only in patients with family history as denovo mutations are not negligible. Further efforts are required to complete the understanding of genotype-phenotype relationships in this field.

P-26
Arterial stiffness determination by oscillometric method in children treated with anthracyclines for malignant disease

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Introduction: Anthracyclines are associated with cardiovascular toxicity and are the major cause of cardiovascular events in the group of survivors of childhood malignancy. Arterial stiffness is an independent predictor of cardiovascular disease. The aim of the study was to determine whether anthracyclines used for the treatment of malignant disease in childhood could increase arterial stiffness measured as the aortic pulse wave velocity (PWVao) and aortic augmentation index (AIxao). In addition, the aim was to identify the clinical parameters correlating with PWVao and the cutoff PWVao value discriminating healthy and diseased subjects.
Methods: A total of 119 children and adolescents aged 7–20 years were examined, 69 of them (mean age 13.69 ± 4.45 years) having completed anthracycline therapy for malignant disease according to various protocols at least a year before. Study patients were free from clinical and laboratory signs of malignant or cardiac disease. Control group included 50 healthy children, mean age 12.68 ± 3.22 years. Arterial stiffness was determined by measuring PWVao and AIxao using oscillometric method on an Arteriograph TensioMed device.
Results: PWVao was significantly higher (6.25 ± 1.31 m/s vs. 5.64 ± 0.66 m/s; $p < 0.001$) and AIxao was higher ($8.7 \pm 9.69\%$ vs. $5.64 \pm 5.15\%$; $P = 0.044$) in subjects with a history of anthracycline treatment as compared with control group. Univariate analysis yielded positive correlation of PWVao with age, body weight, body height, blood pressure, heart rate, mean arterial pressure and central arterial pressure in the group of patients previously treated with anthracyclines. There was no correlation of PWVao with anthracycline dosage and time elapsed from treatment completion. Multivariate regression analysis indicated body height, heart rate and physical activity to be the main PWVao predictors. The cutoff value of PWVao was 6.25 m/s (95%CI 0.591–0.765; $P < 0.0002$; sensitivity 44.9%; specificity 90%).
Conclusions: PWVao and AIxao are significantly higher in patients treated a year or more before with anthracyclines as compared to healthy children. The effect of anthracyclines on late mortality in individuals treated for malignant disease in childhood may not be exclusively due to their cardiotoxicity, but also to the increased arterial stiffness.

P-27
Endocardial fibroelastosis in the current era: associated congenital heart disease and outcome

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Introduction: Endocardial fibroelastosis (EFE) occurs primarily in infants during the first year of life, and is often associated with congenital heart disease (CHD), especially stenosis within the left ventricular outflow tract (LVOT) and hypoplastic left ventricle syndrome (HLHS). Outcome of infants with EFE in the current era has not been well delineated.
Methods: The echocardiography database from 1997 until December 2012 was searched for all children and infants (pt) with echocardiographic signs of EFE (dense echoes along the endocardial surface of the left (and sometimes right) ventricle (LV). Associated CHD, echocardiographic findings, age at diagnosis and survival were analyzed.
Results: There were 35 children (27 males) with EFE. HLHS was present in 18 (51%), bicuspid aortic valve in 14 (40%), and/or other

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

CHD in 22 pt (63%). Age at last echocardiographic examination was 0.9 ± 2 years. At least moderate mitral mitral regurgitation was present in 9 pt (26%). A dysplastic mitral valve was seen in 15 pt (43%). Decreased LV function was present in 57%.
Survival was severely diminished, 5 year survival was 64% but could not predicted by the presence of hypoplastic left heart syndrome, the ejection fraction, gender, age or associated congenital heart disease (p = NS). Cardiac interventions were necessary in 15 pt including hybrid operations, balloon valvuloplasty, Ross procedure.
Conclusions: The outcome of cardiac disease associated with endocardial fibroelastosis in the current era is still severely limited. Death can not be predicted by the underlying etiology of EFE.
Detection of EFE by echocardiography identifies a high risk population.

P-28
Assessment of Fetal Cardiac Function in Fetuses of Pre-eclamptic Mothers
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Introduction: The purpose of the present study was to investigate the detailed fetal cardiac function in mild pre-eclamptic pregnancies.
Methods: Sixty-five fetuses of mild pre-eclamptic mothers and fifty-five fetuses of healthy mothers between 26 and 40 weeks of gestation were included in this study. Fetuses with intrauterine growth retardation were not included in the study. Detailed fetal cardiac function were evaluated by M-mode, pulsed and tissue Doppler.
Results: Both groups were similar in terms of maternal age, gravidity, parity and gestational age ($p > 0.05$). Pulsed wave Doppler derivated ratio of E/A in mitral and tricuspid valves were similar in both group. In addition, deceleration time of early mitral inflow was prolonged in fetuses of pre-eclamptic mothers (FPEM) ($p < 0.001$). Isovolumic relaxation time and right and left myocardial performance index (MPI) were higher in FPEM ($p < 0.001$, $p = 0.02$, $p = 0.03$, respectively). Ea, Aa, ratio of Ea/Aa in interventricular septum, left ventricle lateral wall and right ventricle free wall were found to be signifcantly decreased in FPEM. The ratio of E/Ea in FPEM was higher than in control group ($p = 0.04$). Increased ductus venosus pulsatility index (PI) and decreased middle cerebral arter (MCA) PI were found in FPEM ($p < 0.001$).
Conclusion: We detected subclinic diastolic dysfunction and incresead right and left MPI in FPEM. In addition, we found decreased MCA-PI. We suggest that cause of all changes are the increase in fetal cardiac afterload.

P-29
Agenesis of ductus venosus: whats the real clinical relevance?
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Backgroud: Anomalies of the Ductus Venosus (ADV) are highly associated with poor fetal outcome (aneuploidy, structural malformations and fetal hydrops). Prognosis is variable and depends on such anomalies and the type of drainage (to portal sinus:intrahepatic or extrahepatic).
Objective: Describe real clinical consequences of this anomaly in two tertiary referral centers.
Methods: Prospective study (January 2005 to June 2012) from two tertiary centers where a complete first trimester scan (DV Doppler assessment) has been performed. All cases underwent serial fetal echocardiography and Necropsy or postnatal follow-up.
Results: we report 51 cases of ADV (3 twin pregnancies), 42% of them were diagnosed in first trim scan. 33 (65%) had intrahepatic (IH) umbilical venous drainage and 18 (25%) had extrahepatic (EH) drainage, either directly into right atrium (5), Inferior Vena Cava (7) or the Iliac Vein (2). We registered development of severe hydrops (IH 6%, EH 22% (P0.05)), associated anomalies (IH 24%, EH 39%) and increased NT (IH 24%, EH 16%, among the IH we had 1 CATCH and 1 Noonan Sd. The overall survival was 69% (IH 85%, EH 39% (P0.01)) but Isolated cases of ADV had 97% survival. We had 2 fetal demise and 12% underwent TOP (IH 6%, EH 22%). Survivors are healthy children (1 Aortic Coartation and 1 Ventricular septal defect operated on, 1 left isomerism without CHD and 1 renal policystosis). 2 hydropic newborn had persistent porto-systemic shunt.
Conclusions: Fetuses with IH umbilical venous drainage have better prognosis than EH drainage. The abscence of associated malformations, hydrops or increased NT has excellent prongnosis.

P-30
Antenatal diagnostic spectrum and early postnatal outcome of conjoined twins
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Introduction: Conjoining is a rare event with an incidence of 1:200 000 live births. The organ sharing is the limiting factor for survival and surgical approach. The aims of this study were to evaluate the foetal echocardiographic findings and early postnatal outcome of conjoined twins.
Methods: Retrospective review of the fetal and newborn database and case notes of 11 sets of conjoined twins seen between January 2001 and December 2007 at Hospital das Clínicas-UFMG and diagnosed prenatally by endovaginal and/or transabdominal echocardiography. The confirmation of diagnosis after birth, besides echocardiography, included cardiac catheterization, magnetic resonance and/or surgical or post-mortem findings.
Results: The mean of maternal age was 24,6 ± 5,9y and no maternal complications related to delivery were seen. The gestational age at diagnosis ranged from 12 to 31w (mean = 21, 2 ± X6, 8w; median = 25,0w) with a female predominance of 2,1:1,0. Antenatal and postnatal evaluation of the diagnostic aspects and the possibility of surgical approach were concordant. There were 54,5% thoracopagus or thoraco-omphalopagus, 18,2% parapagus and 9,1% each of omphalopagus, ischiopagus and cephalo-thoracopagus. None with two hearts was classified as thoracopagus. The thorax was involved in the fusion in 9 (81,9%) cases, of whom two showed two separate hearts and common pericardium. In the remaining 7, the findings involved fused hearts with fused atria, including common atrial chambers, common or straddling atrioventricular valves and complex ventricular structure of three or four ventricles with communication between them by

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

VSD. Anomalies in venous connections were frequent. Besides one judicial termination at the 14th week, four deaths occurred during the prenatal period. Five newborns died, two after surgery.
Conclusions: Conjoined twins presented a dismal prognosis. Thoracic-level fusion was associated with high frequency of fused hearts and complex structural heart disease. The endovaginal echo constituted an important tool for early diagnosis and counseling.

P-31
Atrioventricular block (AVB): antenatal diagnosis and late postnatal follow-up
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Objectives: To investigate intrauterine diagnostic spectrum, late postnatal evolution of AVB and risk factors associated to poor outcome.
Methods: From 12/1994 to 12/2010, 33 consecutive fetuses with AVB (age: 18–38w; mean: 31, 3 ± 3, 9w) were identified by ECHO and followed-up after birth (1,1–16,0y; mean: 7,9 ± 3,4y). Variables selected for analysis: maternal disease, gestational age (GA) at delivery, degree of hydrops, atrial and ventricular rates, structural cardiac lesions, cardiomyophaty.
Results: The maternal mean age was 28,4 ± 4,4y and 48,5% had a connective tissue disease (n = 33). The GA at delivery ranged from 28–38 (median: 37w) and the atrial and ventricular rates from 96–162 and 33–91 beats/min, respectively. Significant hydrops occurred in 53,3% and death, including 2 newborns with pacemaker in 11/33 (33,8%) of which 45,4% during the prenatal period. GA < 37w was found in 67,8% of stillborns and in 83,3% of neonatal death. Two groups were analyzed: G1 (n = 24): isolated AVB; G2 (n = 9): AVB plus significant structural heart disease (left atrial isomerism and congenitally correct transposition: 77,8%). Sustained bradycardia was seen in both groups but intermittent bradycardia (3 cases) only in G1. Complete AVB was diagnosed in all fetuses of G2 and in 16 of G1. Of the remaining 8 cases, all with second-degree block, 3 progressed to complete AVB, 3 to sinus rhythm and 2 remained stable. Death occurred in 6/9 in G2 and in 5/24 in G1, of whom 2 with second-degree block. Fourteen pacemakers (8 at first day) were implanted, mostly in G1 (86,7%). From the 22 survivors, 18 are asymptomatic (8 with pacemaker) and 4 with pacemaker and NYHA II or III (1 resynchronization therapy).The risk factors associated to death included hydrops III and IV (p: 0.004), significant structural heart disease (p: 0.010), ventricular rate < 55 beats/min (p: 0.001), and GA < 37w (p: 0.003). Dilated cardiomyopathy was associated with HR < 40beats/min (prenatal period) and with long term pacing (postnatal period).
Conclusions: Considering the high frequency of intervention and death, the AVB is a severe fetal arrhythmia, with early and late important repercussions, mainly in the group with associated congenital heart disease, severe bradycardia and early presentation. The fetal ECHO is very useful for early detection which contributes to optimize the pre and postnatal management.

P-32
Prediction of neonatal surgery or catheter based therapy in fetuses with congenital heart defects: Fetal Echocardiographic Score
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Introduction: There is improved survival and reduced morbidity in certain prenatally diagnosed congenital heart defects (CHD) compared to those with a postnatal diagnosis. On the other hand unnecessary referral brings about needless costs and anguish. The aim of this presentation was to develop a model to predict the probability of neonatal cardiac surgery or catheter based therapy in fetuses with CHD.
Methods: We developed and internally validated a model in a retrospective study from two tertiary centers in women whose fetuses were diagnosed as CHD. Retrospective analysis of ultrasounds and clinical records were performed. The outcome of interest was neonatal cardiac surgery or catheter based therapy. Clinical and ultrasound variables were included in a stepwise backward elimination regression model to predict the outcome. We assessed performance using the area under the curve (AUC) of the receiver operating characteristic (ROC). Standard bootstrapping techniques were used to assess potential overfitting.
Results: Between September 1998 and March 2012; 242 patients with prenatal diagnosis of CHD were included, of whom 142 (58.4%) required cardiac surgery or catheter based therapy. Visualization of only one atrioventricular valve, asymmetry of the great arteries and asymmetry of the ventricles were the strongest independent predictors in multivariable analyses. The model predicted the adverse outcomes within 30 days of life (AUC ROC 0·94, 95% CI 0·92–0·97). There was no significant overfitting. The model allowed the stratification of the sample into clinically relevant categories. The cutoff point of 0.30 estimated probability determined a positive predictive value of 85.8% and a negative predictive value of 97.5%.
Conclusions: In patients with CHD neonatal surgery or catheter based therapy can be predicted from ultrasonographic fetal heart evaluation by a prediction model and risk score, these may help in decision making regarding prenatal management and referral for birth place.

P-33
Prenatal diagnosis of coarctation of the aorta
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Coarctation of the aorta (Coa) is frequently not detected at obstetric scanning, and when suspected prenatally presents a significant rate of false positive cases (right heart chambers enlargement). We report the experience in its prenatal diagnosis in 4 tertiary health care centers in Argentina, Brasil and Cuba.
Methods: We reviewed fetal echocardiogram records and clinical records of 86 patients with prenatal suspition of Coa or Interruption of the aortic arch (IAA) in a ten-year period, fron December 2002 to December 2012. Cases with prenatal suspition of Coa or IAA and postnatal follow up of a minimun of 1 year were included. Patients with associated complex congenital heart and those lost to follow up were excluded.
Results: From 51 selected cases, 32 presented Coa, 4 had IAA, and 15 had mild hypoplastic or normal aortic archs. In the latter group 4 cases had mildly restrictive ductus arteriosus and in one case a restrictive foramen ovale. In the group with Coa, 19 cases had a ventricular septal defect and 16 had bicuspid aortic valve. Five patients developed Coa at or after 15 days of life.
Conclusion: Prenatal diagnosis of Coa or IAA poses many difficulties, false negatives being the main problem, restriction

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

of the ductus arteriosus or of the foramen oval might explain some of the false positive cases.

P-34

Vascular Rings: prenatal diagnosis and implication on postnatal treatment

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Objectives: to describe prenatal diagnosis and management of isolated vascular rings (VRs) at a single tertiary care institution.
Background: VRs refer to a group of congenital anomalies that interferes with and may compress the esophagus and trachea, leading to respiratory distress shortly after birth or to tracheoesophageal compression later in life. Prompt diagnosis and treatment of these congenital abnormalities can be lifesaving.
Methods: the department registry was retrospectively searched for all patients born between January 2002 and December 2012 with a prenatal diagnosis of congenital isolated VR. The variables assessed were indications for referral, gestational age at diagnosis, and postnatal outcome. Karyotiping and prenatal testing for 22q11 microdeletions were offered to all parents. Magnetic resonance imaging or computed tomography was performed immediately in symptomatic patients or electively in the first month of life in those with complete vascular rings, for excluding esophagus or tracheal compression.
Results: 53 fetuses referred to our centre for increased risk or suspicion of congenital heart disease had diagnosis of isolated VR at a mean gestational age of 24 weeks. The most common type of VR was left aortic arch (LAA) with aberrant right subclavian artery (ARSA) seen in 31 patients of whom 2 showed trisomy 21 at karyotiping and parents opted for termination of pregnancy (TOP). The second most common type was right aortic arch (RAA) with left aberrant subclavian artery (ARSA) arising from the Kommerell's diverticulum, seen in 15 patients. In 7 of them a circumflex retroesophageal aortic arch was found. Mild stenosis of ALSA was suspected in one newborn patient but he still didn't need of any treatment. Seven fetuses had double aortic arch with RAA larger than LAA. One of them had associated a 22q11 deletion and parents chose TOP. Three patients with DAA and 2 with RAA and ALSA developed symptoms from tracheal compression and immediately underwent successful surgical correction. In one of them tracheoplasty was performed as well.
Conclusions: Prenatal diagnosis of these vascular anomalies may be useful for improving postnatal management of affected children, who should receive a prompt surgical correction as soon as symptoms appear, avoiding tracheomalacia.

P-35

Impact of antenatal diagnosis on outcomes of neonates with Hypoplastic Left Heart Syndrome

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The objective of this study was to assess the influence of antenatal diagnosis of HLHS on postnatal outcomes.
Material and methods: This is a retrospective review of French single-centre records of neonates with diagnosis of HLHS. Clinical data, therapeutic management, outcomes were assessed. Comparison were made between groups with and without antenatal diagnosis, and between periods before and after 2005 (date of initiation of Norwood Program).
Results: Among 95 neonates (56 males = 59%) with HLHS, 44 (46%) were diagnosed prenatally (PreND group) and 51 (53%) after birth (PostND group). Birth weight was 3 ± 0.5 kg, Age at diagnosis in PostND group was 4 ± 6 days. All patients were free from symptom in the PreND group, while PostND group cases presented with heart failure (42%), cardiogenic shock (24%), cyanosis (12%), heart murmur (8%) or associated symptoms (14%). Age at Norwood procedure was 6 ± 2 days in PreND and 9 ± 4 days in PostND ($p = 0.01$). Time from diagnosis to surgery did not differ between groups (mean 6 days). Survival was similar between groups. Because of less termination of pregnancy, 71% (22 of 31 cases) of neonates with HLHS were diagnosed prenatally since 2005, compared with only 34% (22 of 64) before 2005 ($p < 0.01$).
Conclusion: Antenatal diagnosis may prevent acute postnatal cardiac failure in neonates with HLHS and allow early Norwood procedure, but it does not impact on long-term postoperative outcome.

P-36

Prenatal cardiac tumors: spectrum and outcome

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Introduction: all foetuses in the Czech Republic with suspected heart lesions are referred to a specialised centre for final diagnosis. The aim of the study was to establish aetiology, course and outcome of foetuses and children with prenatally diagnosed cardiac tumors.
Patients and methods: retrospective study of all prenatally diagnosed tumors in the Czech Republic between 1994 and 2011.
Results: 53 foetuses were identified to have single or multiple cardiac tumors: 45 (85%) rhabdomyomas, 3 (5.5%) teratomas, 3 (5.5%) fibromas, 1 (2%) hemangioma and 1 (2%) hamartoma. In 8 cases, parents elected to terminate the pregnancy: 4 rhabdomyomas due to MRI diagnosed tuberous sclerosis, 2 tumors almost completely obturating the cavity of left (fibroma) or right (rhabdomyoma) ventricle leading to univentricular haemodynamics, another 2 (fibroma, hamartoma) associated with severe extracardiac lesions. From 40 continuing rhabdomyomas, 32 (80%) were multiple, 6 (15%) had inflow/outflow obstruction, arrhythmias in 4 (10%), 1 died in utero (hydrops), Five out of 39 born alive were operated at the age of 10 ± 4 weeks, 2 of them died. Tuberous sclerosis was detected in 19/20 (95%). From 2 continuing pregnancies with teratomas, 1 died in utero, 1 with massive pericardial effusion and progressive tumorous enlargement was succesfully drained prenatally and operated in early neonatal period. 1 hemangioma was successfully extirpated in the neonatal period.
Large variability in size of tumors was found ranging from 3 to 55 mm (median 6). Rhabdomyomas (median 14 mm) were smaller than teratomas (42 mm, p 0.03) and fibromas (53 mm, p 0.002). Teratomas rapidly enlarged, 9/15 rhabdomyomas grew since the detection (27 ± 6.5 week of gestation) to birth, size of rhabdomyomas in all survived children postnatally regressed.
3 women had again foetuses with rhabdomyomas in their subsequent pregnancies.
Conclusion: Rhabdomyomas are most commonly prenatally detected cardiac tumors. They show prenatal growth in 56% but are usually not significant haemodynamically and do not

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

require immediate postnatal surgery. For the remaining rare tumors both prenatal and postnatal treatment may be live-saving in case of hemodynamic derangement.
"Supported by the project (Ministry of Health, Czech Republic) for conceptual development of research organization 00064203 (University Hospital Motol, Prague, Czech Republic)"

P-37

Electrophysiologic features of fetal ventricular aneurysms and diverticula

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Background: Congenital ventricular wall defects are very rare and include congenital ventricular aneurysms and diverticula.
Patients and Methods: We report a series of five fetuses: three with congenital ventricular aneurysms (CVAs) and two with congenital ventricular diverticula (CVDs) referred due to fetal arrhythmia.
In addition to routine fetal echocardiography, fetal magnetocardiography (fMCG), a highly effective method of diagnosing cardiac arrhythmia in the fetus, was used. The fMCG recordings were made using a 37-channel SQUID magnetometer.
Results: Incessant premature ventricular contractions (PVC), mainly bigeminy and trigeminy were found in three fetuses with CVAs and in one with CVD, who also had ventricular couplets. The other fetus with CVD, referred because of PVCs, had only sinus tachycardia. ST elevation was noted in two. P: QRS amplitude ratio ranged between 0.22–0.31 (normal value P: QRS ratio = 0.1), indicating early signs of atrial hypertrophy. Fetal movement had a variable impact on PVC's.
Postnatal evaluation demonstrated two persistent left ventricular aneurysms and one CVD; one CVD resolved at 35 weeks gestation, and one fetus remains in utero. Two neonates had incessant PVCs. Both arrhythmias resolved spontaneously while being treated with propranolol.
Conclusion: In fetuses with left ventricular wall defects, precise electrophysiological diagnosis can now be made, including the complexity of ventricular ectopy, arrhythmic response to fetal movement, presence of ST-T wave abnormalities, and atrial amplitude increases. fMCG is complimentary to echocardiographic imaging. Prenatal risk factor assessment using fMCG can support postnatal treatment and follow-up.

P-38

Interrupted inferior vena cava in fetuses with omphalocele. Case series and review of the literature

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Background and aim: Congenital abdominal wall defects (omphalocele or exomphalos and gastroschisis) occur in approximatly 1 of 2000 live births. In omphalocele, congenital heart disease is reported in 15–45% of cases. Associated abnormalities of systemic veins including abnormal angulation or interrupted inferior vena cava (Int-IVC) have occasionally been reported in children but only rarely has this been documented in the fetus. We report a case series of prenatal diagnosis of Int-IVC in association with omphalocele and review the literature.
Method: All cases of omphalocele referred for fetal echocardiography (FE) between 1997 and 2012 were identified from our fetal database. Pre and post-natal medical records were reviewed. A literature search was performed to identify any previous relevant publications.
Results: Of approximately 9,000 fetuses referred for FE 33 had an omphalocele. Mean gestational age at FE was 21+3 weeks (range: 17+5 to 24+0). Seven of the 33 cases (21%) were shown to have an Int-IVC with azygos contiuation to a right-sided superior vena cava (SVC). In six, the heart was structurally normal. One fetus had a ventricular septal defect and suspected coarctation. In all seven, the defect was large, containing liver in 6/7 cases. Since 1975, there have been only 11 publications reporting systemic venous abnormalities in association with omphalocele. We identified 10 cases (with possible cases being reported >once) in >30 years prior to this study. Two of the 10 cases had been misdiagnosed as having an abnormal situs based on int-IVC but post-mortem data confirmed situs to be normal.
Conclusion: In this study we have documented the relatively common association between a large omphalocele and Int-IVC with azygos contiuation to the SCV. We speculate that such an association occurs early in pregnancy as the systemic venous system develops at the same time as abdominal wall defects. Int-IVC in the presence of omphalocele is a developmental abnormality rather than an abnormality of visceral or atrial situs. This may have implications at the time of surgical repair and will influence route of any future cardiac catheterisation.

P-39

Impact of team preparation and procedure technique on outcome of the foetal balloon aortic valvuloplasty

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Introduction: Prenatal interventions (PI) for foetal congenital cardiac malformations such as balloon aortic valvuloplasty (BAV) are performed in several centres worldwide. The centres report variable technical success and complications rates. We present our technical developments regarding the BAV and their impact on technical success of these procedures.
Methods: In years 2011–2012 13 BAV were performed in foetuses aged 20–31 weeks (avg.25). Evolving left heart hypoplasia was an indication for BAV in 7, foetal heart failure in 5 and combination of both in 1 case. Before the BAV program started, the procedure was simulated on the foetal cadaver. Prior to the procedure, aortic valve (AoV) diameter and distance from the apex to the AoV were measured. Safe position of the needle tip was assessed basing on expected position of proximal end of balloon in the left ventricle (LV). The LV was always punctured in a single attempt. The 18 G needle was inserted parallel to the interventricular septum (IVS) as close to the AoV as possible without threatening both aortic and mitral valves. Balance Middleweight (Abbott)

guidewire was used in 4 and Whisper MS (Abbott) in 9 cases. Balloon size was calculated as 120% of AoV diameter. Maverick2 (Boston Scientific) balloons were used in 12 cases and Trek (Abbott) balloon in 1 case with size range 3.5–4 mm.
Results: In all cases the LV was successfully punctured, the AoV was traversed with a guidewire, the balloon was advanced through the AoV and inflated up to 3 times. The Whisper MS curved-tip hydrophilic guidewires showed better manoeuvrability and support. All foetuses survived the procedure. Pericardial effusion (6 cases) and bradycardia (5 cases) were the most common complications. One foetus died in utero one day after BAV due to premature placental separation. In 1 case intrauterine death occurred in late pregnancy due to placental insufficiency not related to the procedure.
Conclusions: Practice in the dissection room, proper selection of the equipment, careful measurements of the LV size and safe needle position help to decrease learning curve and reduce risk of lethal complications.

P-40

Antenatal and postnatal right isomerism of the atrial appendages in a 20-year single institution experience

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Background: Right isomerism of the atrial appendages (RAI), particularly if diagnosed prenatally is thought to be one of the worst forms of cardiac disease.
Methods: Single centre retrospective observational study of all cases with pre- or postnatal diagnosis of RAI during a 20-year period. All prenatal cases with visceroatrial situs ambiguous, severe congenital heart disease and the presence of an inferior vena cava were included. Postnatally RAI was confirmed by the presence of asplenia and/or bilateral right sided bronchial tree in chest X-ray or magnetic resonance.
Results: Between 1992 and 2012 RAI was diagnosed in 36 cases. Prenatal diagnosis was achieved in 18 fetuses. Pregnancy was terminated in 9 (50%), palliative care was chosen in 3 newborns and 6 patients were treated. In total only 4 of 18 (22%) prenatally diagnosen children were alive at end of follow up. 18 had a postnatal diagnosis with 6 choosing palliative care. Four of the remaining 12 died after surgery and 8 patients are alive. Of total 18 patients intended to treat 12 had an AVSD (unbalanced in 7,+pulmonary atresia (PA) in 4), single right ventricle in 3 and compex anatomies in 3 cases (+PA in 2). TAPVD to a systemic vein was diagnosed in 7/18 cases. Non confluent pulmonary arteries were present in 5/18 patients. The 12 survivors had a median follow up of 4.1(0.02–18) years.
Fontan (n = 13) or biventricular (n = 1) repair were planned in 14/18 (77%). 2 patients were considered unsuitable for Fontan completion later and receive palliative care. 2 cases underwent successfully cavopulmonary connection and are waiting for Fontan completion. 9 underwent complete Fontan repair where 2 failed and died. One patient underwent successful biventricular repair. Successful univentricular palliation or biventricular repair were achieved in only 8/18(44%) patients. Fetal diagnosis or anatomical findings, such as TAPVD, non-confluent pulmonary arteries were not predictive for Fontan failure or outcome. (p = NS).
Conclusion: Our data confirm that RAI patients have a dismal prognosis with successful univentricular palliation or biventricular repair being feasible in less than 50% of all cases. Prenatal diagnosis and/or anatomical findings seem not to influence outcome.

P-41

Different hemodynamic patterns in head-up tilt test in 400 pediatric cases with unexplained syncope

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Objectives: The purpose of this study is to assessment different hemodynamic patterns in head-up tilt test in 400 pediatric cases with unexplained syncope
Methods: Head up tilt test was performed 400 pediatric patients with unexplained syncope. Blood pressure and heart rate were monitored simultaneously during 35-min (patients in 65° upright position) passive head up tilt test. According to their different hemodynamic patterns in head-up tilt test, subjcets were divided into orthostatic intolerance (OI) response pattern, postural orthostatic tachycardia syndrome (POTS) response pattern, orthostatic hypotension (OH) response pattern, asymptomatic orthostatic hypotension (AOH) response pattern, vasovagal (VVS) response pattern, and normal response (NR)pattern. Vasovagal response pattern was consisted of vasodepressor (type 3), cardioinhibitory (type 2A, type 2B), and mixed (type 1) pattern. Age, sex, baseline heart rate, baseline blood pressure, duration of symptoms, and number of syncope were recorded in all groups.
Results: The ages of 400 pediatric patient included in the study were in a range from 5 to 18 years (mean 12,6 ± 2,6 years). Two hundered sixty four (66%) were females and 136 (34%) male. Two hundered seventy seven (65%) of subjects displayed the hemodynamic pattern of NR, 28 (%) OI response, 22 (%) POTS response, 7 (%) OH response, 28 (%) AOH response, and 38 (%) VVS response. Vasovagal response pattern was consisted of 9 (%) type 3 response, 10 (%) Tip 2A response, 2 (%) Tip 2B response, and 17 (%) mixed (type 1) response. The most frequently abnormal monitored hemodynamic patterns were Orthostatic intolerance syndromes (OI, POTS, AOI). After that there was VVS. There were no statistically significant differences between the groups with regard to age, gender, baseline heart rate, baseline blood pressure, and duration of symptoms ($p > 0.05$). The syncopal attacks of the children with VVS reponse group was significantly more frequent than that of the children with OI, POTS, and AOI ($p < 0.01$)
Conclusions: We observed nine different hemodynamic patterns in head-up tilt test in 400 pediatric cases with unexplained syncope. The most frequently abnormal monitored hemodynamic patterns were Orthostatic intolerance syndromes.

P-42

Short-Term Efficacy of Oral Rehydration Salts and Propranolol Treatment in Pediatric Patients Showing Orthostatic İntolerance Symptoms During Head Up Tilt Test

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Purpose: The goal of this study is to evaluate short-term efficacy of the therapeutic regimen composed of oral rehydration salts, propranolol in children showing orthostatic intolerance (OI) symptoms and different hemodynamic patterns during head up tilt table test.
Method: Pediatric patients who were exposed to tilt test because of syncope and diagnosed with orthostatic intolerance (OI) disorder were divided into groups based on their distinct

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

hemodynamic patterns. Group I consisted of pediatric OI group, (n = 28). Group 2 consisted of POTS group, (n = 24). Group 3 consisted of patients meeting the criteria of accelerations in heart rate of OI but not showing any symptoms (control group, n = 26). The patients in group 1 and group 2 were administered rehydration salt and propranolol treatment together. The patients in group 3, control subjects, were given rehydration salts treatment alone. The response rates of patients to rehydration salts and propranolol treatment were evaluated.
Results: There were no statistically significant differences between the groups with regard to age, gender, BMI, the frequency of syncope attacks prior to the treatment. Post-treatment frequency of syncope attacks were found to be significantly reduced in all groups in comparison with pre-treatment status. The median number of pre-treatment vs. post-treatment syncope attacks were 3 (min 2 max 20) vs. 0 (min 0 max 3) in group 1 ($P < 0.01$), 3 (min 2 max 5) vs. 0 (min 0 max 1) in group 2 ($P < 0.01$), 3 (min 2 max 10) vs. 0 (min 0 max 1) in group 3 ($P < 0.01$). When treatment response rates of OI group were compared with that of POTS group and control group, it was found to be statistically significantly lower than both POTS group and control group ($p < 0.01$, $p = 0.03$, respectively).
Conclusions: In general, therapy with rehydration salts and propranolol was found to be efficacious to reduce the frequency of syncope attacks in pediatric patients showing orthostatic intolerance disorder. That the patients in OI group had lower rates of treatment response was thought to be due to that this group of patients were likely to have a distinct pathophysiology and hemodynamic pattern.

P-43
Evaluation of Myocardial Functions in Children Receiving Anthracyclines
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Objective: In our study, using echocardiographic measurements in patients with acute lymphoblastic leukemia receiving anthracycline therapy have evaluated presence of anthracycline-induced cardiac dysfunction and it has been carried out to determine the statistical echocardiographic parameters that best predict it.
Methods: Thirty cases (mean age 9.87 ± 3.92 years, 13 males and 17 females) with pediatric hematological conditions receiving anthracycline therapy were enrolled in the study, along with 30 age-matched controls. Their cardiac functions were evaluated echocardiographically with two dimensional, M-mode, PW Doppler and PW tissue Doppler methods.
Results: The left ventricular cardiac functions were significantly reduced in the study group. The fractional shortening (FS) was 36.09 ± 3.17% (range 28–41%) in the study group as opposed to 39.65 ± 3.21% (34–45%) in controls ($p = 0.03$). The Diastolic functions (E/A ratio) weren't significantly change in the study group. (Right ventricular (RV) flow E/A 1.36 ± 0.29 and 1.34 ± 0.69, $p > 0.05$. Left ventricular (LV) flow E/A 1.53 ± 0,31 and 1.58 ± 0,35, $p > 0.05$. The myocardial performance index (MPI) was calculated in the study subjects using both PW Doppler and PW tissue Doppler methods and compared to controls. RV flow MPI 0.39 ± 0.02 and 0.33 ± 0.03, $p < 0.01$. LV flow MPI 0.41 ± 0.08 and 0.34 ± 0.04, $p < 0.01$. RV tissue MPI 0.44 ± 0.04 and 0.39 ± 0.05, $p < 0.01$. LV tissue MPI 0.42 ± 0.02 and 0.36 ± 0.01, $p < 0.01$.
Conclusions: The results of our study show that MPI (especially LV flow MPI, LV tissue MPI) and systolic functional changes are better indicators of anthracycline cardiotoxicity than are diastolic functions. We suggest that LV flow MPI and LV tissue MPI be primarily used in the search for anthracycline cardiotoxicity.

P-44
The Aortic Valve Index as a predictor of a reoperation after Interrupted Aortic Arch (IAA) and Ventricular Septal Defect (VSD) one-stage surgery
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Introduction: IAA is a rare congenital defect of cardiovascular system often accompanied by VSD. The link between results of its early correction including the need of later re-treatment and diameter of aortic annulus could be analysed.
Material: We analysed 28 children with IAA+VSD who underwent single-stage repair in 1996–2010. The mean age at the operation was 19.4 ± 7.4 days. 22 children (78.5%) had IAA type B, 3 - A, 3 - C.
Methods: Patients were divided into two groups based on preoperative echocardiographic measurements of aortic annulus size (AoV) [mm] and body weight (b.w.) [kg] The Aortic Valve Index (Hirata et al.) was calculated : AVI = AoV – b.w. In the group I there were 23 patients (82.1%) with large aortic annulus (AVI > 1.5). Group II consisted of 5 patients with small aortic annulus (AVI < 1.5)
Results: One neonate died in the early postoperative period.
9 patients (32.1%) aged 2 months to 3.2 years (with a mean age of 2.8 years) required re-operation or re-intervention.
5 patients (21.7%) of the group I required re-treatment: 2 - balloon angioplasty, 1 - resection of reCoA, 1 - VSD recanalization, 1 – reoperation for LVOTO and twice aortic arch angioplasty.
In group II 4 patients (all with IAA type B) required re-treatment: 2 patients underwent angioplasty of the aortic arch, one - angioplasty of the aortic arch, then reoperation for LVOTO and in the 10 year the Konno-Ross procedure with success. The last patient underwent balloon angioplasty of LVOTO and in the age of 5 years the Ross procedure. He died in the early postoperative period.
Conclusions: Good medium-term results of one-stage repair of patients with IAA + VSD were found. Almost one-third of patients required re-treatment. In neonates with a relatively small aortic annulus mostly was necessary to perform re-intervention. In children with large aortic annulus, only some required re-intervention. The results indicate of a link between the aortic valve index and the need for re-operation.

P-45
Rhizomelic chondrodysplasia punctata and cardiac pathology
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Introduction: Rhizomelic Chondrodysplasia Punctata (RCDP) is an autosomal recessive peroxisomal disorder characterized by

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

rhizomelia, contractures, congenital cataracts, facial dysmorphia, severe psychomotor defects and growth retardation. Biochemically, the levels of plasmalogens (major constituents of cellular membranes) are low in erythrocytes due to a genetic defect in their biosynthesis. Cardiac muscle contains high concentrations of plasmalogens. Recently cardiac dysfunction was found in a mouse model for RCDP with undetectable plasmalogens levels in all tissues including the heart. This suggests the importance of plasmalogens in normal cardiac development and function. Congenital heart disease (CHD), however, has not been recognized as a major characteristic of RCDP.
Objectives: We aimed to determine the prevalence of CHD found in RCDP patients as well as to describe genetic, biochemical and cardiac correlations.
Methods: We included 23 patients with genetically proven RCDP. The genetic, biochemical and physical data were evaluated. Electrocardiograms and echocardiograms were reviewed.
Results: Cardiac data were available for eighteen patients. Twelve (52%) had CHD. All 12 had type 1 RCDP and 11 (92%) had the PEX 7:c.[875T>A] mutation, of whom 7 homozygous (58%). Plasmalogen levels were significantly lower in the patients with CHD. Cardiac lesions included: septal defects (80% atrial), patent ductus arteriosus, pulmonary artery hypoplasia, Tetrology of Fallot and mitral valve prolapse (mostly older patients).
Conclusion: The CHD prevalence among RCDP patients was at least 52%, significantly higher than among the normal population. Plasmalogen levels were significantly lower in patients with CHD. Routine cardiac evaluation should be included in the clinical management of RCDP patients.

P-46
Evaluation of 316 Children with Acute Rheumatic Fever; A Single Center Study From Turkey
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Objectives: We aimed to investigate the diagnostic findings of acute rheumatic fever patients (ARF) seen over the past seven years in center Anatolia in Turkey. We evaluted long term effects of treatment modalities on cardiac sequela rate.
Method: We analyzed the medical records of 316 patients who were diagnosed with new onset ARF, between January 2005 and December 2011 years, retrospectively.
Results: The mean age of the patients was 10,5 years and the female/male ratio was 1,2. The most frequent complaints were arthritis in 78,1% of the patients. The most frequent major finding was the combined presence of arthritis with carditis in 38,6% of the patients. Monoarthritis was identified in 30,3% of patients with joint involvement. Carditis was detected in 242 cases (76,5%) and 102 cases (32.2%) presented with Sydenham's Chorea. Carditis was found in 64% of the patients as isolated mitral regurgitation. Patients with silent carditis constituted of 27,2% of all cases with carditis. Rheumatic valvuler disease was more frequent during the follow up of patients with more severe carditis at initial attack and in cases with combined valve involvement ($p < 0.05$). Severe form heart failure was detected in 8 cases (3,3%) and valvular surgery was performed in two cases. The side effect rate in patients recevied acetylsalicylic acid (19.7%) was higher than patients received Naproxen sodium ($p < 0.005$). The median follow-up duration was 28 months. The rate of compliance with secondary prophylaxis was 80,7%. Recurrent attacks were detected in 15,8% of patients. The rate of recurrence was 4,2% in patients with a full compliance to secondary prophylaxy, whereas this rate was 65,2% in not compliant patients ($p < 0.001$). The prevalence of chronic rheumatic valvular disease in patients received Naproxen sodium was not different from patients received acetylsalicylic acid. Rebound, recurrence and cardiac sequlea rate in patients received steroid were not different from patients treated with a non steroidal drug (acetylsalicylic acid or naproxen).
Conclusion: This study demonstrated that diagnostic findings of rheumatic fever have not changed for seven years in center Anatolia. Naproxen was an excellent alternative antiinflamatory drug.

P-47
High Anxiety and Depression Scores in School-aged Children Defining Syncope Together with Cardiac Sypmtoms
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Objectives: The aim of this study was to investigate the psychological factors in school-aged pediatric population defining syncope together with palpitation and chest pain.
Methods: Seventy school-aged children defining syncope, palpitation and chest pain were investigated. "The Screen for Child Anxiety Related Emotional Disorders (Birmaher B., J Am Acad Child Adolesc Psychiatry, 1997; 36: 545–53)" and "Beck Depression Invantory (Kovacs M., Psychopharmacol Bull, 1985; 21: 995–8)" scales were applied to patients to determine psychological background. Special features at history (family history, worrying events at near time, negative effect on daily life, duration of complaint), physical findings (heart rate, blood pressure), laboratory findings (hemoglobin, serum glucose and electrolytes, thyroid function), electrocardiography (ECG) (rhythm, QTc duration) and echocardiography (ECHO) findings, 24-hour rhythm Holter monitorization results were also investigated.
Results: The mean age of children was 13.7 ± 2.1 (9–17) years. The female/male ratio was 3.7 (55/15). The median anxiety score was 30 (7–60) (cutt-off point of 25) and depression score was 13 (2–33) (cutt-off point of 19). The anxiety score was high in 54 patients (77%) and depression score was high in 17 patients (24%). In 15 patients (21%) both scores were high, however in 14 patients (20%) both scores were normal. The mean hemoglobin was 13.2 ± 1.3 (9.2–15.8) gr/dl, the mean QTc duration was 383 ± 31 (320–491) ms. Other clinical and laboratory findings are listed in Table 1. There were significant correlations between anxiety scores and female sex (r: 0.3, $p < 0.01$), and negative effect on daily life ($r = 0.3$, $p < 0.01$). There were also significant correlations between depression scores and female sex (r: 0.3, $p < 0.05$), family history (r: 0.3, $p < 0.05$), worrying events at near time (r: 0.2, $p < 0.05$), and negative effect on daily life (r: 0.2, $p < 0.05$). The correlation between anxiety and depression scores was also significant (r: 0.5, $p < 0.0001$).
Conclusions: Psychological factors causing anxiety and depression may result in psychosomatic disorders. It could be more reasonable to direct these patients with high anxiety or depression scores to psychotherapy before making some advanced laboratory studies and prescribing medicines.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

P-48

Cardiovascular Dysfunction and Pulmonary Edema in Children with Scorpion Envenomation: A Report of 5 Patients

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Introduction: Scorpion envenomation is a life-threatening emergency and a common public health problem in children. Severe scorpion envenoming is characterized by cardiocirculatory failure which may lead to pulmonary edema. Children are at greater risk of developing these complications. We aimed to evaluate clinical, laboratory, treatment, and outcome characteristics of children with scorpion envenomation and the need for intensive care unit admission.
Cases: Five patients aged between 4–7 years were admitted for scorpion envenomation. Of the five patients four were stung from feet while one was stung from the head. Three patients had respiratory distress, four had tachycardia and hypotension and three had somnolence. Arterial blood gas examination showed acidosis in one patient. Three had X-ray findings consistent withpulmonary edema. All patients had sinus tachycardia and ST-T changes on ECG. Troponin values were increased in four patients (5–11 (normal range 0.02–0.06 ng/ml)). Echocardiographic evaluation showed evidence of moderate to severe sistolic dysfunction and reduced ejection fraction in four patients. Two patients had serious cardiomyopathy (EF:30%). Scorpion venom were given to all patients. Intensive inotropic treatment were given to patients with serious cardiomyopathy. Prazosin, diuretics and oxygen treatment were given to patients with pulmonary edema. The clinical course of all patients was satisfactory and the laboratory, electrocardiographic and echocardiographic changes returned to normal within 6 days (mean: 3.5 days).
Conclusion: Severe scorpion envenoming is characterized by cardiocirculatory failure which may lead to pulmonary oedema. Involvement of the heart has been attributed to the massive release of catecholamines and/or to a direct toxic effect of the venom on cardiac fibres, while pulmonary oedema has been considered to be of cardiogenic or non-cardiogenic origin. Based on the clinical course of our patients we think that patients with scorpion envenomations should be observed in the intensive care unit for the risk of developing serious cardiomyopathy and pulmonary edema.

P-49

Assessment of cardiac functions in patient with Trisomy 21

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Introduction: Extra genetic material in Down syndrome (DS) may affect the function of any organ system. We wanted to evaluate cardiac functions using two-dimensional, M-mode and Doppler echocardiography even in absence of congenital and acquired heart disease in patients with Down syndrome.
Methods: 115 patients with DS between 6 and 13 years of age with clinically and anatomically normal heart and 55 healthy children were included in this cross-sectional. DS was diagnosed by a karyotype test. Patients with mosaic type were not included in this study. Systolic and diastolic functions were evaluated by echocardiography.
Results: Systolic pulmonary artery pressure was significantly higher in patients with DS than in healthy group ($p < 0.001$). They had significantly higher left ventricular mass, ejection fraction, mitral annular plane systolic excursion values. Pulsed waved Doppler transmitral E (early diastolic velocity)/A (late diastolic velocity), tissue Doppler mitral annular Ea (early diastolic velocity)/Aa (late diastolic velocity), transtricuspid E/A and tricuspid valve annulus Ea/Aa, pulmonary venous Doppler S (systolic)/D (diastolic) wave ratio were lower in patients with DS than in control group ($p = 0.04$, $p = 0.001$, $p < 0.05$, $p < 0.001$, $p < 0.001$ respectively). Mitral and tricuspid annular Ea were lower in patients with DS ($p < 0.001$). The right and left ventricular myocardial performance index were higher in patients with DS than in controls ($p < 0.01$).
Conclusion: Conventional and tissue Doppler echocardiography is important in determining the subclinic cardiac diastolic dysfunction in patient with DS. We think that these changes may be due to autonomic dysfunction, pulmonary hypertension, myocardial fiber structure.

P-50

Assessment of Arterial Stiffness Influence on Aortic Root Dilation in Children with Repaired Tetralogy of Fallot

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Aortic root dilatation is a known feature in tetralogy of Fallot (TOF). Progressive aortic root dilatation has been reported in some cases with tetralogy of Fallot even after reparative surgery. Aortic root dilatation was thought to be due to increased aortic flow resulted from right to left shunting and dextraposition. Besides, intrinsic histological abnormalities of the aortic wall that present since from infancy are important causative factors leading to aortic root dilatation. This study was conducted to explore relation of arterial stiffness with aortic root dilation in cases with repaired tetralogy of Fallot.
The study population consisted of 50 cases with repaired TOF and 27 age-matched healthy control children. The cases with repaired tetralogy of Fallot were divided into two groups as with and without dilated aortic root (group 1 and 2, respectively). Aortic strain was lower in group 1 than group 2 and controls ($p = 0004$, $p < 0.001$), aortic distensibility was lower in group 1 than controls ($p = 0.016$), and beta stiffness index was higher ($p = 0.016$). Aortic distensibility was lower and beta stiffness index was higher in group 1 compared with group 2, although statistically insignificant ($p = 0.06$, $p = 0.06$). These results suggested that aortic strain which indicates elasticity of the aorta may be affected earlier than the distensibility and beta stiffness index.
Arterial stiffness may contribute to progressive dilation of aortic root in patients with repaired TOF. A better understanding of the pathophysiology will help to treatment strategies in TOF which patient with aortic dilation.

P-51

Five years of cardiologic follow-up of children with acute Chagas disease infected by the oral route in Venezuela

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Background: Chagas disease (ChD) is an endemic disease, frequent cause of morbidity and mortality in Latin America. Acute clinical cases have high mortality if not treated promptly, with life-threatening arrhythmias and severe myocarditis. Recent outbreaks of oral transmission reveal its importance and highlight historical underestimation of this form of infection. Early diagnosis and treatment with benznidazole or nifurtimox can warrant better outcomes in this acute phase since low rate sequels occur. There are not previous studies of follow-up of children orally infected with Trypanosoma cruzi.
Objective: The aim of this study is to describe 5 years of cardiovascular follow-up in a group of children with ChD orally acquired.
Methods: During 2007–2012 period, 51 ChD pediatric patients were followed by clinical visits, annuals electrocardiograms (ECG), echocardiograms (ECHO) 24-hour ECG Holter monitoring, and laboratory tests (ELISA, Indirect hemagglutination, PCR, lytic antibodies).
Results: 51 children (68% male, 32% female), mean 13 years old, received specific treatment. During the acute phase none had congestive heart failure (CHF). Electrocardiographic findings were not specific during all follow up: T-wave inversion in right precordial leads with normal progression (84.3%), indeterminate T-wave abnormality (18%), pathologic elevation of ST segment > 2,5 mm (4%) early repolarization, and sinus bradycardia (4%). Holter monitor findings (11%); sinus tachycardia, premature supraventricular beats and ventricular polymorphic arrhythmia. Echocardiographic findings (first evaluation) (9%): pericardial effusion and left atrial and ventricular dilatation. Between 2010–2011 a second course of anti-parasitic treatment was given to 51% of these patients and in spite that 78% persist with positive lytic antibodies and some have positive PCR, all these cardiac manifestations disappeared in the five years follow up. All had normal systolic function.
Conclusion: In pediatric patients with confirmed acute ChD associated with oral transmission treated early, we are able to obtain fair outcome with low risk of complications or sequels. Few non-specific cardiovascular findings have been found 5 years later, however positive serology suggest infection persistence even in absence of relevant cardiovascular manifestations. We consider necessary to perform long term follow up, with routine clinical surveillance, serial electrocardiograms, and echocardiograms in order to rule out cardiac disease progression of infected children.

P-52
Prognostic Factors In Pulmonary Atresia With Ventricular Septal Defect

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Objectives: The main purpose of this work is to evaluate the impact of different anatomical and therapeutical factors in the prognosis of patients with pulmonary atresia with ventricular septal defect (PAtr + VSD).
Materials and Methods: Retrospective analysis of 84 patients with PAtr + VSD and balanced ventricles consecutively treated in our center. Demographic data, pulmonary artery (PA) anatomical features, including PA dimensions evaluated by McGoon Index (McGoonI), presence of non-confluent PAs, surgical and percutaneous approach required and health condition on follow-up were recorded.
Results: 42 (50%) patients were male. Major aortic pulmonary collateral arteries (MAPCAs) were found in 48 (57,1%) patients, PA hypoplasia with McGoonI <1,5 in 50 (59,5%) and non-confluent PAs in 20 (23,8%). There was a late referral to our center (beyond the neonatal period) of 20 patients (24%). Corrective surgery (Corr Surg) was performed in 35 patients (41,7%) between 0,9 and 23 years of age, and all had a McGoonI ≥1,5. In 15 patients (17,9%), right ventricle to PA continuity was established without VSD closure as there was pulmonary hypoplasia. The initial McGoonI predicted the possibility of corrective surgery with an odds-ratio of 12.1 ($p = 0.04$); the best cut-off was a McGoonI >1.3 (by ROC curve analysis). Final McGoonI was 1,8 in the group who underwent corrective surgery and 1,4 in the group who was considered unsuitable for correction ($p = 0.03$). Age at the time of first surgery was not different between the group who had Corr Surg and the remaining patients. Presence of non-confluent pulmonary arteries did not influence negatively the possibility of total correction (p NS). The actuarial survival of the whole group was 96,4% at one year of age; 92,8% at 2 years; 90,4% at 5 years and 88% at 10 years. At last follow up, 38 (55,1%) patients were in NYHA class I or II.
Conclusions: In patients with PAtr + VSD and balanced ventricles, the initial McGoonI predicted the possibility of corrective surgery with a best cut off value of 1,3. Age at time of initial surgery and presence of non-confluent pulmonary arteries did not influence the possibility of corrective surgery.

P-53
The importance of cardiac biomarkers and echocardiography in the evaluation of heart failure children younger than 3 years

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Introduction: N-terminal pro-hormone brain natriuretic peptide (NT-proBNP) and brain-natriuretic peptide (BNP) are cardiac biomarkers that provide diagnostic information in heart failure (HF) patients. We aim to assess the serum levels of NT-proBNP and BNP in HF children younger than 3 years with heart abnormalities and to evaluate the correlations with NYHA/Ross functional class and the left ventricle ejection fraction (LVEF).
Methods: 24 children with HF due to congenital heart diseases and dilated cardiomyopathy were enrolled. Children were analyzed based on NYHA/Ross functional class. We measured the serum levels of NT-proBNP and BNP and LVEF was calculated in all cases.
Results: Patients with cyanotic heart diseases recorded the highest values of median NT-proBNP level and median BNP level (248.0 pg/mL and 3000.6 pg/mL, respectively). Both heart biomarkers NT-proBNP and BNP had a negative relationship with LVEF: coefficient of correlation Spearman was −0.165 (95% CI −0.682–0.26), with $p = 0.44$ for NT-proBNP and −0.066 (95% CI −0.418–0.345), with $p = 0.76$ for BNP. We found a very good correlation between NT-proBNP and BNP, with a coefficient of correlation Spearman of 0.903 (95% CI 0.526–0.887), and with $p < 0.001$. *Conclusions:* The cardiac biomarker BNP was found related to the severity of HF in infants and small children younger than 3 years with HF due to congenital heart diseases and dilated cardiomyopathy. The 3 patients with dilated cardiomyopathy proved an association between the highest values of NT-proBNP and BNP with the lower LVEF. The other patients from our study group did not prove this association, data being without statistic significance.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

P-54

Early echocardiographic improvements and ROC analysis to detect cut-off levels of laboratory and echocardiographic values in patients with rheumatic carditis

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Objectives: To evaluate the improvement in rheumatic valvulitis just after the inflammation has subsided and to determine the cut-off values associated with rheumatic carditis.
Methods: The medical records of the patients diagnosed with rheumatic carditis, including history, physical examination, electrocardiogram, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), echocardiography, and medications were retrospectively evaluated. All the cases with carditis had prednisolone treatment for two weeks. If they responded to therapy, salicylate was started and prednisolone dose was gradually decreased and stopped within two weeks. Salicylate treatment continued at least 4–6 weeks. The cut-off values for the variables associated with valvulitis were determined by Roc analysis.
Results: The number of patients with any valvulitis was 38. The mean age at diagnosis was 11.9 ± 2.6 years and most of them were male (58.3%). Thirteen had subclinic carditis. Thirty-four had mitral regurgitation (MR), nineteen had aortic regurgitation (AR). Fifteen of those had insufficiency in both valves. No valvulitis was determined in ten cases (26.3%) three months after diagnosis. When evaluated separately; MR disappeared in twelve cases, AR in five, and combined MR and AR in seven cases. The determined cut-off values via Roc analysis for the variables associated with valvulitis such as CRP, ESR, left vetricular end diastolic dimension (LVEDd), LVEDd indexed to body surface area (BSA), left ventricular end systolic dimension (LVESd), and LVESd indexed to BSA were 1.62 mg/dl (sensitivity: 0.87, specificity: 0.81), 48 mm/hr (sensitivity:0.82, specificity: 0.90), 40.5 mm (sensitivity: 0.71, specificity: 0.32), 31.8 mm/m^2 (sensitivity: 0.66, specificity: 0.29), 24.5 mm (sensitivity: 0.53, specificity: 0.45), 19 mm/m^2 (sensitivity: 0.74, specificity: 0.34), respectively. The most significant cut-off values found were the ones associated with ESR and CRP.
Conclusions: Rheumatic valvulitis may disappear in significant proportion of the cases within few months. Factors associated with the course of valvulitis vary widely according to the valve involved. The determined cut-off values related to valvulitis may help the clinicians in the management of rheumatic carditis.

P-55

Spontaneous closure of a symptomatic coronary artery fistula just within a few days of newborn period

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Introduction: We present a a rare case of spontaneous closure of a fistula between the left coronary artery (LCA) and the right ventricle with in a few days of newborn period.
Case: A 14-day-old male newborn was referred to our clinic for investigation of tachypnea and cardiac murmur which had been discovered on routine postnatal examination. His medical history was unremarkable. Physical examination revealed mild tachypnea and a continous murmur at the left lower sternal border. The electrocardiogram was normal while the chest roentgenogram showed mild cardiomegaly. Mild right ventricular enlargement and dilated left coronary artery (LCA) with the inner diameter of 3.3 mm were determined by two dimensional echocardiography. A color flow Doppler echocardiography revealed turbulent flow of a large CAF between the left circumflex artery and the right ventricle (RV), together with resultant continuous turbulent flow pattern (Figure 1 and 2). The neonate was hospitalized in intensive care unit and furosemide was started. During follow-up, his clinical status remained stabile and tachypnea gradually resolved. The murmur disappeared four days after hospitalization. Repeat echocardiography was performed and interestingly, no fistula was detected. There have been no sypmtoms on clinical examination and no evidence of a coronary fistula on follow-up echocardiograms for 10 months of duration.
Conclusion: The spontaneous closure of CAF was found to be more likely in the cases younger than two years with small-sized fistulas opening into the right-sided structures, especially to the RV. Nevertheless, the spontaneous closure is very rare in cases with significant shunt. To the best of our knowledge, this is the first case with documented spontaneous closure of CAF just within the newborn period.

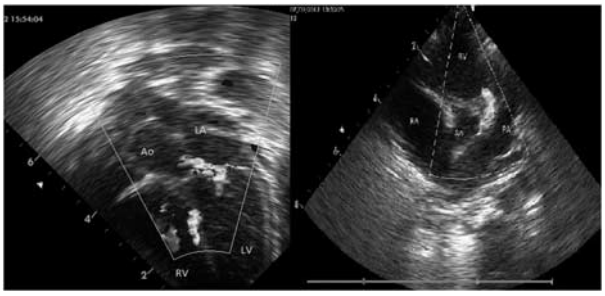

Figure 1: Apical five chamber view showing a coronary artery fistula arising from the left coronary artery that drainage into the right ventricle
Figure 2: Parasternal short axis view demostrating a turbulent flow between left circumflex artery and right ventricle

P-56

Congenitally Corrected Transposition of the Great Arteries: Single Center Experience

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Introduction: There are several rhythm and conduction disturbances associated with congenitally corrected transposition of the great arteries (ccTGA). The purpose of this study is to determine the incidence of rhythm and conduction disturbances in ccTGA patients with two adequate-sized ventricles.
Methods: Retrospective analysis of records of 49 patients from single center were reviewed to determine long term results of ccTGA patients.
Results: The study comprised 49 patients (15 girls, 34 boys). The median age of the patients at initial presentation was 3 months

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

(1 day–34 years) and mean period of follow-up was 4.5 ± 1.8 years (1 month–22 years). Forty seven of them had associated heart anomalies. The most common associated lesion was VSD (38 patients). Pulmonary valve abnormalities were second most common lesion. Pulmonary stenosis was more common than pulmonary atresia (17 versus 6 patients). As usual Ebstein anomaly and tricuspid regurgitation were quite common among our patients. During the follow-up period 18 patients had a total of 22 operations. Systemic to pulmonary circulation shunts were the most common procedures (9 patients). Conventional biventricular repair was and double switch procedure were performed equally (5/5 patients). Tricuspid valve replacement was performed in 2 patients. At initial examination, two patients had first-degree AV block, one second-degree AV block and one congenitally complete AV block, Additionally, one patient had atrial ectopic rhythm, one left bundle branch block. Supraventricular tachycardia was detected in 3 patients. At follow-up, complete AV block developed in 5 patients after intracardiac surgery. Pacemaker implantation was required for these patients and one patient with congenitally complete AV block.
Conclusions: Patients diagnosed as ccTGA should be followed lifelong. During the disease course they may need different type of surgical procedures and ccTGA may complicate with different types of rhythm and conduction disturbances at any time.

P-57
Kawasaki disease in Germany: A nationwide survey of 202 children
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Introduction: Kawasaki disease (KD), an acute febrile illness of unknown aetiology, is most common in children of Asian origin, but affects all ethnic groups. Incidence, age distribution, course and complication rate in Germany is unknown.
Methods: Prospective study of the German surveillance unit for rare paediatric diseases (ESPED) of children (<5 years) with KD, treated with iv Immunoglobulins (IVIG).
Results: 1/2011–9/2012 there were 202 patients (137 male) with KD. We found a significant peak incidence during winter but no difference in age distribution. Compared to older children, infants <1 year significantly less developed the complete form of KD (47,5% (19/40) vs. 74,7% (121/162); $p = 0.007$), especially cervical lymphadenopathy, and enanthema were missing. Instead in this age group hematological disorders (e.g. anemia: 65% (26/40) vs. ≥43,2% (70/162), $p < 0.02$) and coronary artery aneurysms (30% (12/40) vs. 12,3% (20/162) $p = 0.0082$) were more frequent. There was no difference in time to first IVIG-treatment initiation. 49 patients received a second course of IVIG and 18 patients were additionally treated with steroids. However, these patients did not indicate an increased rate of coronary artery aneurysms.
Conclusions: Infants <1 year present in the acute state of Kawasaki disease with fewer clinical symptoms. In contrast, hematological parameters were more frequently pathologically altered. The higher incidence of coronary artery aneurysms in this age group needs to be taken into account in the cardiological follow-up.

P-58
A retrospective study of cardiomyopathy in children and adolescents: etiology and function of oxidative phosphorylation (oxphos)
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Introduction: Cardiomyopathy (CMP), a heterogeneous group of myocardial disorders, is rare in children but involves a high mortality rate. The etiology is divers and often hereditary. The main causes are isolated mutations, neuromuscular dysfunctions, malformation syndromes and metabolic disorders. The severity and familial recurrence of CMP highlights the importance of genetic knowledge and consequent prenatal diagnostics. The aim of this study is to describe patients with CMP followed at the pediatric cardiology department of Ghent University Hospital (UZG) with special attention for the clinical and laboratory characteristics of mitochondrial disease.
Methods: A retrospective study in children and adolescents with CMP, followed between 2003–2011 in the UZG, with registration of clinical presentation, diagnostic results, etiology (if known) and treatment. Based on a literature study, criteria that raise the suspicion of underlying defects in oxidative phosphorylation (oxphos), were described.
Results: Between 2003 and 2011, 66 patients with CMP were followed (40 boys, 26 girls), diagnosed with CMP between 1984 and 2011. Eighteen patients died at a median age 10 months (ranging from 6 days to 13,6 years). At the time of the study in 2011, the age of the surviving patients varied between 15 months and 29 years (median age 11,9 years). Hypertrophic cardiomyopathy (HCM) was diagnosed in 32 patients (48,5%) and dilated cardiomyopathy (DCM) in 27 (41%). The remaining 10% showed a restrictive or other non-classified form of CMP (4 left ventricular non-compaction, 1 takotsubo cardiomyopathy and 1 atypical right ventricular cardiomyopathy). Etiology was known in 50% of the patients and was divided into genetic (25/33) and non-genetic (8/33) causes. Genetic etiology consisted of isolated mutations (8/25, 32%), neuromuscular syndromes (3/25, 12%), malformation syndromes (3/25, 12%) and metabolic pathology (11/25, 44%). In 64% (7/11) of the metabolic cases, CMP was caused by was a respiratory chain defect. After implementation of the criteria for oxphos defects in the group with unknown origin, more elaborated metabolic examinations were recommended in 13 more patients.
Conclusion: Presentation and etiology of CMP in children is heterogeneous. Deficiency in oxphos-mechanism are relatively frequent. In our study population these disorders are probably under diagnosed.

P-59
Longitudinal systolic left ventricular-right ventricular interaction in pediatric and young adult patients with TOF: a magnetic resonance imaging and M-mode echocardiography study
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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Introduction: In operated Tetralogy of Fallot (TOF) patients there is clear evidence that a possible impact of right ventricular (RV) dysfunction on left ventricular (LV) function must be considered. Aim of this study was to evaluate the longitudinal systolic left ventricular (LV) – RV interaction in operated patients with Tetralogy of Fallot (TOF).
Methods: Biventricular measures of indexed ventricular end-diastolic volume (EDVi), ejection fraction (EF), LV longitudinal function parameters determined by magnetic resonance imaging (MRI) were investigated and compared to established normal z-score values.
Results: In our patients we found a good correlation between mitral annular plane systolic excursion (MAPSE) and LVEF values ($r = 0.788$; $p < 0.001$). While LVEF was normal in patients with mildly reduced right ventricular RVEF, the LVEF was decreased in patients with significantly reduced RVEF after 22 postoperative years. Patients with RVEDVi $\leq 150\,ml/m^2$ had a mean MAPSE of 1.43 ± 0.20 cm, and patients with RVEDVi $> 150\,ml/m^2$ a mean MAPSE of 1.30 ± 0.26 cm, the latter significantly reduced when compared to normal MAPSE z-score values. LV longitudinal function is decreased below the -2 SD of normal MAPSE z-score values 22 years after surgical repair in our TOF population. *Conclusions:* Our data show that simple M-mode measurement of the systolic LV function (i.e. MAPSE) is a sufficient surrogate for the LVEF. Therefore, when the endocardium is suboptimal for tracing, MAPSE data can be used for LV longitudinal systolic function investigations and reduce the need for expensive investigations such as cardiac MRI.

P-60
Clinical use of the Kid-Short Marfan Score (Kid-SMS) as an additional tool for proper diagnosis of Marfan syndrome in children
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Background: Due to age dependent manifestations diagnosis of Marfan syndrome (MFS) in children is sophisticated. Although revised Ghent Criteria (GC) is a major step forward, its utility in children is still restricted due to expensive and technically advanced diagnosis. As early diagnosis submits long-term benefits concerning prognosis, the need of appropriate diagnostic tool for risk stratification is justified. In 2012 we developed the Kid-Short Marfan Score (Kid-SMS) to improve and simplify diagnosis in young patients. This study verifies the benefit of this new risk score.
Methods: Overall 187 paediatric patients were subject to a standardized diagnostic programme. Frequency of diagnosis with the revised Ghent nosology, genetics and the Kid-SMS were analysed and compared in all patients.
Results: During the verification process the Kid-SMS identified more suspected patients with MFS compared to revised Ghent nosology and genetics alone. The developed Kid-SMS covered for more than 96% of our patients with MFS.
Conclusion: Whereas diagnosis of MFS is sophisticated, Kid-SMS is a useful tool for risk stratification of suspected children with MFS by easy executable diagnosis, especially for paediatricians and paediatric cardiologists. The clinical use may be recommended.

P-61
Clinical features and current management of ventricular septal defects (VSD): a Tunisian experience
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Introduction: Ventricular septal defects (VSD) are the congenital heart defects most frequently. Their isolated form represents more than 20% of congenital heart disease. The aim of our study is to analyze the epidemiological, clinical features of VSD and to assess the survival and outcome of affected babies after surgery in a Tunisian pediatric population.
Methods: It is a retrospective study of 30 patients admitted in the Pediatric department of Sahloul hospital from January 2005 through December 2011. Inclusion criteria were: isolated VSD diagnosed by echocardiography and hospitalized in the service.
Results: VSD accounted for 16% of the congenital heart diseases hospitalized in our department. The mean age at diagnosis was 4 months $\frac{1}{2}$. The consanguinity rate was 43,3%.
The primary clinical symptoms were dominated by failure to gain weight (53%), heart failure (37%) and recurrent wheezing and respiratory distress (33%). Eight children (26.6%) had Trisomy 21. Echocardiography showed that membranous defects were by far the most common type (63,3%). Pulmonary hypertension was already present at diagnosis in 63.3% of patients. Twenty one children (70%) received symptomatic medical treatment. Only 12 (40%) underwent surgery: 11 had a surgical closure of VSD and a single child had a pulmonary artery banding. Postoperative complications were: postoperative heart block, persistent pulmonary hypertension or residual VSD.
Conclusion: VSD still suffer from the delay of the diagnosis and the surgical treatment in our country. Unfortunately patients operated late have worse short and long-term outcome.

P-62
Comparison of semi-automatic border detection software with manual electronic calipers in the quantifi-cation of arterial layer thickness with very-high resolution ultrasound
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Background: Non-invasive very-high resolution vascular ultrasound (VHRU, 25–55 MHz) has recently been developed but images are currently analyzed with manual electronic calipers (EC). The aim was to compare the use of semi-automatic border detection software (AMS; Artery Measurement System) with EC in the analysis of arterial images obtained with transcutaneous VHRU.
Methods: 100 images from central elastic (common carotid) and peripheral muscular (brachial, radial, femoral, tibial) arteries were obtained on two separate days from 10 healthy subjects of different sizes and ages including both adults and children. AMS and EC were independently used to measure lumen dimension (LD) and intima-media thickness (IMT) for all arteries and intima-media-adventitia thickness (IMAT) for muscular arteries. Adventitia thickness (AT) was calculated as the difference between IMT and IMAT. The intra-, inter-, and test-retest variability for each measurement were assessed for both systems.
Results: No bias between AMS and EC was found. The intra and inter coefficients of variation (CV) for carotid LD (mean 5.60 mm) was lower with AMS compared with EC (0.4 vs. 1.9%, $p = 0.033$ and 1.9 vs. 4.1%, $p = 0.037$, respectively; $N = 20$) while no difference in IMT or in test-retest comparisons were found. No consistently significant differences in intra, inter or test-retest CVs were observed for muscular artery LD, IMT,

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

IMAT or AT between AMS and EC overall. The intra CV for AT (15.6 vs. 24.8, p = 0.011; mean 0.111 mm; N = 41) and inter CV for IMT (14.3 vs. 21.2, p = 0.001; mean 0.219 mm; N = 58) obtained with AMS in higher quality muscular artery images was lower compared with EC.
Conclusion: Minor, although statistically significant, differences in the precision of AMS and EC-systems may be found in the analysis of arterial images obtained with VHRU. The improved precision of AMS for AT and IMT in higher quality images may be explained by a decrease in the technical variability imposed by the observer.

P-63
Precision of semi-automatic border detection software for the quantification of arterial layer thickness with very-high resolution ultrasound
Sundholm J.K.M. (1), Gustavsson T. (2), Sarkola T. (1)
Children's Hospital, Helsinki University Central Hospital, Helsinki, Finland (1); Chalmers University of Technology, Gothenburg, Sweden (2)

Background: Non-invasive very-high resolution vascular ultrasound (VHRU, 25–55 MHz) has recently been developed but images are currently analyzed with manual electronic calipers. The aim was to evaluate the use of semi-automatic border detection software (AMS; Artery Measurement System) in the analysis of arterial layer thickness in images obtained with VHRU.
Methods: 100 images from central elastic (common carotid) and peripheral muscular (brachial, radial, femoral, tibial) arteries were obtained on two separate days from 10 healthy subjects of different sizes and ages including both adults and children. AMS was used to measure lumen dimension (LD) and intima-media thickness (IMT) for all arteries and intima-media-adventitia thickness (IMAT) for muscular arteries. Adventitia thickness (AT) was calculated as the difference between IMT and IMAT. The intra-, inter-, and test-retest variability were assessed for the different measurements.
Results: Intra, inter, and test-retest coefficients of variation (CV) were 0.4, 1.9, and 7.2% for carotid LD (mean 5.601 mm; N = 20) and 9.2, 13.6, and 14.8% for carotid IMT (mean 0.373 mm). Intra, inter, and test-retest CVs were between 2.3–4.1, 2.6–7.4, 7.9–13.5% for different muscular artery LD (mean 1.965–7.751 mm; N = 80), between 7.2–12.3, 5.8–13.5, 12.9–18.0% for muscular artery IMT (mean 0.143–0.325 mm), between 3.8–10.0, 7.8–12.9, 14.7–16.7% for muscular artery IMAT (0.225–0.564 mm), and between 9.4–28.8, 13.4–31.0, 23.4–32.5% for muscular artery AT (0.079–0.239 mm), respectively.
Conclusion: The quantification of arterial LD and wall layer thickness from images obtained with transcutaneous VHRU is reliable using semi-automatic border detection software.

P-64
Clinicopathological investigation on cardiomyopathy with special reference to fatal cardiomyopathy in childhood using pathophysiological parameters: The impact of noncompaction cardiomyopathy on pediatric cardiomyopathy
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Purpose: This study examines the demographic, clinical and histopathological features of fatal cardiomyopathy in childhood with special reference to its subtype and pathophysiologic parameters. Early diagnosis of cardiomyopathy enables the start of effective treatment with the purpose of better outcomes in this population. Noncompaction of the ventricular myocardium (NCVM) has gained increasing awareness and attention and become a widely recognized cardiomyopathy. The clinical spectrum of this complex pathology is highly varied. Accurate diagnosis of NCVM is clinically important as NCVM can present as sudden unexpected death. In this aspect, we try to make clear the prognostic factors in pediatric cardiomyopathy with special reference to fatal cardiomyopathy in childhood. Cinical symptoms were compared with baseline ECG, 2DE, MRI, biomarkers and endomyocardial biopsy (EMB).
Patients and method: Between 1990 and 2011, a total of 46 cardiomyopathy patients were enrolled, including 21 hypertrophic cardiomyopathy (HCM) and 25 dilated cardiomyopathy (DCM); 7 idiopathic DCM, 4 NCVM, 8 myocarditis, 6 tachycardia induced cardiomyopathy (TIC). Selected biochemical markers were high-sensitive CRP, myoglobin, Creatin Kinase MB, troponin T, heart-type fatty acid binding protein, ANP and BNP. Histopathology was evaluated with semiquantitative morphometry.
Results: Resuscitated sudden death occurred in 2 out of 7 idiopathic DCM and in 3 out of 4 NCVM and 7 out of 21 HCM. Ablation was 1 for TIC. Pacemaker implantation was one for HCM, ICD implantation was one for d-HCM. Histopathology on EMB showed abnormalities of inflammatory cell infiltration, vacuolar degeneraton, lysis of myofibrils and higher % fibrosis in cardiac death patients. Fatal cardiomyopathy was more frequent in NCVM with LQTc on baseline ECG.
Conclusions: Although clinical severity did not reveal statistic correlation with biochemical markers and histopathological severity, symptomatic patients showed raised concentration of biochemical markers, so biochemical markers might be one of the plausible predictors for the severity of myocardial damage and EMB may still be helpful to determine etiology in undiagnose cardiomyopathy. High incidence of sudden cardiac death was found in NCVN with long QTc of ECG. Long QTc could be predictive of a poor prognosis. Therefore, early therapeutic implications for this condition may lead to better outcomes in this population.

P-65
Familial non-syndromic thoracic aortic aneurysms and dissections – a life threatening disease. Identification of novel genetic mutations of the ACTA2- and MYH11-gen in two families
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Introduction: Aneurysms of the thoracic aorta (TAAD) occur sporadically in about 80% and familiar in about 20% of the cases. Non-syndromic familial thoracic aortic aneurysms and dissections (FTAAD) are characterized as TAAD in absence of clinical syndromes such as Marfan-, Loeys-Dietz- or vascular Ehlers-Danlos syndrome. FTAAD is a functionally heterogeneous disorder with autosomal dominant transmission. It is identified to date by mutations of TGFBR2 as well as of gens encoding structural proteins of the smooth muscle cells such as ACTA2

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

and MYH11. Mutations in the ACTA2 gen furthermore predispose to occlusive vascular disorders of the coronary and cerebral arteries as well as a bicuspid aortic valve and a PDA; extracardiac involvement may be present in the skin (livedo reticularis) and the eye (iris flocculi). Mutations of gen MYH11 are rare; in most patients a PDA is present.
Results: This report is on two families with an ACTA2- and a MYH11-mutation each, in which both a novel mutation has been identified. The ACTA2 family members are affected with PDA, dilation and aneurysm of the ascending and descending aorta including death because of dissection [missense mutation c.229A>T (p.lle77Phe) in exon 3 of the ACTA2 gen]. The MYH11 family members suffered from valvular aortic stenosis and dilation and dissection of the thoracic aorta, associated with death [heterozygote mutation c.4975.5T>C within the intron 35 of the MHY11 gen].
Conclusions: FTAAD disease is a rare hereditary disorder with increasing clinical importance because of its potential lethal outcome. Patients with dilation/ dissection of the aorta should be thoroughly screened for involvement of the coronary and cerebral arteries, a PDA, or ocular and cutaneous signs; investigation of first degree family members is mandatory and a genetic counselling is obligatory.

P-66
The value of investiogation of cardiac involvement in human immunodeficiency virus infection in children
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Introduction: Cardiac involvement in infection with human immunodeficiency virus (HIV) has particularly severe for morbidity and mortality in these patients and therefore requires early detection for appropriate treatment.
Objectives: The research of the main clinical aspects and diagnostic problems of cardiac involvement induced by human immunodeficiency virus infection (HIV infection/AIDS) in children.
Methods: Patients: 51 children, 2–16 years old, with HIV infection/AIDS in various stages of evolution, with diverse symptoms affecting many organs. Evaluation of patients: clinical, ECG, Chest X-ray, echocardiography (echo). Staging of HIV infection/AIDS by clinical exam and CD4 lymphocytes values.
Results: 60% of patients were included in group P2f clinical staging:. Signs of cardiac involvement: heart failure (11 cases), tachycardia (20 cases), deafness of the heart sounds ± gallop rhythm ± systolic murmur of mitral regurgitation (12 cases), dyspneea (14 cases), other non-symptomatic (14 cases) or with signs of others diseases. ECG : disturbances of ventricular repolarisation, sinusal tachycardia. RxCT: cardiomegaly (30% cases) ± aspects of pulmonary infections. Echo exam: cardiac involvement in 66% cases: dilated cardiomyopathy, the most severe changes (14 cases), pericarditis (10 cases), isolated dilation of the left and right ventricle (6 cases), LV diastolic dysfunctional (14 cases), pulmonary hypertension (6 cases). The severity and incidence of cardiac disease was associated with significant reduction of CD4 value < 400/mmc. Hystological exam performed in 28 patients died by pulmonary infections: aspects of myocarditis, pericardial and myocardial inflammatory infiltration, necrotic lesions.
Conclusion: The high incidence (66% of Cases) and severity of clinical manifestations, cardiac suffering during HIV infection/ AIDS is one of the most important problems of these patients. Cardiological evaluation of patients is necessary in all the stages of the infection, even non-symptomatic, for the diagnosis and follow-up of evolution. Echocardiography is the most sensitive noninvasive method useful for highlighting cardiac damage in these patients.

P-67
The Operative Outcome of Left and Right Atrial Isomerism: Single Centre Experience from Turkey
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Aim: In this article, we report outcomes of our institutional experience about the patients with left (LAI) and right atrial isomerism (RAI) during a 3-year period.
Patients and Methods: We studied the databases of the patients with LAI and RAI between January 2010–January 2013, retrospectively.

Characteristics	Right atrial isomerism (n = 16)	Left atrial isomerism (n = 12)
RVOT obstruction	81% (13/16)	83% (10/12)
Complete AVSD	75% (12/16)	58% (7/12)
MAPCA and/or PDA	63% (10/16)	16% (2/12)
DORV	38% (6/16)	50% (6/12)
Persistent superior vena cava	25% (4/16)	33% (4/12)
Dextrocardia	31% (5/16)	1
TAPVD	19% (3/16)	0
Ventriculoarterial discordance	13% (2/16)	25% (3/12)
Common atrium	0	17% (2/12)
PAPVD	1	17% (2/12)
LVOT obstruction	0	17% (2/12)

Surgical Operation Type	Patient number (n = 27)
Systemic-pulmonary shunt	13
Kawashima	6
Glenn shunt	6
Pulmonary banding	3
TAPVD repair	3
Fontan operation	1
VSD closure	1
Arterial switch	1
TOF total repair	1

Results: 28 patients were included the study (39% Female). The cardiac statuses and echocardiographic findings of patients are summarized in Table 1. Surgical approach could be done for 27 patients because one of the patients was inoperable. The characteristics of patients who underwent surgery for cardiac abnormalities are shown in Table 2. Biventricular repairs were possible for 3 patients. One/one and a half ventricle repairs were done for 12 patients. Other patients still waiting for one and a half ventricle repair. General survival rate was 77%(6/28). 37%(6/16) of the patients died; 2 because of Glenn failure, 1 because of intractable

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

heart failure and 2 because of sepsis and all of these were with RAI. One patient with RAI was died at home. Total early and midterm mortality was 23%. Mean follow up period was 12.6 ± 10.6 months (range, 5–28). In the follow up period; 1 patient received pacemaker implantation because of sick sinus syndrome, ventricular tachycardia and atrial fibrillation developed in one patient.
Conclusion: Atrial isomerism is one of the severe forms of the congenital heart defects. A mortality ratio in patients with RAI is higher than in patients with LAI. It seems that early and mid- term survival ratios can be improved with advanced surgical techniques and good postoperative intensive care conditions.

P-68
Right Ventricle Function in Obese Adolescents with or without Hypertension: Insights from Tissue Doppler Imaging
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Objective: The aim of this study was to present right ventricle echocardiographic parameters in asymptomatic adolescents with obesity and/or hypertension.
Materials and Methods: 93 obese (n = 36 obese-nonhypertensive and n = 57 obese-hypertensive; body mass index >30 kg/m^2) and 14 non-obese hypertensive (body mass index = 17–25 kg/m^2) adolescent along with 23 age-matched non-obese, non-hypertensive (body mass index = 17–25 kg/m^2) adolescent for the control group, were included in the study. Pulsed Doppler and tissue Doppler parameters were studied using transthoracic echocardiography for right ventricular function.
Results: There was significant difference in tricuspid valve (TV) myocardial performance index (MPI), TV isovolumic relaxation time (IVRT), and TV isovolumic contraction time (IVCT) when the hypertensive and normotensive cases in the non-obese group were compared. While there was significant difference in all tissue Doppler parameters between the obese and non-obese cases in the hypertensive group, there was also significant difference in TV E/A ratios and TV MPI parameters between the hypertensive and non-hypertensive cases in the non-obese group.
Conclusion: According to our findings, conventional and tissue Doppler echocardiography is useful when demonstrating the effects of obesity on the right ventricle, both in the presence and absence of accompanying hypertension, and echocardiographic indicators of right ventricule disfunction vary depending on whether obesity is unaccompanied or not.

P-69
Efficacy of Very Low Dose Prostaglandin E1 in Duct-Dependent Congenital Heart Disease
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Introduction and Aim: Prostaglandin infusion treatment is of vital importance for keeping the ductus arteriosus patency while waiting for either open surgery or invasive transcatheter intervention in duct-dependent congenital heart disease. Our study aimed at defining the lowest effective prostaglandin E1 dose in patients with inadequacy of both pulmonary blood flow and blood mixing (Group 1) and those with inadequate systemic blood flow (Group 2).
Methods: Patients referred to our center within two weeks from birth following the diagnosis of duct-dependent congenital heart disease and the start of intravenous prostaglandin E1 infusion were included in the study. Patients with inadequacy of both pulmonary blood flow and blood mixing (Group 1) and those with inadequate systemic blood flow (Group 2) were retrospectively evaluated in two separate groups with regard to the prostaglandin E1 starting dose given in the referring facility, the lowest and the highest dose administered in our center, treatment duration, adverse effects and administered treatment.
Results: Of the 95 patients considered in the study, 69 (72.6%) belonged to Group 1 and 26 (27.3%) to Group 2. No difference between the groups could be detected as to sex or birth weight ($p = 0.95$ and 0.42, respectively). A statistically significant difference could not be established for prostaglandin treatment duration, 9.73 ± 0.81 days in Group 1 and 11.6 ± 1.05 in Group 2 ($p = 0.064$). While the prostaglandin starting dose given to both groups in the referring facility was 0.067 ± 0.003 micrograms/kg/minute, it was reduced after titration to 0.039 ± 0.002 and 0.081 ± 0.005 micrograms/kg/minute, respectively, and this difference between the two groups was significant ($p < 0.001$). The dose administered to Group 1 while ductus patency was being maintained was 0.0031 ± 0.0001 compared to 0.0042 ± 0.005 micrograms/kg/minute for Group 2, also a statistically significant difference ($p < 0.001$). No adverse effects, including apnea, were observed during prostaglandin E1 infusion.
Conclusion: Our findings indicate that the infusion of prostaglandin E1 at a very low dose (0.003–0.005 micrograms/kg/minute) is sufficient to maintain the patency of the ductus arteriosus. A higher dose of prostaglandin E1 may be necessary in patients with inadequate systemic blood flow.

P-71
Our 10 years experience; Clinical and epidemiological characteristics of juvenile myocardial infarction
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Background: Elevated incidence of acute myocardial infarction (AMI) in young age and its correlation with growing population of obesity and hypertension in the age due to change of lifestyle have recently been reported. However, precise picture of the correlation is undetermined. In addition, involvement of Kawasaki disease, another increasing risk factor, in AMI is unknown.
Objectives: Backgrounds of early-onset AMI including history of Kawasaki disease and coronary risk factors were investigated.
Methods: Among 1687 patients with AMI who admitted in St. Marianna University School of Medicine hospital from January 2000 to December 2010, 22 patients under 40 year-old (1.3%) were inspected for obesity, hypertension, hyperlipidemia, smoking, diabetes and family history as well as a history of Kawasaki disease.
Results: Twenty cases (91%) of the early-onset AMI patients were male. Average number of coronary risk factors per patient was 3.09 ± 1.4, with following incidence: obesity (BMI > 25: 71%, BMI > 30: 38%), hypertension (55%), hyperlipidemia (73%) and diabetes (23%), smoking (55%), and family history of AMI (38%). Percutaneous coronary interventions were performed to 22% of

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

patients with double stenoses. As underlying diseases, collagen disease (2 patients), venous sinus thrombosis (1 patient), familiar hyperlipidemia (1 patient), primary aldosteronism (1 patient), dilated cardiomyopathy (1 patient) and suspicion of Kawasaki disease (2 patients) were identified.
Conclusions: The early-onset AMI in Japan is characterized as a male disease preceded by multiple untreated coronary risk factors. Kawasaki disease has less implication than expected. Our results strongly suggest that the risk factors need to be treated at an early stage.

P-72
Importance of NT-proBNP in monitoring acute rheumatic carditis

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Introduction: To detect the correlation of N-terminal Pro Brain natriuretic peptide (NT-ProBNP) levels with clinical and laboratory findings by measuring them at diagnosis, during and after treatment in children with acute rheumatic carditis.
Methods: A total of forty children including 20 acute rheumatic carditis patients aged between 5 and 16 years and 20 healthy children as controls were included in the study. Blood was drawn from patients at diagnosis and in the first week, first month and third month after treatment in order to detect proBNP, C-reactive protein levels and erythrocyte sedimentation rates. All patients underwent echocardiography.
Results: The NT-ProBNP levels of children with acute rheumatic carditis were significantly higher than those of the control group at diagnosis and in the first week of treatment ($p < 0.05$). Echocardiographic evaluation of acute rheumatic carditis patients revealed that left atrium diameter continued to decrease during the study and that mean left atrium diameters measured at diagnosis and in the 1st week were statistically higher than the mean left atrium diameters measured in the 3rd month. There was no correlation between left atrium diameters at diagnosis and in the 1st week, 1st month and 3rd month and NT-ProBNP levels during the same periods in the patient group.
Conclusions: Although increased serum NT-proBNP levels acted as a marker of cardiac inflammation in patients with acute rheumatic carditis in this study, this increase was not correlated with enlargement in the left atrium. NT-proBNP levels were found to be a valuable determinant indicating cardiac inflammation and hemodynamics.

P-73
Clinical Characteristics and Incidence of Acute Rheumatic Fever in Kayseri, Turkey, A Retrospective analysis of 624 patients between 1998 and 2011

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Introduction: Determine the incidence and clinical characteristics of acute rheumatic fever (ARF) in Central Anatolia within the past 14 years and make a comparison of two periods of seven years.
Methods: We made a retrospective analysis of 624 patients who were diagnosed with ARF at the Department of Pediatric Cardiology in the Medical Faculty of Erciyes University between January 1998 and December 2011.
Results: The mean age of patients was 10.9 ± 2.7 (2.5–18) years, with 60% ranging from 10 to 14 years of age. The female/male ratio was 1.4. The estimated incidence rate of ARF was 7/100000/year, and estimated rheumatic heart disease (RHD) prevalence was 60/100,000 in Central Anatolia.

	1998–2004 [295 patients (47%)]	2005–2011 [329 patients (53%)]	Statistical difference ($p < 0.05$)
Mean age	11.0 ± 2.7	10.6 ± 2.9	-
Gender (F/M ratio)	1.41	1.35	-
Major manifestations			
Carditis	56.6%	52.2%	-
Arthritis	36.6%	33.4%	-
Chorea	27.4%	23.1%	-
Subcutaneous nodules	1%	0%	-
Erythema marginatum	0%	0.3%	-
Minor manifestations			
Fever	19.6%	15.5%	+
Arthralgia	35.2%	32.5%	-
Elevated CRP	48.2%	41.1%	-
Elevated ESR	62.4%	57.2%	-
Prolonged PR	21.5	13.5	+
Elevated ASO	80.7%	82.4%	-

The most common presenting complaint was arthralgia (34%). Among the major findings, the most common included carditis at 54%, arthritis at 35%, Sydenham's chorea at 25% and subcutaneous nodules at 0.5%, respectively. The valve that was most commonly affected was the mitral valve alone 63%, followed by combined aortic valve and mitral valve 32%, and the aortic valve alone 5%, respectively. Although the number of patients diagnosed with ARF was 1.3 times more in the second 7-year period than in the first 7- year period and incidence of ARF had decreased in last 3 years.
Conclusions: Although there has been socioeconomic development in Turkey during recent years, the incidence of ARF is still high in Central Anatolia. A national program is needed for the prevention of this disease and the taking of relevant precautions.

P-74
Two Hennekam Syndrome Cases Presenting with Massive Pericardial Effusion and Hydrops

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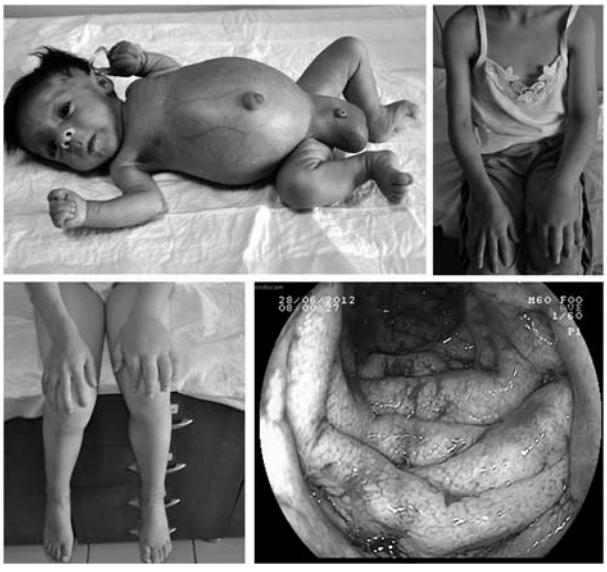
Introduction: Hennekam syndrome is a rare autosomal recessive syndrome first described in 1989, characterized by congenital lymphedema of the limbs, genitalia, and face, intestinal lymphangiectasia, mild growth retardation, variable mental retardation, seizures and craniofacial dysmorphic features.
Case one: At birth the patient presented with severe respiratory distress due to non-immune hydrops. Generalized edema was seen especially prominent in his genitals and periorbital region. After resolution of the hydrops fetalis his facial features became apparent: flat face, prominent forehad, somewhat down-slanted palpebral fissures, epicanthal folds, hypertelorism, a broad, depressed nasal bridge, a bulbous nasal tip, small mouth, low-set

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

ears, micrognatia and high-arched palate. Bilateral pleural effusion was seen in his chest X-ray and massive ascite in abdominal ultrasonography. Repeated examination of fecal excretion of alpha-1 antitrypsin showed positive, suggesting intestinal lymphangiectasia. Repeated intravenous supplementation of albumin has given according to his albumin level. His symptoms get better with a special fat-free diet with added medium chain triglycerides and fat soluble vitamins but ascites continued.
Case two: 13 years girl patient applied to emergency with chest pain, edema in her limb appearance in left side. Previous doctors told that lymphedema in her left side of the body could be due to allergy. She had used multiple ointments, attacks of diarrehea continued for one or two days but resolved spontaneously. Dysmorhic features were: small mouth, thin limb, low-set ears and long philtrum. There was left hemifacial, left sided upper and lower extremity edema. She had normal mental development. Massive pericardial effusion was seen in echocardiography. Radionuclide lymphoscintigraphy scan showed abnormal drainage of the lower and upper (significantly left side) limb. Gastroduodenoscopy showed snowflake appearance of the duodenum. Intestinal biopsy revealed lymphangiectasia. His symptoms improved with a special fat-free diet with added medium chain triglycerides and fat soluble vitamins.
Conlusion: Massive pericardial effusion and non-immun hydrops were rare clinical presentation of Hennekam syndrome. In addition to 31 cases in literature; these two cases cause of massive edema even as fetal hydrops were presented in order to take attention to this rare autosomal recessive disorder.



P-75
Assessment Of Left Ventricular Functions By Strain And Strain Rate Echocardiography In Children With Type I Diabetes Mellitus
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Introduction: Left ventricular (LV) dysfunction can occur in children with type I diabetes mellitus (DM). We evaluated regional LV myocardial functions by strain and strain rate echocardiography in children with type I DM
Methods: 101 type I DM patients (49 girls, 52 boys; mean age 14.3 ± 3.3 years) and, 32 healthy controls (13 girls, 19 boys; mean age 13.6 ± 2.6 years) were studied. Patients were divided into 3 groups according to the duration time of diabetes (group 1: <5 years, group 2: 5–10 years and group 3: ≥10 years). LV functions were evaluated by conventional echo, tissue Doppler, and 2D strain and strain rate echocardiography.
Results: The mean duration of diabetes was 3.1 ± 1.1 years, 6.5 ± 1.3 years and 12.2 ± 1.7 years in group 1, 2 and 3, respectively. No significant difference was found between the groups in age, sex, height, weight, body mass index, body surface area, systolic blood pressures. Heart rate was significantly high (group 1: 83 ± 13, group 2: 84 ± 14, group 3: 92 ± 17 beats/min, control group: 75 ± 10 beats/min; $p = 0.058$, $p = 0.020$ and $p = <0.001$, respectively) in the patients groups. Ejection fraction and shortening fractional were similar in all groups. Mitral A velocity was significantly different in the group 3 compared to controls (0.67 ± 0.13 vs. 0.55 ± 0.11 cm/sec, $p < 0.001$). Deceleration time was significantly increased (144 ± 31 vs. 116 ± 33 msn; $p = 0.001$) in group 2 compared to control group. Tei index was significantly decreased (0.35 ± 0.06 vs. 0.41 ± 0.11; $p = 0.0.05$) in the group 2. The global average of longitudinal peak systolic train values was not statistically different between all groups. Longitudinal and circumferential strain and strain rate values was significantly increased in the different regions of the left ventricle in both group 2 and group 3 compared to control group ($p < 0.05$). There was not any correlation between the global longitudinal strain values and both HbA1c values and the duration time of diabetes.
Conclusion: 2D strain echocardiography is a useful tool to detect the effects of diabetes on the heart, especially in which those conventional methods cannot present a more detailed analysis on regional and global myocardial function.

P-76
Small ventricular septal defect considered not requiring surgical closure; can we wait to develop complications?
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Objectives: The purpose of the study was to assess the outcome of patients with small ventricular septal defects (VSDs) considered not to require surgical closure during childhood.
Methods: A descriptive study was conducted on children undergoing echocardiography from January 2008 to December 2011 at Children Hospital and Institute of Child health, Lahore. The data with isolated VSD considered too small to require surgery during childhood as defined by gradient across VSD more than 50 mm of Hg, normal LVEDd according to age, size less than 3 mm, no PR and asymptomatic, on all children below 15 years was reviewed. The data was analyzed with SPSS 16 version.
Results: The total of 883 patients of restrictive VSDs considered not to require surgery, 60.6% (n = 535) were males and 39.4% (n = 348) were females. The significant number of patients i.e. 18.7% (n = 166) developed complications. Aortic cusp prolapse developed in 13.6% (n = 120) p = .001, 2.3% developed aortic regurgitation secondary to aortic cusp prolapsed. 1.8% developed right ventricular track outflow obstruction (RVOT) p = .021 and 0.3% of patient developed left ventricular outflow track obstruction (LVOT) p = .018. Similarly 0.8% patients developed endocarditis. Regarding types of VSD, we found Perimembranous in 65.8%, muscular in 12.6%, Subaortic in 8.3%, doubly committed in 6.0%, Inlet in 5% and outlet in 1.7% of patients.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Conclusions: Although patients with small VSDs have generally been considered not to require surgery, data suggests that a significant percentage of these patients developed complications later in their life i.e., 18.7%

Keywords: Ventricular septal defect, Aortic cusp prolapse, Aortic regurgitation, Right ventricular outflow track obstruction, Left ventricular outflow track obstruction.

P-77

Spironolactone versus Amiloride: Time for change?

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Introduction (or Basis or Objectives): Potassium sparing diuretics are used frequently in paediatric cardiac patients to supplement loop diuretics and minimise hypokalemia. Diuretics in infants and children are largely used in the context of 'heart failure' due to pulmonary over circulation from structural left to right shunts, and to a lesser extent for myocardial dysfunction. Spironolactone is the drug used at East Midland's Congenital Heart Centre (EMCHC). Liquid Spironolactone is used at a dose of 1–2 mg/kg/day and is expensive (at 50 mg in 5 mls, a 125 ml bottle must be thrown away after a month, and costs £60). Amiloride is an alternative potassium sparing diuretic & is used at some other children's heart units at 0.2 mg/kg/day. A 150 ml bottle of Amiloride 5 mg/5 ml costs £45 and can be kept for 12 months. This study was aimed to identify which potassium sparing diuretics are being used at the various paediatric cardiac units in UK and also the evidence base for use of either of these agents.

Centre	K sparing diuretic
Birmingham	Amiloride
Bristol	Spironolactone
Glenfield	Spironolactone
GOS	Amiloride
Guys	Spironolactone
Leeds	Spironolactone
Liverpool	Spironolactone
Newcastle	Spironolactone
Royal Brompton	Spironolactone
Southampton	Spironolactone

Methods: In November 2012 we surveyed all paediatric cardiac centres in UK to enquire about current practice of prescribing potassium sparing diuretics in children. We also did a database search looking at studies comparing Amiloride & Spironolactone using OVID interface for Pubmed & Medline.

Results: Telephone survey of all ten English Paediatric Cardiac Surgical units revealed that only two centres are currently using Amiloride as the preferred potassium sparing diuretic. The rest of the units are using spironolactone (table below).

Discussion: Literature search found no paediatric studies comparing both the drugs. 15 adult studies were found of which only 6 studies were partially relevant. Overall both the agents appear comparable with no statistically significant differences in reduction of potassium excretion. Spironolactone has been shown to be of additional benefit in long term treatment of heart failure due to myocardial dysfunction but there is no evidence this is relevant to its short term use for left to right shunts.

Conclusions: Amiloride is a useful & cheaper alternative instead of spironolactone. More paediatric studies are needed to compare both these drugs in children with congenital heart diseases.

P-78

Measurement of ultrasensitive Troponin T levels in cord blood for early detection of myocardial cell damage after birth

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Background: Myocardial cell damage after neonatal asphyxia is underestimated. Indeed, its occurrence has not been systematically investigated that far.

With this respect, the possibility to detect it early after birth by measuring blood levels of markers of organ injury would allow early treatment and improvement of patient outcome.

This prospective study was designed to collect normal values and analyse levels of the new markers of tissue damage and inflammation that are the ultrasensitive (u) cardiac Troponine T (TnT) and copeptine, Brain Natriuretic Peptide (BNP) and u C-reactive Protein (CRP), respectively, in neonates.

Methods: In 60 consecutive mature neonates (mean age: 39 gestational weeks) cord blood levels of uTnT, Copeptine, BNP and uCRP were measured.

Data were analysed with respect to mode of birth, presence of dystocia and were related to the Apgar score given at 5 and 10 minutes.

Results: uTnT levels averaged 35 ± 18 ng/L, Copeptine $483{,}5 \pm 669{,}9$ pmol/L, BNP $706{,}8 \pm 447{,}6$ ng/L and uCRP $0{,}064 \pm 0{,}11$ mg/L, respectively.

There was no influence of gestational age on marker levels.

There was no difference between neonates born by vaginal delivery or by caesarean section.

In contrast, the 7 neonates born after dystocic delivery showed higher values of uTnT than neonates born after uncomplicated delivery: 44 ± 22 versus 30 ± 19 ($p < 0.1$).

uTnT values correlated with the Apgar score calculated at 5 minutes ($P < 0.1$) and with BNP values ($p < 0.02$).

Blood concentrations of Copeptine, BNP and uCRP were not influenced by dystocia demivery.

Conclusions: The results of this study suggest that measuring cord blood concentrations of uTnT might allow detecting neonates with myocardial cell damage due to intrapartal asphyxia.

The relationship between uTnT- and BNP levels we report here suggests that uTnT elevation is the expression of a myocardial damage that is functionally relevant.

We expect that early recognition of neonates with subclinical myocardial cell injury should lead to early protective strategies in order to improve post-natal outcome.

P-79

Merits and complications of the vertical vein in superior total anomalous pulmonary venous connection. A transcatheter closure series of 5 further cases

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The vertical vein (VV) is frequently left non ligated after supracardiac TAPVC rechanneling, to avoid hemodynamic

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

complications due to small left heart especially in case of obstructed connection.
Objective: We report here one centre experience in TAPVC rechanneling, and follow up of the VV.
Patients and methods: We have done a retrospective study to illustrate the outcome of supracardiac TAPVC since 1998. VV follow up was performed by colour-Doppler echocardiography and CT scan. Transcatheter closure of VV was performed under local anaesthesia.
Results: We had a total of 11 patients (7 males and 4 females). Diagnosis was done between the age of 1 and 120 days, all of them had undergone rechanneling. VV was not ligated in 4 out of 5 patients with obstructed connection, versus 3 out of 6 patients without obstruction. VV patency had no immediate post operative benefits in non obstructed TAPVC. On the opposite in obstructed connection, the ligation was associated with longer ICU stay duration as observed in our sole patient (33 days).
During follow-up, VV remained patent in 6 of 7 patients and required transcatheter closure in 5 patients because of significant left to right shunt. The shunt was negligible in the remaining patient.
The closure was successfully performed without complications with Amplazer Vascular Plug (type 2) at a median age of 4 years and 8 months. The device diameter was chosen up to 1.5 fold of the diameter of VV. The radiation doses ranged between 2 to 17 $Gycm^2$. After closure, VV occlusion was confirmed in all patients.
Conclusion: Patent VV in non obstructed TAPVC does not change post operative course. Most non ligated VVs remain patent. Transcatheter occlusion with Amplazer Vascular Plug is a safe and effective procedure to suppress the left to right shunt.

P-80
"Headache with an interruption"
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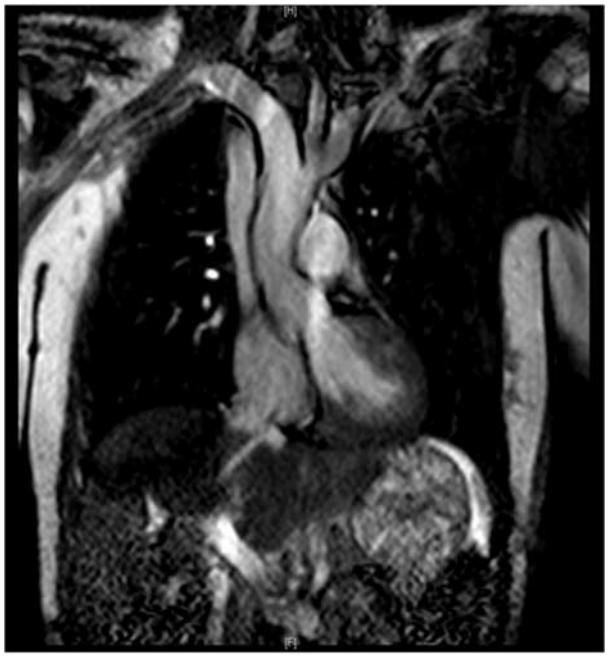
Introduction: We present an interesting case of a 14 year old girl with history of intermittent headaches and "feeling tired all the time".
Case history: Patient was referred for a cardiology consultation in view of pulsations noted at the base of the neck on the right side, intermittent episodes of headaches from age of 8 years (diagnosed as migraine) and always feeling tired with lack of energy.
In the past during hospital visits doctors had commented that it was difficult to feel her left brachial and radial pulses. This was also noticed during a biology class in school on the circulatory system but no further investigations were done. There was no family history of congenital heart disease.
On examination of her pulses the right brachial and radial pulses were well felt along with both her carotids. It was not possible to feel her left brachial or radial pulse. The femoral pulses were bilaterally palpable. Her heart rate and saturations were normal. Blood pressure measurement showed she was hypertensive in her right arm (160 mm Hg systolic), left arm (100 mm Hg systolic). On cardiovascular examination she had normal heart sounds and no murmurs. The rest of the examination was unremarkable.
The following investigations were performed

- CXR — Rib notching on right side with prominent soft tissue shadow
- ECG — sinus rhythm, normal axis and rate with no abnormality
- Echocardiogram — difficult to visualise entire aortic arch with interruption after the left common carotid
- Cardiac MRI — Normal intra-cardiac anatomy. 4 cm interruption of aortic arch between left common carotid and left subclavian artery
- MRI Brain with MRA — Multiple collaterals with descending aorta supplied by these and also by retrograde flow through left vertebral artery through circle of Willis

The patient underwent single stage surgery and has done well on follow up.
Conclusions: Interrupted aortic arch is a rare congenital malformation that occurs in 3 per million live births. The presentation in teenage/adulthood is even rarer but is well recognised. Clinicians dealing with children must consider this in their differential diagnosis in someone with headaches, hypertension and weak/absent pulses.



P-81
Normal values of atrioventricular valves diameters in neonates
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Background: Normal values of various cardiovascular diameters are needed for facilitate proper diagnosis and decision about treatment of children with congenital heart defects. The larger is analyzed group, the conclusions and received norms are more accurate and helpful.
The aim of this study was to assess the normal values of tricuspid and mitral valves in healthy, term neonates and to correlate this with age (1–30 days), weight and body surface area.
Material and Methods: 771 transthoracic echocardiographic examinations performed between years 2002–2008 were reviewed and data were collected retrospectively. All examinations were performed in healthy, term neonates without any structural heart defect. Measurements were taken from four chamber axis view in diastole. Statistical analysis was performed using Statistica 10 software.
Results: The average age of neonates at echocardiographic examination was 10,6 days (SD −7,8), weight 2,6 kg (SD −0,7).

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Mitral valve diameter - average 9,2 mm; SD-1,45; 95%CI 9,1-9,4 – was statistically significant correlated with body surface area (BSA) ($r = 0,18$; $p < 0.001$) and weight ($r = 0,2$; $p < 0.001$). Tricuspid valve the observed diameters - average 9,9 mm; SD -1,74; 95%CI 9,8–10,1 – was also correlated significantly with BSA ($r = 0,19$; $p < 0.001$) and weight ($r = 0,24$; $p < 0.001$). All significant corelations were week, however correlations with weight were in each case stronger than with BSA. We calculated the normal values of mitral (7,3–11,6 mm) and tricuspid (7,5–13 mm) valves for our population contains values between 5 and 95 percentile.

Conclusion: The diameters of mitral and tricuspid valves in neonatal period doesn't have strong correlation with age, weight or BSA, despite the fact, that it is statistically significant correlation in case of BSA and weight. It allows to use the normal values range for whole neonatal period. In case of borderline values, the normal values calculated for weight should be taken under consideration, because it has the strongest correlation with atrioventricular valves diameters.

P-82

Implications of new recommendations about Sildenafil in children with PAH. Is reduction of Sildenafil to recommended doses safe?

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Introduction: Sildenafil has been used off-label for treatment of PAH in the paediatric population, however data on dosing has been limited. A recent extension to a randomised control trial of sildenafil in children with PAH (STARTS – 2) reported an unexplained excess mortality in patients randomized to higher doses. Consequently the European Medicines Agency (EMA) recommended children weighing 8–20 kg should be treated with 10 mg three times a day and patients weighing 20 kg or more should be treated with 20 mg three times a day. We sought to assess the clinical impact and safety of reduction of Sildenafil to the recommended doses.

Methods: Twenty-three patients (11 females) with median age 9.2 (range 1.2–15.4) years were included. PAH was idiopathic in 7, associated with lung disease in 9, associated with congenital heart disease in 5. Ten patients were on monotherapy with Sildenafil and 13 patients were combination therapy. Nine patients weighed <20 kg requiring mean reduction of 18.7 mg (range 10–45) sildenafil per day; 14 patients weighed ? 20 kg requiring mean reduction of 43 mg (range 20–100). Patients were assessed prior to reduction of dose and after reduction with median time interval of 13 weeks (IQR 10–17).

Results: No additional PH treatment was required during the weaning period. Following dose reduction 3 patients improved functional class, 2 worsened, there was no change in functional class in the remainder. No patient was in functional class IV and there were no episodes of syncope.

There was no significant fall in six-minute walk distance (mean(SD) baseline distance 414(56)m, at follow-up 412(40)m; mean change −2.5(92), $p = 0.88$).

On MRI ($n = 5$) there was no significant change between baseline and follow-up in RVEDV (mean RVEDV = 77 ml/m^2 and 83.6 ml/m^2 respectively, p-value = 0.58), RVESV (mean RVESV = 32.8 ml/m^2 and 39 ml/m^2 respectively, p-value = 0.0579) or RVEF (mean RVEF 56.8% and 53.6% respectively, p-value = 0.1).

Conclusion: These early data may support physicians trying to implement the new dosing recommendations into clinical practise. However, close monitoring for clinical deterioration remains paramount for this progressive disease.

P-83

Cytomegalovirus post paediatric heart transplantation: a prospective study

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CMV remains a major infectious complication after transplant. In paediatric heart transplantation it is implicated in accelerated coronary vasculopathy, which may be accentuated by lack of substantial CMV specific immunity.

We designed a prospective study of CMV in paediatric cardiac transplantation. CMV-specific immunity was investigated in relationship to CMV viral load. Cell-mediated immune response and the presence of soluble markers of inflammation were determined in blood samples of heart-transplanted children at regular intervals up to one year.

30 recent transplant recipients were enrolled prospectively. The presence of CMV-specific T cells and their ability to produce inflammatory cytokines was determined in peripheral blood mononuclear cells isolated from whole blood. Soluble markers of inflammation were measured in plasma, including CX3CL1 (fractalkine), an atypical chemokine relevant to the pathology of atherosclerosis and other vascular diseases.

CD8+T cells from children who had viremia within 12 weeks after transplant, were able to respond to viral stimulation between 16–32 weeks post-transplant. Lymphocytes from patients who had been immunologically primed by CMV following natural infection, but did not show reactivation/reinfection, responded to CMV stimulus earlier. The CMV specific responses were similar in those groups exposed to the virus at one year post-transplant.

From week 16 post-transplant, the levels of CD57+ CD8-T cells were increased particularly in the viremic group, where this cell population maintained effective cytotoxic potential, producing increasing amounts of Granzyme B. Furthermore, both the CMV seropositive and the viremic cohort showed augmented plasma levels of CX3CL1 when compared to the CMV negative group at the same time point. Titres remained high throughout the one year study period.

Our data indicate that T-cell immunity can be used to monitor the level of immunosuppression and to modulate the outcome of CMV infection, thus reducing the risk for long-term complications post-transplant. Elevated levels of CX3CL1 in the CMV exposed patients suggest that this chemokine has a central role in the development of cardiac allograft vasculopathy.

P-84

Pulmonary artery reintervention following the arterial switch operation for transposition of the great arteries: a single centre experience

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Background: To determine rates of pulmonary artery (PA) reintervention following arterial switch operation (ASO) for transposition of the great arteries (TGA) and to identify factors associated with reintervention.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Methods: Retrospective data collection via an electronic database and individual patient records.
Results: 178 patients underwent ASO for TGA between 2005–2011. Fifty-two patients followed-up overseas were excluded from further analysis. Of the remaining 126 patients, ASO was performed at a median age of 9 (2–2067) days with median weight 3.6 (2.4–17.6)kg.
Simple TGA (including muscular VSD) accounted for 109/126 (87%) patients. Fourteen (11%) underwent palliative procedures (PA banding ± modified BT shunt (mBTS)) before ASO. Over 98% of patients underwent a Lecompte manoeuvre, whilst 43% had primary PA augmentation during ASO.
Over a median follow-up of 4.0 (0.7–7.7) years, 4 patients died, whilst 12/126 (9%) required reintervention for PA stenosis, half of whom required multiple reinterventions. Those undergoing reintervention all underwent Lecompte manoeuvres and 7/12 (58%) had PA augmentation at ASO. Only 1/12 had a preceding palliative procedure.
First reintervention following ASO occurred at a median interval of 16 (5–216) weeks; 11/12 (92%) reinterventions were within the first postoperative year. First reintervention was via transcatheter balloon angioplasty in 7/12 (58%) patients, of whom, only 2/7 (29%) have required no further intervention. Comparatively, 4/5 (80%) undergoing initial surgical reintervention remain free from subsequent reintervention.
Conclusion: In our experience, PA stenosis requires reintervention in 9% of patients following ASO, most frequently within the first postoperative year. Need for reintervention is not significantly influenced by pre-emptive PA augmentation at initial ASO, preceding palliative procedures involving the PAs, or age at ASO when adjusted for weight. Balloon angioplasty is successful in some patients, though the majority require future surgical reintervention.

P-85
Aerosolized iloprost and oxygen for assessment of pulmonary vasoreactivity in children with pulmonary hypertension

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Background: The evaluation of pulmonary vascular reactivity plays a significant role in the management of patients with pulmonary hypertension. Inhaled nitric oxide in combination with oxygen has become widely used as an agent for pulmonary vasodilator testing. However, inhaled nitric oxide is not available in many developing countries. Recently, aerosolized iloprost was suggested as an alternative to nitric oxide for this purpose.
The present study employed aerosolized iloprost together with oxygen to identify pulmonary vasoreactivity in children with pulmonary hypertension and made use of the synergistic effect of both vasodilators.
Methods: The study registered a total of 16 children whose mean age was 5.06 ± 3.88 with severe pulmonary hypertension. Hemodynamic parameters were quantified before and after the vasoreactivity test. At catheterization, patients who were found to have either $PVR > 6\,WU\,m^2$ or $Rp/Rs > 0.3$ received aerosolized iloprost (Ilomedine^R; Schering AG, Berlin, Germany). Aerosolized iloprost was administered at a dose of $25\,ng\,kg^{-1}\,min^{-1}$ diluted in 1.5 ml of isotonic saline solution and nebulized for 10 minutes with O_2 through face mask to achieve alveolar deposition of the drug. Increased left-to-right shunt, pulmonary vasculer resistance being <6 $WU.m^2$ and pulmonary-systemic resistance ratio being <0.3, as well as a decrease $>10\%$ in the pulmonary vascular resistance and pulmonary-systemic vascular resistance ratio after the vasoreactivity test were accepted as a positive response.
Results: Eleven children gave a positive response to the vasoreactivity test, while 5 children did not respond. Pulmonary vascular resistance dropped from $9.98 \pm 1.39\,WU.m^2$ to $5.08 \pm 1.05\,WU.m^2$ ($p = 0.013$) and the pulmonary-systemic vascular resistance ratio fell from 0.68 ± 0.08 to 0.32 ± 0.05 ($p = 0.003$) in the children who were responsive.
Discussion: Administration of inhaled iloprost in combination with oxygen for pulmonary vasoreactivity testing can be useful to correctly identify pulmonary vasoreactivity without extending the duration of cardiac catheterization.

P-86
Ambulatory blood pressure monitoring: Importance for determination of white coat hypertension in children

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Introduction: Identification and treatment of hypertension should be an important focus of physicians caring for children. White coat hypertension is defined as BP levels that are the 95th percentile or higher when measured in the physician's office or clinic but are completely normal average BP 90th percentile outside of a clinical setting. Although intensively discussed and finally accepted in recent years, white coat hypertension (wch) still raises the question of the benefit of diagnosing it in pediatric patients as its possible link with the subsequent essential hypertension is not clear yet.
The aim of the study: Selected use of ambulatory blood pressure monitoring to identify children with white coat hypertension, thus avoiding unnecessary diagnostic testing and treatment of these children.

Dg.	n	SBP	t-test	DBP	t-test
HtS	92	121	$p < 0.001$	70	$p < 0.001$
WCH	68	110	$p < 0.001$	66	$p < 0.001$

BP indicates blood pressure, SBP - systolic blood pressure, DBP - diastolic blood pressure, MAP - mean arterial blood pressure.

Methods: 160 children (aged 12 to 19 years) participated in the study. Based on office systolic and diastolic blood pressure measurements and 24-hour ambulatory blood pressure monitoring, subjects were placed into one of two groups: first, normotensive, and second, white coat hypertensive.
Results: Forty three percent (68 children) of 160 subjects with systolic or diastolic blood pressures greater than or equal to 95th percentile were reclassified as white coat hypertension; 57% remained hypertensive. The ambulatory blood pressure monitoring patterns of white coat hypertensive patients were significantly different from those of hypertensive patients. The mean values of systolic, diastolic and mean arterial pressure were significantly higher in children with essential hypertension than in the group with white coat hypertensionä tab.1).
Conclusions: This study documented the existence of white coat hypertensive children and showed that white coat hypertensive children were significantly different from hypertensive children in comparisons of 24-hour ambulatory blood pressure monitoring data.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

P-87

Use of Sildenafil in Pulmonary Hypertension associated with Bronchopulmonary Dysplasia in Premature Infants. Single Center Experience

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Pulmonary hypertension (PH) is an emerging potentially lethal complication of bronchopulmonary dysplasia (BPD) of prematurity (O2 dependency at 36 weeks GA), Its incidence is unknown and treatment is not yet standardized. We present retrospectively the experience with sildenafil (S) in a single large Center.

We analyzed all very low birth weight (VLBW) infants admitted at our Neonatal Intensive Care Unit (NICU) between 01/2007 and 12/2011 who developed BPD and PH and were treated with S for PH defined as systolic PA pressure (PAP) above 40 mmHg by Echo (mean >25 mmHg). S was discontinued when PAP was normal at 2 subsequent evaluations. Safety was assessed by adverse events while on S and the discontinuation of treatment for reasons other than improvement. Data were analyzed with SPSSv 18. Continuous variables are expressed as mean ± standard deviation and dichotomic variables as number (percentage).

Between 2007 and 2011, 649 VLBW infants were admitted to our center. Mean gestational age (GA) and birth weight were 29 ± 2.8 weeks and 1136 ± 274 grams respectively. 45 patients (pts) died before 36 weeks GA, Of the 605 survivors, 87 (14,2%) developed BPD, severe in 51 (8,4%). PH was detected in 14/51 (27%) pts with severe BPD (14% of all BPD), and they were treated with S (2.5 ± 1.25 mg/kg/day) Mean GA at diagnosis was 46 ± 9.7 weeks and mean systolic PAP 53 ± 16 mmHg. Three pts (21%) died before discharge because of chronic respiratory failure and PH. Three patients (14.3%) recovered for PH during hospital stay and discontinued S before discharge. Eight patients (57%) were discharged on S and O2 therapy and monitored as outpatients. Sildenafil was successfully weaned by 5.2 ± 4.7 months of corrected age in 5 patients. Three patients are still under treatment age 18, 22 and 39 months.

There were no adverse effects related to S treatment PH is emerging as a serious common problem in pts with severe BPD of prematurity Chronic use of S seem to be safe in these pts, although larger series will be needed to establish this firmly.

P-88

Regular assessment of the quality of life in a therapeutic education program: example of INR self-monitoring among children

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Introduction: Children under vitamin K antagonist (VKA) should benefit from self-monitoring of their International Normalized Ratio (INR). In France, since the 2008 Public Health Law, children and/or their family who wish to participate to INR self-monitoring must integrate a formalized authorized education program to anticoagulation therapy led by a pediatric cardiologist. Our national reference center for congenital heart diseases, while building this program, aimed to regularly evaluate the quality of life (QoL) of children.

Methods: All children and parents participating to our INR self-monitoring education program were invited to complete a QoL questionnaire during each group session. Generic pediatric QoL questionnaires were used (QUALIN for infants <2 years old, PedsQL for children aged 2 to 18). Both parents independently participated. PedsQL Child self-report QoL questionnaires were used for children above 5 years, under trained nurse supervising. This study received the approval of the Ethics Committee. Relations between QoL and patients' characteristics were studied.

Results: 111 children (54 girls) participated to our INR self-monitoring program between 2010 and 2012. Indications for VKA were classical within pediatric population: valve replacement (n = 47), total cavo-pulmonary connexion (n = 33), dilated cardiomyopathy (n = 13), Kawasaki disease (n = 8), others (n = 10). No family refused to be enrolled in this study. 476 QoL questionnaires (27 QUALIN, 449 PedsQL) were completed by 265 different persons (80 children, 107 mothers, 78 fathers), depending on the number of group sessions for each family (1 to 3). There were no significant relationships between QoL and patient's sex, type of AVK (warfarin or fluindione), number of group sessions, chronic illness duration or moment of diagnosis (prenatal or postnatal). Qol scores were significantly lower among children with congenital heart disease. Fathers and mothers' QoL scoring are rather well correlated but are significantly lower than their child's self- assessment.

Conclusion: Routine QoL assessment well applies to education programs with strong joining of families and children. Our center leads 5 official education programs (pulmonary hypertension, anticoagulation, transition to adulthood, pacemaker-Defibrillator and chronic cardiac failure) and aims in further studies to compare QoL of children participating to such programs to those who don't.

P-89

Changes in Longitudinal Myocardial Contractility and Electromechanical Interval during the First Month of Life in Healthy Neonates

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Background: During the neonatal period, the heart is subjected to significant hemodynamic changes. This study aims at documenting the changes in ventricular tissue velocities, longitudinal strain and the electromechanical coupling during the first month of life.

Methods and Results: We prospectively studied the ventricular performance of 16 healthy neonates at three time-points over the first month of life: at day 3, week 2 and week 4 of life. When measured at the mitral valve annulus, the tricuspid valve annulus and the interventricular septum, we found both right and left ventricular systolic tissue velocities to increase significantly by 21–28% between subsequent visits ($P < 0.001$). Early and late diastolic velocities at the same sites increased between the first and third visit by 1.5–1.7 times ($P < 0.001$) and 1.4–1.7 times ($P < 0.001$), respectively. Tissue velocity E/A ratio did not change significantly during the first month of life. Similar to the increase in systolic tissue velocities, peak systolic longitudinal strain of the right and left ventricle increased significantly during the first month of life. However, no significant changes in longitudinal strain rate were found. Finally, the electromechanical

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

interval, measured as the time between Q (on ECG) and the peak of the systolic wave, significantly shortened with advancing age of the patients: being measured at 12 points throughout the left ventricle and the lateral mitral and tricuspid annulus, time to peak systolic velocity decreased on average to 89% in the 2nd and to 80% in the 4th week of life (22.3 ± 0.2 ms vs. 19.8 ± 0.3 ms vs. 17.8 ± 0.5 ms, $P < 0.001$). When comparing opposing walls of the left ventricle, no dyssynchrony in time to peak systolic contraction was found.
Conclusions: During the neonatal period, increasing tissue velocities and shortening of the time to peak systolic contraction reflect the increasing efficiency of the excitation-contraction coupling in the maturing myocardium. Throughout this time, there appears to be no dyssynchrony in ventricular contraction. Longitudinal strain increases during this period; however, this may not be explained by changes in the loading condition alone. These novel findings could have significant implications in understanding the normal cardiac function during the neonatal period and in certain disease states.

P-90

Myocardial involvement and cardiac troponin T level in infants with Respiratory Syncytial Virus bronchiolitis

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Background: Respiratory Syncytial Virus (RSV) lung disease in infants is associated with myocardial involvement. In this study we aim to investigate the prevalence and the nature of associated myocardial involvement in children with RSV bronchiolitis.
Methods: The study was a prospective observational study. We assessed the myocardial involvement in infants with RSV bronchiolitis admitted to pediatric department and intensive care unit at the Western Galilee hospital during winter season 2011-2012. Cardiac Troponin T (cTnT), C Reactive Protein (CRP), Electrocardiogram, Chest X-ray, and Echocardiography were done for infants with RSV bronchiolitis.
Results: In our study 38 children were included. All infants were without congenital heart disease. Cardiac troponin T level was elevated in 8 infants out of 34 (23.5%). Level range was 0.056–0.209 Pg/ml. Seven infants out of eight (87.5%) with elevated cTnT level were younger than age of two months. Range of age was 0.75–2 months. Average 1.15 months. Echocardiography was possible for 24 infants. Cardiac function and rhythm were normal in all infants. Pericardial effusion was found in 11 out of 24 infants (45%). CRP was elevated in 21 infants (55%). Chest X-ray revealed pulmonary infiltrate in 13 infants. There was no significant difference between the patients with or without elevated cTnT with regard to the presence of pericardial effusion or pulmonary infiltrate, intensive care unit history or myocardial shortening fraction.
Conclusion: Myocardial involvement is common in infants with RSV bronchiolitis. Neither elevated cTnT level nor the presence of pericardial effusion were associated with myocardial dysfunction. Elevated cTnT was significantly correlated to younger than 2 months age.

P-91

Prevalence of subclinical rheumatic heart disease in urban and rural areas of north India: The e-RHEUMATIC (Extended-Rheumatic Heart Echo Utilization and Monitoring Actuarial Trends in Indian Children) Study

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Background: Studies have reported a higher prevalence of subclinical rheumatic heart disease (RHD) using echocardiographic screening in many countries including India. The prevalence may be significantly different in rural and urban areas of India, but is not studied. The objective is to compare the prevalence of subclinical RHD among rural and urban school children in India.
Methods: We carried out a cross sectional echocardiographic screening study among 8,454 randomly selected school children aged 5–15 years (10.6 ± 2.8 years; 57.9% male). A total of 6345 students were from rural areas and 2109 were from urban areas of New Delhi. We used the World Heart Federation criteria for diagnosing RHD by echocardiography.
Results: Clinical examination detected mitral regurgitation (MR) in 6 patients (clinical prevalence of RHD 0.7/1000 school children). Echocardiography-Doppler diagnosed definite or borderline RHD in an additional 75 cases. Hence a total of 81 children had RHD, giving a prevalence of 9.6/1000 school children (95% CI - 7.7–11.9/1000 children). The prevalence of definite or borderline RHD was 6.6/1000 school children in urban areas as compared to 10.6/1000 in rural areas. Even among the rural children there was a difference among children studying in government schools (12.2/1000) as compared to those in private schools (9.5/1000). Studying in a government school may be taken as a surrogate marker of lower socio-economic status.
Conclusions: The prevalence of RHD is higher among rural children as compared to urban children. The programs to control RHD should perhaps focus on such high prevalence zones.

P-92

Brain natriuretic peptide rises in response to different cardiac performance of every first strategy before Glenn procedure

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Background: The levels of plasma brain natriuretic peptide (BNP) are elevated highly in patients before Glenn procedure (pre-Glenn hBNP). Significant cardiac stress exists in patients with pre-Glenn hBNP. We predicted myocardial impairments were different in each first strategy. The purpose of this study was to investigate the cardiac performances which influenced pre-Glenn hBNP in every first strategy.
Method: The medical records of 126 patients before Glenn were reviewed. Cardiac catheterization was performed with Glenn in view between 2003 and 2011. Venous blood samples were obtained at the same point in time. We defined BNP levels 100pg/ml over as pre-Glenn hBNP. First strategies were divided into five categories. We examined what cardiac performances of each strategy affect pre-Glenn hBNP.
Results: After multiple logistic regression analysis pre-Glenn hBNP in all 126 patients were significantly associated with moderate atrioventricular valve regurgitation; increased ventricular volume of end-systole (ESV); increased ventricular pressure of end-systole; increased ventricular pressure of end-diastole (EDP); decreased ejection fraction of ventricle (R-square = 0.46). In monovariate analysis pre-Glenn hBNP were additionally related to increased ventricular volume of end-diastole (EDV). In separate analysis with first strategy, EDV, ESV and EDP were significantly related to pre-Glenn hBNP in group of shunting from systemic artery to pulmonary artery; EDV in native pulmonary stenosis; EDP in pulmonary artery banding. No cardiac performances were related to pre-Glenn hBNP

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

in groups of pulmonary flow relaying on ductus arteriosus; in groups of systemic flow relaying on ductus arteriosus which underwent bilateral pulmonary artery banding. *Conclusion:* Our study showed that variant cardiac strains were associated with pre-Glenn hBNP. And cardiac strains were different in every strategy. In ductus-depending groups cardiac performances were not clear which related to pre-Glenn hBNP. There may be latent damages of myocardium which were not represented by cardiac performances. If high levels of BNP were sustained, we should provide relief from different strains in every strategy based on this investigation. This may lead to stable condition before Glenn procedure.

P-93
High-levels of uric acid occurs in asplenia patients regardless of the degree of cardiac dysfunction

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Backgound: High-levels of uric acid (high UA) are associated with heart failure in adult patients. In children only one syndrome is known as gene disorder which takes on high UA without heart disease. In asplenia syndrome children have high UA often but they have cardiac dysfunction in varyng degrees. We investigated whether high UA exists without the levels of cardiac dysfunctions in asplenia.
Methods: The medical records of 124 patients after Glenn procedure were reviewed. Twenty-three patients were asplenia syndrome. We used other 101 non-asplenia as control. Cardiac catheterization was performed in stabilized period after Glenn between 2003 and 2011. Blood tests were performed within a few days before catheterization. We defined high UA as uric acid levels in the top quartile of 124 patients (UA $\geq$ 5.9 mg/dl). First, clinical data which influenced high UA were determined in all 124 by multivariate analysis. Second, the relationship between high UA and clinical data were investigated for asplenia, and for non-asplenia separately.
Results: Study age was not different between in asplenia and in non-asplenia (3.8 years vs. 3.7 years). Uric acid levels were higher in asplenia (5.9 mg/dl vs. 4.8 mg/dl, $p < 0.001$). In 124 patients after Glenn high UA was independently associated with odds ratio of 11.4 for levels of brain natriuretic peptide ($\geq$68.9pg/ml), 5.1 for asplenia syndrome and 3.9 for creatinine level ($\geq$0.29 mg/dl). In mono-variate analysis aging was additionally related to high UA. Polycycemia was not associated with high UA. Only for non-asplenia group study age, creatinine level and value of brain natriuretic peptide in patients with high UA were significantly higher than those in patients with non-high UA. For asplenia group these indexes were not different between in high UA and in non-high UA.
Conclusions: Asplenia syndrome was independently associated with high UA in young patients after Glenn procedure. Additionally high UA was not related to the degrees of heart dysfunction and kidney dysfunction in asplenia. Congenital factor may be responsible in part for high UA in asplenia. We should interpret the cause of high UA and consider the adoption of therapy in asplenia syndrome.

P-94
Clinical pictures of newborn infants who have been prenatally diagnosed with congenital heart disease but resulted in hospital mortality

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Introduction: It is expected that the prospective clinical care based on prenatal diagnosis using ultrasonography enables us to set up well-planned management and contributes to improvement of prognosis. While the prevalence of prenatally diagnosed congenital heart disease (CHD) has risen over the past decade, we experienced many severe cases die in early postnatal period.
Purpose: We reviewed clinical pictures of newborn infants who have been prenatally diagnosed with CHD (includes fetal arrhythmia) but resulted in hospital mortality.
Methods: Study population consisted of 89 consecutive newborn infants who were diagnosed with CHD (includes fetal arrhythmia) prenatally in our hospital between January 2011 and November 2012. We retrospectively analyzed the anatomical diagnoses, clinical course and cause of death in the patients who resulted in hospital mortality.
Results: Most common fetal diagnoses were fetal arrhythmias (19%), variant hypoplastic left heart syndrome (HLHS) (11%), tetralogy of Falllot (9%), coarctation of aorta or interruption of the aortic arch (9%), single-ventricular lesions (8%) and heterotaxy syndrome (4.5%). Thirteen infants (14.6%) resulted in hospital mortality. Of those, the cause of death was perioperative mortality in 9 (69.2%), chromosomal abnormality (18 trisomy or 21 trisomy) in 3, and the other one died of metabolic disorder. Of the 9 cases who resulted in hospital mortality, the underlying anatomical diagnosis was HLHS in 4, Ebstein's anomaly with circular shunt in 2, critical aortic stenosis (cAS) with endocardial fibroelastosis (EFE) in 1, pulmonary atresia with intact ventricular septum and cAS in 1 and remain was left atrial isomerism with congenital complete atrio-ventricular block (cAVB). Two cases with HLHS died of fatal capillary leakage syndrome.
Conclusions: All the 13 (14.6%) infants resulted in hospital mortality had severe heart defect or systemic disorder. Especially in Ebstein's anomaly with circular shunt, cAS with EFE and left atrial isomerism with congenital cAVB, the further development of new therapeutic methods or strategies are strongly desired in future. In severe heart defects, it is still difficult to save them even if they have been diagnosed prenatally.

P-95
Natural history of Barth syndrome: A national cohort study of 22 patients

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This study describes the natural history of Barth syndrome (BTHS).
Methods: The medical records of all patients with BTHS living in France were identified in multiple sources and reviewed.
Results: We identified 16 BTHS pedigrees that included 22 patients. TAZ mutations were observed in 15 pedigrees. The estimated incidence for BTHS was 1.5 cases per million births (95%CI: 0.2–2.3). The median age at diagnosis was 3.1 weeks, and the median age at last follow-up was 4.75 years (range, 3–15 years). Eleven patients died at a median age of 5.1 months; 9 deaths were related to cardiomyopathy and 2 to sepsis. The 5-year

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

survival rate was 51%, and no deaths were observed in patients ≥3 years. Fourteen patients presented with cardiomyopathy (dilated DCM and/or hypertrophic HCM), and cardiomyopathy was documented in 20 during follow-up. At diagnosis, the median LVEDD z-score was 4.5 (p 25: 1.9–7.5), the median LV-mass z-score was 3.5 (p 25: 1.7–p 75: 5.5), the median SF was 16% (p 25: 12.8–p 75: 25) and the median EF was 32.5% (p 25: 25.2–p 75: 43). At diagnosis, 6/11 patients had associated DCM and HCM (54.5%), 2 had DCM and 1 had HCM. In addition, 7 patients (32%) had prominent trabeculations of the LV, either on echocardiogram or on MRI, and were considered to have left ventricular noncompaction (LVNC). Left ventricular systolic function was very poor during the first year of life and tended to normalize over time. Nineteen patients had neutropenia. Metabolic investigations revealed inconstant moderate 3-methylglutaconic aciduria and plasma arginine levels that were reduced or in the low-normal range. Survival correlated with two prognostic factors: severe neutropenia at diagnosis (<0.5 G/L) and birth year. Specifically, the survival rate was 70% for patients born after 2000 and 20% for those born before 2000.
Conclusions: This survey found that BTHS outcome was affected by cardiac events and by a risk of infection that was related to neutropenia. Modern management of heart failure and prevention of infection in infancy may improve the survival of patients with BTHS.

P-96
Anatomy of the ventricular septal defect in congenital heart defects: a random association?
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Introduction: A ventricular septal defect (VSD) is part of most congenital heart defects (CHD).
Aim of the study: To determine the distribution of the anatomic types of VSD in CHD.
Material and methods: We analyzed morphologically 1178 heart specimens with CHD from the anatomic collection of the French Reference Center for Complex CHD. Special attention was paid to the localization of the VSD: muscular, membranous, outlet located between the two limbs of the septal band, inlet. The specimens were classified according to the anatomic and clinical classification of CHD (ACC-CHD).
Results: A VSD was present in 67% of all hearts and was:

- Constant, of a single type, in tetralogy of Fallot and variants and common arterial trunk: outlet, in complete atrioventricular canal (CAVC): inlet, and in double-inlet left ventricle (DILV): muscular.
- Not constant with a predominant type, in 96% of double discordance (DD,inlet 82%), 62% of heterotaxy syndromes (Hetx, inlet 93%), 93% of interrupted aortic arch (outlet 80%), 87% of double outlet right ventricle (outlet 77%).

Not constant, of variable type, in 68% of aortic coarctation (CoA: outlet 44%, membranous 35%, muscular 21%), 54% of transposition of the great arteries (TGA: outlet 40%, membranous 25%, muscular 25%, inlet 10%).

- Rare, in anomalies of pulmonary veins (5%), Ebstein anomaly (14%), double-inlet right ventricle (10%), coronary anomalies (25%).
- Isolated in 10% of all VSD: outlet 44%, membranous 36%, muscular 18%, inlet 2%. Associations according to VSD type:
- outlet : 60% "conotruncal" defects (CTD), 10% TGA
- inlet : 57% CAVC, 13% DD, 10% Hetx
- muscular : 33% DILV, 26% TGA, 13% isolated
- membranous : 30% TGA, 28% isolated, 16% CoA.

Conclusion: The VSD is an integral part of the phenotype in some CHD. In CoA and TGA the VSD is not constant and its anatomic distribution is similar to that in isolated VSD, indicating a likely random association. This reinforces the hypothesis of different genetic mechanisms in TGA and CTD. This original approach, using the anatomic characteristics of one part of the phenotype, could provide new insights in the grouping and aetiology of CHD.

P-97
Congenital Heart Disease in a Population of Twins: the role of Assisted Reproduction Technology
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Introduction: There is increased incidence of congenital anomalies in neonates resulting from twin gestations. Assisted reproduction technology which is related to increased rates of twinning has also been associated with increased risk of birth defects. We studied the incidence of congenital heart disease in a population of twins and its relationship with assisted reproduction.
Methods: Data concerning 849 live-born twin neonates (of which at least one of the co-twins) was admitted in the Neonatal Intensive Care Unit during 1995–2011 were analysed. Forty-four % (189/429) of the gestations were the result of assisted reproduction (which was defined as in vitro fertilisation or intracytoplasmic sperm insertion).
Results: In the assisted reproduction group 29/373 (7.8%) had congenital heart disease compared to 20/476 (4.2%) neonates conceived naturally ($p = 0.037$). The spontaneous conception gestations had higher incidence of monochorionic placentation (47/240 versus 4/189, $p < 0.0001$) and included mothers who were younger (29.1 ± 4.7 years versus 33.9 ± 5.2 years, $p < 0.0001$) and had a higher parity (median 2, range 1–7 versus 1, range 1–4; $p < 0.0001$) compared to the assisted reproduction gestations. Multinomial logistic regression analyses using fetal chorionicity and gender, maternal age and parity as covariates, showed that assisted reproduction and monochorionic placentation were both significant determinants of congenital heart disease ($Exp(B) = 2.534$, $p = 0.016$ and $Exp(B) = 3.058$, $p = 0.007$, respectively) and major birth defect ($Exp(B) = 2.287$, $p = 0.004$ and $Exp(B) = 2.195$, $p = 0.022$, respectively].
Conclusions: There is increased incidence of congenital heart disease and birth defects as a whole in twins resulting from assisted reproduction. Although in vitro fertilisation and intracytoplasmic sperm insertion are considered safe procedures both for the mother and the offspring, detailed antenatal screening including fetal echocardiography should probably be offered to pregnant women carrying multiple fetuses, especially post artificial reproduction.

P-98
Three siblings with the extremely rare geleophysics dysplasia and different severity of cardiac involvement
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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Background: Geleophysic dysplasia (GD) is an extremely rare autosomal recessive skeletal dysplasia resembling lysosomal storage disease, characterized by short stature and short limbs, joint contracture and cardiac involvement. The most frequent cardiac lesions described being the mitral valve, the aortic valve followed by pulmonary valve and the least is tricuspid valve. It has been described in few cases worldwide.
Aim: To study the progression of cardiac lesions in patients with Geleophysic dysplasia.
Methods and Results: We reviewed the cardiac lesions in three female siblings of a Saudi family who showed different severity of the valvular involvement. The three sisters are seven, six and two years old. They were born to first cousins parents who also have 3 years healthy son. They had history of respiratory problems requiring frequent hospital admissions. For last 3 years weight and height grew parallel but considerably below 3rd centile. Furthermore they shared dysmorphic features of small and broad hands and feet, hypertelorism, depressed nasal bridge and anteverted narse. They have limitation of flexion of their hands with bilateral contractures of both elbow and knee joints. They have happy and friendly personality with normal intelligence. Genetic studies confirmed the heterogenicity of the ADAMTSL2 gene.
All sisters have associated cardiac lesions with different severity. The eldest has mild pulmonary and aortic valve stenosis. The middle sister has thickening of mitral valve leaflets without stenosis. The youngest sibling has severe aortic valve stenosis and mild pulmonary valve stenosis.
We observed that the valvular involvement remained static in first two patients over period of 3 years. This observation helped to manage the third case in a more conservative way despite severe aortic stenosis and after one year follow up the aortic valve gradient remained the same.
Conclusions: In geleophysics dysplasia cardiac valves are usually thickened leading to stenosis. Most of the reports suggested progression of the disease. In our three cases the youngest has the most severe form of valve involvement. This suggests that the expression of the gene could determine the severity of the disease, rather than the belief of progression with time.

P-99

Exercise training improves fitness without adverse cardiac remodelling in patients after repair of tetralogy of Fallot: Preliminary results of the TOFFIT study

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Objective: To assess a) whether aerobic exercise training (AET) improves exercise capacity and b) if AET results in adverse cardiac remodelling, in children and young adults after repair of tetralogy of Fallot (TOF).
Methods: Design: multicenter randomized controlled trial. Forty-seven participants after repair of TOF were recruited from 5 tertiary referral centres. Participants were assigned to an interventional or control group. All underwent cardiopulmonary exercise testing and cardiac MRI before and after a 12 week period. Patients in the intervention group performed supervised aerobic exercise training (AET) for 12 weeks. AET consisted of 2–3 sessions/week, for 1 hour/session at an exercise level predicted by 60–70% of heart rate reserve. The control group did not change their life style.
Results: Preliminary results of the 47 participants after TOF repair: baseline characteristics of the intervention (AET) group ($n = 27$): age 15.5 ± 2.8 years; 21 males; controls ($n = 20$): age 16.0 ± 2.6; 12 males. After the 12 week AET program peak VO_2 of the intervention group improved slightly and significantly (35.6 ± 7.1 vs 37.8 ± 8.1 ml/kg/min, $p = 0.02$). In the control group no changes were noted (33.8 ± 8.0 vs 34.8 ± 8.0 ml/kg/min). Maximal work load of the intervention group improved significantly (170 ± 56 vs 179 ± 53 Watt, $p = 0.001$). The control group did not show any change (167 ± 36 vs 168 ± 39 Watt). The oxygen uptake efficiency slope (OUES) did not change in either group.
Cardiac MRI did not reveal significant changes for right and left ventricular size and function in either group. For the AET TOF group vs TOF controls: RVEDV AET group: pre-intervention 127 ± 37 vs 122 ± 30 ml/m^2 post-intervention; controls 132 ± 39 vs 130 ± 42 ml/m^2. RVSV AET: pre-intervention 61 ± 14 vs 61 ± 14 ml/m^2 post-intervention; controls 66 ± 17 vs 65 ± 19 ml/m^2. RVEF AET: pre-intervention 50 ± 9 vs 51 ± 7% post-intervention; controls 51 ± 7 vs 51 ± 6%. LVEDV AET: 82 ± 13 vs 84 ± 21 ml/m^2; controls 83 ± 7 vs 81 ± 12 ml/m^2. LVSV AET: 48 ± 6 vs 47 ± 6 ml/m^2; controls 51 ± 7 vs 50 ± 9 ml/m^2. LVEF AET: 60 ± 10 vs 60 ± 9%; controls 51 ± 7% vs 51 ± 6%.
Conclusion: Aerobic exercise training improves exercise capacity of children and young adults after repair of tetralogy of Fallot. Interim analysis did not show adverse cardiac remodelling.
Supported by NHF grant 2008B026.

P-100

Congenital heart disease in Tunisia: what can we learn from an echocardiographic study in a developing country?

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All patients with congenital heart disease (CHD) diagnosed in the cardiology department at hedi chaker hospital (Sfax) from july 2009 to june 2012 were included in the study based on echocardiography findings. Patients were followed up to September 2012.
Results: 674 patients, 330 (49%) males with congenital heart disease were studied. All patients were evaluated with echocardiography. Left-to-right shunts were the commonest defect in 342 patients (50,7%), caused by ventricular septal defect in 154 patients (22,8%), atrio-ventricular septal defect in 45 patients (6,6%), atrial septal defect in 75 patients (11,1%) and persistence of ductus arteriosus in 67 patients (10%). Tetralogy of Fallot was the most common cyanotic defect, present in 29 patients (4,3%). Transposition of great arteries was the second cyanotic defect with 19 cases.
Coarctation of the aorta were diagnosed in 27 cases, fourteen of which died.
428 (63%) children presented for echocardiography before the age of one year, and 532 (79%) presented before the age of five years.
121 children died during the study, a case fatality of 17,9% if we consider all cases of CHD but it reaches 37% if we consider only severe CHD.
Conclusions: The situation of pediatric cardiology is still worrying in Tunisia. This study has revealed a high case fatality rate among

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

children suffering from CHD. Much remains to be done to guarantee that every child born in Tunisia with cardiac anomalies can have access to appropriate medical and surgical care.

P-101

Long term follow up of patients with Ebstein's anomaly

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Introduction: Ebstein's anomaly (EA) is a rare congenital cardiac anomaly characterised by downward displacement of the septal and posterior leaflet of tricuspid valve (TV), which may result in cyanosis, right heart failure and tachyarrhythmia during the newborn period or after adolenscence. Due to the rarity of the diseases, the data regarding life-expectancy are still limited, escpecially in a small country.
Aim: To define the demographic parameters, clinical manifestations and long-term outcomes in Hungarian cohort of EA.
Patients and Methods: The downward displacement of the TV from the atrioventricular ring? 0.8 cm/m body surface area was set as the criterion for EA. Patients with EA+congenitally corrected transposition of the great arteries were excluded. Data of 32 (20 male) patients with EA born between January 1994 and December 2012 in Hungary were studied.
Results: The frequency of EA during this period was 1.67/100.000 life birth. The clinical signs leading to the diagnosis were: abnormal echo scan in foetus (5), cardiac murmur (13), cyanosis (7), dyspnoe (7), heart failure (6), extracardiac malformation (4) in the perinatal period (all of 26 cases), and cardiac murmur (3), cyanosis (1), dyspnoe (1), cardiac arrythmia (1) in childhood (all of 6 cases). Average gestational age was 38.5 ± 2.2 weeks, birth weight was 3280 ± 667 g. Extracardiac malformations occur in 10 cases. The follow-up time was 87,0 months (1–218). Majority of the associated cardiac anomalies disappeared spontaneously during the follow-up period: ASD-II (28–11), PDA (9–0), VSD (2–0), PS (6–6), PA (1–1). The QRS duration increased from 76.9 ± 17.4 (58–120) ms to 98.4 ± 24.1 (60–161) ms, occurance of incomplete RBB increased from 10 to 13, complete RBB from 2 to 10, preexcitation from 1 to 3, respectively, during the follow-up period. It happen 2 cardiac intervention, 1 RF ablation, in 8 cases one cardiac operation, in 4 cases two, and in 3 cases three. There were 3 death (49,6, 6–108 months) related to the EA.
Conclusion: The occurance of EA in Hungary in this period is higher, the ratio of extracardial anomalies is higher, the complexity and the mortality is much more lower, than is in the literature.

P-102

Role of intravenous immunoglobulin in the treatment of acute myocarditis

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Background: Acute myocarditis is rare disease in infants and childhood, the role of Immunosuppressive agents and Intravenous Immunoglobulin in the treatment of this disease was investigated by many researchers. The immunosuppressive agents found to be not effective. The potential benefits of Intravenous Immunoglobulin (IVIG) are promising in the treatment of patients with acute myocarditis and recent onset Dilated Cardiomyopathy (DCM).
The aim of this study: Is to investigate the benefit of IVIG in the patient with acute myocarditis and dilated cardiomyopathy as indicated by the analysis of the improvement in the ventricular contractility.
Patient and method: A nonrandomized controlled clinical trial conducted in the cardiology department at Tripoli Children's Hospital from 2002–2005. Forty patients (40) were diagnosed with acute myocarditis and Dilated Cardiomyopathy based on clinical features, laboratory data and echocardiograph findings. Left Ventricular Ejection Fraction (LVEF) <55% were the main inclusion criteria. The Age at presentation, sex and the presenting symptoms, of the patient were recorded they were followed up at 3, 6, and 12 months after the treatment.
Results: The median for the age at presentation was 9 months, Male to female ratio is 1:1.5, and congestive heart failure was the common presenting symptom.
Thirteen patients (13) were treated with Intravenous Immunoglobulin (IVIG) (0.4 g/kg/day) for 5 days; all patients were followed to see the changes of left Ventricular Ejection Fraction (LVEF). In this group the LVEF determined by Echocardiogram improved from 0.44 ± 0.025 (mean± S.D) at base line to 0.59 ± 0.36 at follow up over the course of one year ($P < 0.01$). LVEF improved 15 EF units in those patients received IVIG compared with the other group who did not receive IVIG where the LVEF changed from 0.36 ± 0.029 at base line to 0.40 ± 0.06 at the end of one year follow up ($P = 0.08$) and the LVEF improved just 9 EF units.
Conclusion and Recommendation: IVIG seems to be a promising agent in the treatment of acute myocarditis. The effectiveness of intravenous immunoglobulin therapy in acute myocarditis should be evaluated further in randomised multicenter trials.
Key words: Acute myocarditis, dilated cardiomyopathy, IVIG.
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P-103

RSV Immunoprophylaxis treatment for RSV in Children with Congenital Heart Disease in Sweden.

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Introductions: Respiratory Syncytial Virus (RSV) is the most common cause of lower respiratory tract infection in infants and young children. Children with congenital heart disease (CHD) have an increased risk of severe RSV disease. Palivizumab, a monoclonal antibody, is an effective, safe and well-tolerated prophylaxis for serious RSV disease. Swedish National Guidelines for prophylactic treatment for CHD children were published 2003. Aim of this study was to study, during the seasons of 2010–11 and 2011–12, if the prophylactic treatment corresponded to the guidelines and to study the morbidity of children that received prophylaxis and yet got a RSV infection.
Methods: All Swedish paediatric cardiology centres were invited to the study. A questionnaire for each treated child with information on CHD-diagnosis, age at start, number of injections and RSV-infection was retrieved. Medical journals of all RSV cases treated were obtained from each hospital.
Results: A total of 219 children were included in the study. Overall there were 869 doses of Palivizumab given during the study-period. The majority of children started treatment during their first six months of life. Each child received in average 3.97 (1–7) injections. About 51% of the children started treatment

according to recommended time of the year. Single ventricle was the most common type of CHD. A total of ten children (4,5%) were tested positive for RSV infection. Five of them had received non or one injection prior the infection and in three cases the infection prolonged time to operation.
Conclusions: The majority of children were treated according to the Swedish National Guidelines. However, adverse time and age at start of treatment was seen. The epidemiological status of RSV in society and guidelines for RSV prophylaxis in other countries may have affected the clinicians' decision of starting time. Ten of the children treated were affected by RSV, which is more than compared to resent studies. This study indicates that even if the infant is treated with Palivizumab immunoprophylaxis, the time of start of treatment and identification of CHD children is essential. The National Guidelines are now being reviewed.

P-104
The Value of Carina Angle Measurement for the Diagnosis of Patent Ductus Arteriosus
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Introduction: Bedside diagnosis of patent ductus arteriosus (PDA) continues to maintain its importance particularly when echocardiography is not available at the moment In our study we aimed to assess whether the widened carina angle (CA) displayed on X-ray supports the presumptive presence of PDA.
Methods: The study group was consisted by 60 infants under 37 weeks diagnosed with hemodynamically significant PDA, based on clinical and echocardiographic findings. The control group was consisted by 60 infants with no clinical or echocardiographic evidence of PDA. In both groups, the location of the left main bronchus was assessed by measuring the angle between two main bronchi at the level of carina.
Results: In the comparison between two groups, a significant widening of carina angle was found in PDA group and while the interquartile range (IQR) was found as 69– 108°, the median was found as 89° and the mean was 87.26° (±7.01 °) in PDA group, these values were found as 57–89°, 66.5° and 67.4° (±7.33°) respectively, in the control group ($p < 0.001$). A cut-off point of 73.5 ° signified the highest sensitivity (97%) and specificity (55%). We found a significant and positive correlation between the increased CA values and PDA occurrence ($p < 0.01$). When PDA was closed these values were respectively 63–88°, 74.5° and 74.7° (±6.4°) ($p < 0.001$).
Conclusions: We demonstrated that the probability of the appearance of a widened CA on X rays was increased by the presence of PDA. Similarly, a CA angle narrower than 73.5° and a negative predictive value of 93% eliminate the diagnosis of PDA. CA is a reliable and widely available tool in making the diagnosis of PDA.

P-105
Aggressive Respiratory Therapy for chronic pulmonary disease optimizes clinical outcomes following surgery for complex congenital heart defects
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Introduction: Pulmonary complications in children with complex congenital heart disease (CCHD) can have a profound adverse influence on the final clinical outcome following surgery for CCHD. Early recognition and aggressive respiratory management has a critical role in management of these patients.
Methods: A retrospective database search was performed to identify patients who attended both cardiology and respiratory outpatient clinics from January 2010 – December 2012.
Entry criteria: 1) CCHD following surgery on home ventilation (Oxygen/CPAP/BiPAP); 2) prolonged ventilation >14 days following initial surgery; 3) pleural effusion/chylothorax >14 days following initial surgery. Respiratory therapy included the use of inhalers (bronchodilators and steroid), antibiotics and oral steroids. Clinical data including demographics, cardiac diagnosis, co-morbidities, surgical procedures, respiratory complications, respiratory and cardiac management Ejection fraction (EF) and pulmonary artery pressure measurements (PAP), pulmonary function tests (PFTs) were recorded.
Results: 24 children in total were identified, 56% were male, median age 7.2 years (range 2–20 yrs). 54% presented with right side lesions,16/24 had multiple congenital anomalies. 40% met entry criteria of prolonged pleural/chylous effusion or ventilation >14 days post initial surgery. 18/24 had normal EF, mean 63% (range 48–74%). 21/24 had normal PAP (<25 mmHg) with no significant RVOT obstruction (<20 mmHg) indicating satisfactory surgical result. 6/24 required cardiac transplantation. Formal PFTs were obtained in 9/24 children (3 transplant). Pre-intensive respiratory therapy mean Forced Vital Capacity (FVC) was 47% (34–80%) predicted and Forced Vital Capacity (FVC) was 47% (FEV) was 46% (16–86%). Following respiratory therapy both increased significantly, FVC 59% (21–82% $p < 0.001$), FEV 54% (16–81% $p < 0.03$). At time of initial review 12/24 required some form of home ventilatory support despite normal EF and optimal cardiac repair; 8/12 had known upper airway lesions. At completion of the review period 4/8 were off ventilatory support.
Conclusion: Despite obtaining an adequate mechanical repair with satisfactory ventricular function, significant respiratory dysfunction as sequelae from a complicated cardiac repair has a profound adverse influence on final clinical outcome. Aggressive respiratory therapy can result in significant improvement in lung function (confirmed by PFTs) and allow completion of multistage surgical procedures. In this group of patients the lungs have a tremendous capacity to heal.

P-106
Comparison between tilt table and active orthostatic testing for syncope simulation and hemodynamic pattern investigation in children
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Objective: To evaluate effectiveness of different orthostatic tests (tilt table test and active lying-standing test for syncope simulation and hemodynamic pattern's investigation in children suffering from recurrent syncope.
Methods: 205 children in age from 4 to 17 years old (mean age 13, 08 ± 2,88) after at least the 3rd episode of syncope and having no contraindications revealed on physical examination, 12-lead electrocardiogram, underwent orthostatic testing: 72 – tilt table testing, 133 – active standing testing according to random selection. Blood pressure and electrocardiogram were recorded by monitors DASH 2000 and Datascope DuoTM. Results were compared with those in control group.
Control group: 92 healthy children (mean age 12,64 ± 3,04) never had syncope or presyncope episode, underwent physical

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

examination, 12-lead electrocardiogram and orthostatic testing. Tilt testing was performed in 40, active standing testing – in 52 control group's children. There was no statistically significant difference of groups in gender and age division. Data were processed with SPSS 17,0 statistical packet. Comparison between groups was made using Student's t-test, F (ANOVA), and $\chi 2$ test.
Results: Tilt table test was positive in 72,2%, and active test was positive in 71.4% of investigation group children. There was detected no statistical significant difference, $p = 0.904$, $\chi^2 = 0.015$. Tilt testing was positive in 27.5% children from control group, and active standing test – 23.1% accordingly. Statistical significance of control group was $p = 0.627$, $\chi^2 = 0.236$. Difference of positivity in orthostatic testing between children with recurrent syncope and children never had the episode was significant: $p = 0.000$, $\chi^2 = 56.6$. Patient's age had no significant influence for orthostatic test results ($p = 1.19$, $\chi^2 = 2.36$). No statistical significant difference for orthostatic test results was noticed in patient gender ($p = 0.34$, $\chi^2 = 0.89$) and reflex or atypical circumstances of syncope ($p = 0.31$, $\chi^2 = 1.03$).
Conclusion: Tilt table testing and active orthostatic testing showed no statistical significant difference for syncope simulation and it's hemodynamic pattern investigation in children suffering from recurrent syncope.

P-107
Genes' condition in infants with congenital heart diseases and ischemic stroke

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Introduction: Cerebral stroke in children is very rare and can be caused by huge number of diseases. Congenital heart malformations and thrombophilia are described to be the most frequent reasons for stroke's debut in infants. But interaction between them is not investigated thoroughly.
Methods: Type of study: case series. Inclusion criteria: boys or girls between the age of 0 and 12 months with congenital heart disease; acute ischemic stroke, which has developed immediately after cardiac surgery and confirmed by brain CT scan; slavic origin; informed consent form. We identified single nucleotide polymorphisms (SNPs) of thrombophilia (8 SNPs), folic acid cycle's enzymes (4 SNPs) and arterial hypertension (9 SNPs) in 10 children's (6 girls, 4 boys) blood samples by polymerase chain reaction.
Results: All children had more than three SNPs of thrombophilic genes in the homozygous or heterozygous state: F2: G20210A (n = 1), F5: G1691A (n = 0), F7: G 10976A (n = 7), F13: G103T (n = 3), PAI-1: -675 5G/4G (n = 10), FGB: G-455A (n = 8), ITGA2: C807T (n = 9) and ITGB3: T1565C (n = 4). All patients had combinations including SNPs platelets receptors (ITGA or ITGB) and fibrinolytic system (PAI); 8 children had combinations with coagulation factors, platelets receptors and fibrinolytic system. Each patient carried two and more mutations of folic acid cycle: MTHFR: C677T (n = 7), MTHFR: A1298C (n = 4), MTRR: A66G (n = 8), MTR: A2756G (n = 4). Homocysteine's level exceeded the norm in two-five times; average level was 13,3 ± 2,1 umol/l.
All children had more than 4 SNPs of arterial hypertension: ADD1: G1378T (n = 1), AGT: T704C (n = 7), AGT: C521T (n = 2), AGTR1: A1166C (n = 4), AGTR2: G1675A (n = 9), CYP11B2: C344T (n = 9), GNB3: C825T (n = 4), NOS3: T786C (n = 8), NOS3: G894T (n = 3). Combination AGTR2: G1675A and CYP11B2: C344T was identified in 9 children.
Conclusion: We assume prothrombotic and procoagulant genes' polymorphisms to be the main reason of early life stroke's debut: eight and more candidate genes were identified and have been realized as stroke and hyperhomocysteinemia. The operation technique can be considered to be an essential risk factor for stroke in children with congenital heart disease. Combination of "sticky platelets" syndrome with defective fibrinolytic system and vessels' tonus regulators, have the most diagnostic value in these patients.

P-108
Analysis of polymorphisms in genes (ADD1 1378 G>T, AGT 704 T>C (Met235Thr), AGT 521 C>T (Thr174Met), AGTR1 1166 A>C, AGTR2 1675 G>A, CYP11B2 344 C>T, GNB3 825 C>T, NOS3 786 T>C, NOS3 894 G>T (Glu298Asp)) associated with hypertension in children of Ural

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Introduction: During the last few years the studies on the genetic basis of essential hypertension in adults have been numerous but there is little data of association of gene polymorphisms with hypertension in children to date.
Objective: To investigate the role of gene polymorphisms in predicting of essential hypertension in male and female children.
Methods: The study subjects consisted of 31 essential hypertensive children (21 boys, 10 girls), aged from 6–18 years old. We identified the following genetic variants of ADD1 1378 G>T, AGT 704 T>C (Met235Thr), AGT 521 C>T (Thr174Met), AGTR1 1166 A>C, AGTR2 1675 G>A, CYP11B2 344 C>T, GNB3 825 C>T, NOS3 786 T>C, NOS3 894 G>T (Glu298Asp) in children. Gene DNA was extracted from blood samples and amplified by polymerase chain reaction (PCR).
Results: The study showed association of ADD1 1378 G>T, AGT 704 T>C (Met235Thr), AGT 521 C>T (Thr174Met), AGTR1 1166 A>C, AGTR2 1675 G>A, CYP11B2 344 C>T, GNB3 825 C>T, NOS3 786 T>C, NOS3 894 G>T (Glu298Asp) genes with hypertension in all children. The combinations of three to seven gene polymorphisms were found more frequently. Our study has revealed that female children have the following gene polymorphisms: AGT 704 T>C and AGTR2 1675 G>A more frequently than male children ($p<0.05$). From the other side, the polymorphism CYP11B2 344 C>T was found in 76% of boys. The identification of nitric oxide gene polymorphisms (NOS3 786 T>C, NOS3 894 G>T) hasn't depended on the sex of children.
Conclusion: The gene polymorphisms AGT 704 T > C and AGTR2 1675 G > A are associated with hypertension and may be a genetic markers of early onset of disease in female children, the gene polymorphism CYP11B2 344 C > T may be a predisposing factor of essential hypertension in boys. Further investigations are required.

P-109
Croatian clinical epidemiological study (2008–2011): the use of standardised risk scores in pediatric congenital cardiac surgery for a case complexity selection and gradual progress of cardiosurgical model in developing countries

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Objective: The evaluation of the quality of care delivered to patients with the congenital heart diseases relies heavily on the analysis of outcomes. The EACTS database was created to assess the outcomes of congenital cardiac surgery, enable comparison of results, definition of risk factors and targeting research activities. By employing the widely used and accepted methodologies of case mix complexity adjustment in congenital cardiac surgery, we tried to evaluate our performance, and to use the ABC scores for a case complexity selection that may have different outcomes in various centres.
Methods: In this report, we analysed outcomes of cardiac surgical procedures performed in our institution between January 2008 and December 2011. Data were collected in the EACTS database and represent surgical procedures performed with or without cardiopulmonary bypass. This section contains information on patient, early mortality, occurrence of postoperative complications, and procedural complexity presented by ABC scores. Together with prospective collection of these data, all patients sent for operation in foreing cardiosurgical centers were recorded.
Results: During the period of study, 634 operations were done, among them 60% were performed in Croatia, 40% in foreign cardiosurgical centers. The number of operations performed in Croatia showed linear increase: 55 operation, 78, 121 and 126 operations performed in a year 2008, 2009, 2010, 2011 respectively. Early mortality was 1.82%, 5.41%, 3.64%, 3.48% in 2008, 2009, 2010, 2011 respectively. Increase in number of operation was followed with satisfactory low average mortality of 3.85%. The mean complexity, according to the risk adjustment methodology for cardiac procedures performed in Croatia is 5.77, with no statisticaly important changes during years of study. When analysing the procedures performed in our country, assigned by higher ABC score, we determined statistically significant connection between ABC score and mortality rate.
Conclusion: The use of standardised risk scores allows selection of complex cardiac diseases which may have very different outcomes in various centres. In our case, those with higher ABC scores were correctly identified and referred for treatment abroad. In this way, we allowed gradual progress of cardiosurgical model in our country and maintain an enviable low mortality.

P-110
Activated clotting time (ACT) for monitoring anticoagulant effect of heparin in children undergoing cardiac catheterization – Are there age-dependent differences?

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Objectives: Activated clotting time (ACT) is a useful parameter to monitor anticoagulant effect of unfractionated heparin (UFH) in children undergoing cardiopulmonary bypass surgery or cardiac catheterization (CC). The aim of the study was to evaluate UFH effect on ACT in different age groups during CC.
Methods: We analyzed the effect of initial bolus administration of UFH (100 IU/kg) in children undergoing CC on ACT comparing three different age groups (0–1 year, 1–5 years, and 6–18 years).
Results: Between April and December 2012 in 104 patients (58 male) we performed CC at median age of 3.1 years (range 0.1–18.1) and a body weight 13.4 kg (2.5–90.0). The age groups included 33 patients between 0–1 years (I), 38 between 1–5 years (II), and 33 between 6–18 years (III). The ACT was 196 sec (160–395) in group I, 211 sec (129–344) in group II, 212 sec (118–344) in group III, respectively, at the end of the procedure 60 min (17–200) after bolus injection of UFH (spearman rho $= -0.33$, $p < 0.05$). There were no significant differences of ACT value comparing the three age groups, despite a trend towards more frequent lower outliers in the subtherapeutic range for older children at the end of the procedure.
Conclusions: The effect of bolus administration of UFH is comparable in all age groups from the neonate to the adolescent resulting in desired ACT values of ≥ 200 sec during CC procedures for at least one hour. Second repetitive UFH injection during longer-lasting procedures should be controlled with preceding ACT measurement. Further prospective trials are needed to determine the impact of variables affecting ACT such as platelet count (thrombocytopenia) and platelet function (use of Aspirin) as well as factor deficiencies (liver disease, critical ill infants during severe periods of postoperative ICU), anticoagulants (low vitamin K dependent clotting factors), and hemodilution (volume overload during CC).

P-111
Risk of pneumonia in adult patients with atrial septal defect

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Objectives: According to medical textbooks pneumonia is common for patients with atrial septal defect (ASD). The pathogenesis and incidence are, however not well characterized and it is not known whether ASD closure protects against pneumonias. In this nationwide cohort study we describe the incidence of pneumonia in patients and compare it with the background population. The patients investigated are those with an open ASD, those having the ASD closed interventionally or surgically, and those where the ASD has been judged insignificant and has been left untreated.
Methods: All patients registered in Denmark between 1977 and 2009 with an ASD were included. Follow up ended December 2011. Patients aged below 18 years at present, patients with other congenital heart disease except PDA and those only diagnosed once before the age of one year were excluded. For every patient in the cohort 10 persons from the Danish Civil Registration were matched on gender and year of birth. Data from the Danish National Patient Registry and the Danish Registry for Congenital Heart Disease were used. All hospital contacts with pneumonia (bacterial, viral, abscesses or empyema) were identified.
Results: 2391 patients of whom 789 had their ASD closed (421 by surgery and 368 by catheter) were included. Pneumonia was seen at least once in 463 (19%) of the patients with ASD (Closed ASD N = 135 (17%), not closed ASD N = 328 (20%)). Nine percent of the patients with ASD had more than one pneumonia with a total of 1008 pneumonias (Closed ASD N = 189 before closure, N = 96 after closure). In the general population comparison cohort 2176 persons (9%) had pneumonia at least once with a total of 4063 pneumonias (3,5% had more than one pneumonia). Comparison of prevalence per year before and after ASD closure and comparison the general population cohort is still pending as well as time-to-event analysis.
Conclusion: We have found that the proportion of ASD patients with pneumonia is increased compared with the general population. We will at the time of presentation have further data on the impact of ASD closure on incidence of pneumonia.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

P-112

Early introduction of self-educational program for the early adolescents with congenital heart disease in outpatient clinic of the local independent children's hospital-nursing perspectives

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Background: Although the patient care for the adults with congenital heart disease (ACHD) has been emphasized in centralization of ACHD medical center, the self-educational program of the patients preparing their independence is also mandatory but often passed away.
Aim: To investigate the self-assessment status of the ACHD patients by questionnaires and to assess the nursing patient-aid program to help postsurgical children learn their disease to be an independent adult.
Methods: The subjects were 415 young children and early adolescents with CHD in outpatient clinic, age ranged 10–34 years; (194 < 15y, 114: 15–18y, 104: 18y<). After they learned their disease by drawing figures and explanations of charged doctors, they filled the questionnaire regarding to their diagnosis, surgical procedure, hemodynamics, medications, social matters and the issues of pregnancy in our hospital which was established 20 year ago. Then their achievement degree was reassessed by the nurses and encourage them to make a plan to learn by themselves.
Results: All patients were once informed from the age of 6 years old about their diagnosis and the content of treatment including the surgery and medications with figures in outpatient clinic. At the second visit after having information, more than 70% of the patients could answer their timing of the surgery. Of 415, the correct answer of their diagnosis and treatment of the patients was 62% (18y<): 23% (<15y), 42% (18y): 19%(<15y), respectively. The prevention of infective endocarditis and pregnancy were poorly recognized as 34%, 19% even in 18y<. After the aid of nursing support program, 67% of patients could improve their achievement scores.
In conclusion, self-educational program for CHD children/adolescents is indispensable to train them to be independent in adult, which could be introduced even in the local children's hospital.

P-113

Congenital left ventricular aneurysms or diverticula: Impact of age, gender, and ethnicity on prognosis

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Objectives: Congenital left ventricular aneurysm (LVA) and diverticulum (LVD) are rare cardiac anomalies. We sought to investigate the impact of age, gender, and ethnicity on the outcome in such patients.
Methods: Scientific databases and reference lists of relevant articles were searched for publications reporting on patients diagnosed with LVA/LVD. Full text articles were analysed for cardiac events (arrhythmic events, rupture, sudden cardiac death, congestive heart failure, cardio-embolic events, syncope, and change in size) during follow-up (FU).
Results: We identified 374 patients published since 1816 [185 (49.5%) LVA, 189 (50.5%) LVD] providing at least 1 year (y) FU. Mean age at diagnosis was 34.1 ± 27y (LVA) and 29.7 ± 27.6y (LVD; $p = 0.05$). 56.4% were male. Mean FU was 5.2 ± 4y (LVA) and 4.1 ± 3.1y (LVD). There was a trend for higher incidence of cardiac death in LVA patients (14.1% versus 7.9%; $p = 0.06$). Cardiac death was significantly more frequent in the younger (≤18y versus >18y) age groups (LVA: 22.0% versus 10.3%; $p = 0.04$, and LVD: 16.1% versus 1.9%; $p < 0.001$) Figure 1. Cardiac event rate per year (CER) differed with respect to type of anomaly (LVA/LVD) and age group (Figure 2). Symptoms (arrhythmia-related symptoms, syncope, and embolic events) at time of diagnosis increased the incidence of adverse events during FU in both groups (LVA: 35.4% vs. 11.6%; $p < 0.001$, and LVD: 22.6% versus 6.6%; $p = 0.004$). Black patients with LVA had a significantly increased CER compared to Caucasians (39.3% versus 14.3%; $p = 0.006$); gender was not predictive for cardiac events in LVA/LVD groups.

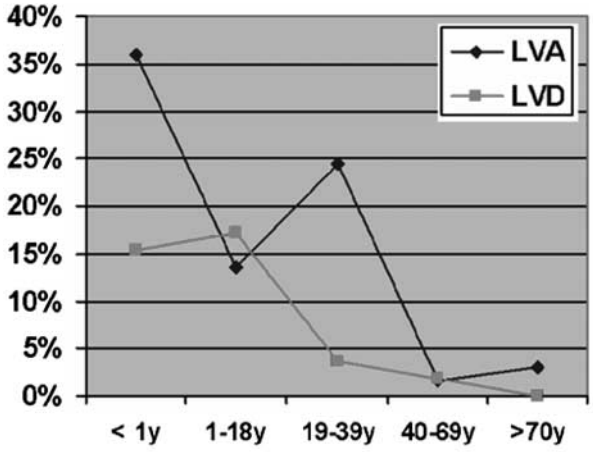

Fig. 1: Mortalityin LVA/LVD patientsrelated to age group

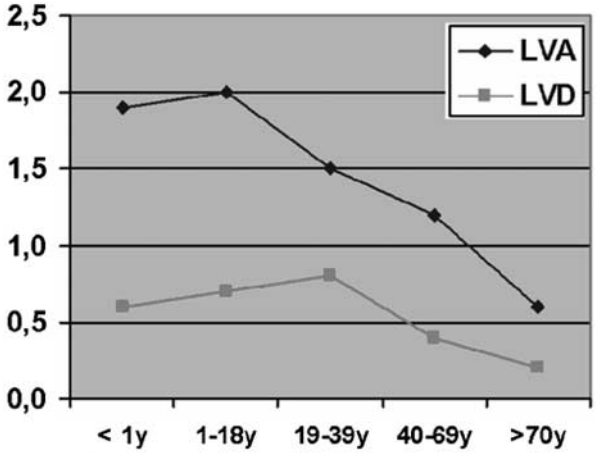

Fig 2: Eventrate per year in LVA/LVD patientsrelated to age group

Conclusions: Young patients (<18y), black patients, and symptoms at diagnosis were associated with worse outcomes in LVA or LVD.

P-114

Congenital left ventricular aneurysms or diverticula: Presentation, Characteristics, and Prognosis

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Objectives: Congenital left ventricular aneurysm (LVA) and diverticulum (LVD) are rare cardiac anomalies. We sought to investigate the clinical characteristics and outcome in all ever published patients.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Methods: MEDLINE, Web of science, Google and EMBASE, and reference lists of relevant articles were searched for publications reporting on patients diagnosed with LVA or LVD.
Results: We identified 664 patients published since 1816 [326 (49.1%) LVA, 336 (50.6%) LVD, 2 (0.3%) both]. Mean age at diagnosis was 34.1 ± 27 (LVA) and 29.7 ± 27.6 years (LVD; p = 0.05). 56.4% were male. LVA were larger (38.7 ± 22.5 mm versus 31.4 ± 21.2 mm; p = 0.002) and frequently found in submitral location (33% versus 4.9%; p < 0.001), LVD were frequently located at the LV-apex (61.2% versus 28.7%; p < 0.001). LVD were often associated with cardiac (34.2% versus 2%; p < 0.001) or extracardiac anomalies (32.7% versus 3%; p < 0.001). Pts with LVA presented more frequently with sustained ventricular tachycardia/ventricular fibrillation (16.6% versus 8.3%; p = 0.001), the incidences of rupture (4% versus 4.5%; p = 0.9), syncope (8.3% versus 5.1%; p = 0.1), and embolic events (4.9% versus 3.6%; p = 0.4) were not different between LVA and LVD. Mean follow-up was 56.3 ± 43 months. Cardiac death occurred more frequently in the LVA group (11% versus 5%; p = 0.05) at a mean age of 8.9 ± 11.6y and 14.1 ± 23.4y, respectively. The leading cause of cardiac death was congestive heart failure in the LVA-group (48% versus 0%: p = 0.03), and rupture in the LVD-group (75% versus 28%; p = 0.04).
Conclusions: Congenital LVA and LVD are two distinct entities with regard to clinical presentation, associated cardiac anomalies, anatomical location, and prognosis.

P-115
Enddiastolic right ventricular volume exceeding 160 ml/m² – a reasonable value for every Fallot patient regarding pulmonary valve replacement?

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Introduction: Body surface area is accepted and used as one possible means for normalization of right ventricular volumes (RVi); clinically it is the gold standard to define RV volume overload. We tested the hypothesis if greater variations of BSA have an influence on decision making of pulmonary valve replacement (PVR) in patients with Tetralogy of Fallot (TOF) after repair.
Methods: Retrospective analysis of CMR volumes from 80 TOF patients (group A: BMI 22.7 ± 4.1, BSA 1.77 ± 0.3 m², RV-EDVi = 138 ± 39 ml) as well as 20 healthy persons (group B: BMI 23 ± 4.5, BSA 1.83 ± 0.25 m², RV-EDVi = 85.2 ± 13 ml) older than 16 years. In a mathematical model both groups gained and lost weight (+20 kg, −10 kg). Changes of indexed volumes were compared using a Student's T-Test. Additionally a prospective CMR-investigation of 10 obese and 12 anorectic adult individuals with normal RV-function was performed and the results were compared with height-matched controls.
Results: With increasing or decreasing weight RV-EDVi-volumes changed significantly in group A (148 ± 42 ml/m² vs 123 ± 35 ml/m²) and group B (91.42 ± 13 ml/m² vs 76.2 ± 12 ml/m², p < 0.001). The number of patients with RV-EDVi > 160 ml/m² in Group A decreased from 16 to 6 by gaining weight and rose to 30 by weight loss. Comparison of anorectic (BMI 16.7 ± 3.6, BSA 1.5 ± 0.19 m², RV-EDV 124 ± 31 ml) and obese (BMI 33 ± 3, BSA 2.0 ± 0.18 m², RV-EDV 124 ± 29 ml) individuals to matched controls did not result in significant differences of not indexed RV-Volumes. RV volumes of TOF patients correlated more with height (r = 0.72, p < 0.01) than with weight (r = 0.38, p < 0.01).
Conclusions: Weight changes influence BSA and consequently indexed RV volumes affecting indication for PVR in TOF patients. Absolute RV volumes of anorectic, healthy and obese people do not differ much. Indexed volumes to an expected "normal" BSA or normalization of volumes to height should be used in those patients for decision making of PVR.

P-116
Evaluation of Contraceptive Methods in Women with Congenital Heart Defects in Germany, Hungary and Japan

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Introduction: The 2011 'ESC Guidelines on the management of cardiovascular diseases during pregnancy' recommend that maternal risk assessment for women with heart diseases is carried out according to the modified World Health Organization (WHO) risk classification. Particularly in women with congenital heart defects (CHD) who are at a higher pregnancy-related risk for cardiovascular complications or in which pregnancy is contraindicated an early and effective contraception is necessary.
Methods: In a multicentre cross-sectional questionnaire-based study, 634 women with CHD (Germany 61%, Hungary 24%, Japan 15%) were surveyed over a period of twelve months concerning contraception. Median age was 30 years. According to the modified WHO classification of maternal cardiovascular risk patients were grouped into three risk groups (low – medium – high/pregnancy contraindicated). The contraceptive methods (CM) used by each group were determined. In this study CM with a Pearl index ≤2 (at ideal use) was classified as 'safe'.
Results: In all three risk groups almost one third of the women was using a CM classified as unsafe. In 29% of all cases an unsafe CM was used. There has been no significant difference between the participating countries.

	Low risk n (%)	Medium risk n (%)	High risk n (%)	Total n (%)
CM safe	220 (72.1)	146 (69.5)	47 (70.1)	413 (71.0)
CM unsafe	85 (27.9)	64 (30.5)	20 (29.9)	169 (29.0)
Total	305	210	67	582

missing values = 52

Conclusions: Alarmingly, almost one third of the women with CHD and increased pregnancy-related risk of cardiovascular complications or contraindication for pregnancy is using a contraceptive method deemed as unsafe. A more efficient education regarding contraception in women with congenital heart defects is necessary.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

P-117

Asymptomatic patients with repaired tetralogy of Fallot from quality of life and exercise tolerance perspective

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Introduction: In early adulthood there is a high proportion of patients after tetralogy of Fallot (ToF) repair remain asymptomatic. These may have an impact in timing an intervention for a significant residual lesion. This study analyses the self-reported QoL and exercise tests results in asymptomatic patients after TOF repair.
Methods: In a prospective study, 47 consecutive patients with ToF had a 6 minutes walk test and a cardiopulmonary exercise test (CPT), and completed a SF-36 health survey questionnaire (health related QoL) in a same day. Asymptomatic patients were those in NYHA functional class I. SF-36 health dimensions in a scale 1–100, $\geq$80 were considered normal. Six-minute walk distance (6WD) $\geq$ 450 m, and predicted peak oxygen consumption (VO2%) $\geq$80 were also documented as normal values.
Results: At a mean age of 30 $\pm$ 8 years 70% (n = 33) were reported asymptomatic (NYHA class I). Forty-nine percent of them had >1 thoracotomy for heart surgery, and 18% a history of arrhythmia. Among the asymptomatic patients normal values were achieved in 97% for 6WD, 75% for SF-36 health dimensions, and only 21% for peak VO2%. NYHA class correlated with 6WD (r = 0.40. p = 0.009), however, did not correlate with SF-36 health dimensions or peak VO2%. The number of surgeries or the history of arrhythmia had no impact on patients self-reported QoL. There was no significant difference between normal SF health dimensions in patients with or without normal peak VO2% (70% vs. 75%, p = 0.25).
Conclusions: Only 20% of patients considered asymptomatic and/or with a good 6MWD result proved to have a good exercise capacity by CPT. In asymptomatic patients CPT may be a useful diagnostic tool in decision making before a therapeutic intervention. Overall self-reported QoL did not reflect the exercise tolerance.

P-118

Specialized care in complex ACHD patients: transfer to adult care

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Introduction: Adult congenital heart disease (ACHD) centers are created to assure the continuity of the pediatric care and to offer a specialized follow-up in this population. We aimed to describe the characteristics and changes during a decade of ACHD patients care.
Methods: We use our ACHD patient database (n = 1107), where active patients has been followed since 2000. Only complex patients were considered for further analysis. Clinical data, complexity of the diagnosis (moderate or severe), age at transfer to ACHD program, and lapse of care (>3 years) were documented.
Results: Sixty - nine percent of our active ACHD patients were considered complex in severity: moderate, n = 425, and severe, n = 350. The mean age at last follow-up was 31 $\pm$ 10 years, male/female ratio was 1.08. The mean age at transfer to adult care was 27 $\pm$ 6 years. In the last decade there was a major increase in the number of patients transferred to the ACHD program, in 2000 there were n = 80, and in 2012 n = 775 complex patients. The number of new patients/year for 1 physician in 2000 vs. 2012 vas 30 vs. 53 respectively. Lapse of care was registered in 15% of patients. During the study period the mortality among the complex patients was 1.7%.
Conclusion: In our tertiary ACHD center 2/3 of patients are complex in severity. The number of new patients/year has almost doubled during the last decade. Despite the disease complexity patients are still transferred to ACHD program very late. However, once they took part in the ACHD program the lapse of care among these patients is low.

P-119

Impact of fetal development on neurocognitive performance of adolescents with cyanotic and acyanotic congenital heart disease (CHD)

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Purpose: Our aim was to evaluate the neurocognitive performance in adolescents with CHD and to determine whether parameters of fetal development evaluated in neonates, such as head circumference, length, weight and Apgar scores, are somehow related to their neurocognitive performance.
Methods: 77 CHD patients (43 males) aged from 13 to 18 years old (mean = 15.04 $\pm$ 1.86), 46 cyanotic (23 Tetralogy of Fallot, 23 Transposition of the Great Arteries) and 31 acyanotic (Ventricular Septal Defect) enrolled in this study. The control group included 16 healthy children (11 males) ages ranging from 13 and 18 (mean = 15.69 $\pm$ 1.44). All assessment measures for CHD patients were once obtained in a tertiary hospital; the control group was evaluated in school. Demographic information and clinical history were collected. Neuropsychological assessment included Wechsler's Digit Test, direct and reverse (WDD, WDR) and Symbol Search (WSS), Rey's Complex Figure (RCF), BADS's Key Searching Test (BKS), Color-Word Stroop Test (CWS), Trail Making Test (TMT) and Logical Memory Task (LMT).
Results: CHD patients compared to control group showed lower scores in WDD (u = 262.500; p = 0.000) and WDR (u = 166.500; p = 0.000) versions, in RCF, copy (u = 152.500; p = 0.000) and memory (u = 149.000; p = 0.000), in WSS (u = 852.000; p = 0.016), in BADS's Key Searching Test (u = 160.500; p = 0.000) in CWS, words (u = 147.000; p = 0.000), colors (u = 225.000; p = 0.000) and interference (u = 133.500; p = 0.000) and in TMT, A (u = 1140.500; p = 0.000) and B (u = 1101.500; p = 0.000). Cyanotic patients, when compared to acyanotic, showed lower scores in all neuropsychological tasks, although the only differences that were significant were in RCF, copy (u = 935.500; p = 0.020), memory (u = 989.000; p = 0.004) in CWS, interference (u = 903,000; p = 0.048). Several correlations were apparent between fetal/neonatal parameters and neuropsychological abilities in each type of CHD. However, head circumference at birth stands as a main correlation with cognitive development later on in all kinds of CHD (WDD: rho = 0.339, p = 0.011; RCF, copy: rho = 0.297, p = 0.027; BKS: rho = 0.264, p = 0.051; CWS, interference: rho = 0.283, p = 0.036; TMT A: rho = −0.321, p = 0.036; LMT: rho = −0.263, p = 0.052).

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Conclusion: Adolescents with CHD have worse neuropsychological performance than the control group, mainly the cyanotic patients. Fetal circulation seems to have impact on cerebral and somatic growth, predicting cognitive impairment in adolescents with CHD.

P-120

Causes of late death in patients with congenital heart defects after paediatric cardiac surgery aiming at biventricular repair

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Objectives: The aim of the study was to identify those patient groups at risk for late mortality after surgery for congenital heart defects (CHD) aiming at biventricular repair by assessing the mode of their late deaths.
Methods: All 2132 patients operated for CHD aiming at biventricular repair at our institution before the age of 18 years between Jan 1st 1994 and Jan 1st 2009 were cross-checked against the Swedish National Population Registry on January 1st 2012 to reliably identify all dead patients. Of all 101/2132 (4,7%) deceased patients 70/2132 (3,3%) had died more than 30 days after the last surgery and 6/2132 (0,3%) had undergone a heart transplantation. 21/2132 patients (1,0%) were lost to follow-up (emigration). The circumstances of late death were analysed by reviewing clinical charts and autopsy reports. The mode of death was defined as the condition that initiated a clinical course of deterioration leading to death.
Results: The mode of death was considered to be CHD-related in 42 and most likely CHD-related in further 9 cases together accounting for 51/70 (73%) of late deaths. The 9 deaths without a definite cause occurred in patients with complex heart defects and in 7/9 patients suddenly in an ambulatory setting. In the majority of these 9 cases an arrhythmia is highly probable. The dominating mode of CHD-related deaths was ventricular failure in 20/42 (48%), which was in 11/20 (55%) due to pulmonary hypertension of different aetiologies including pulmonary venous stenosis (5) and AVSD or VSD in Down syndrome (3). 8/42 (19%) died because of a failing shunt-circulation (shunt-occlusion 2, circulatory failure 6). 19/70 (27%) of all late deaths were not related to the underlying congenital heart defect. Of all patients 49/70 (70%) had a syndrome, chromosomal and/or congenital defects.
Conclusion: Late mortality after surgery for CHD aiming at biventricular repair is low and in nearly one third of cases the deaths were not related to the underlying cardiac defects. Syndromes and relevant comorbidities were common complicating factors. The dominating modes of death were right heart failure and in patients with primary palliation a failing shunt circulation.

P-121

Assessment of quality of life in families with a child after staged treatment for hypoplastic left heart syndrome (HLHS) – one center experience

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Background: In spite of increasing number of survivors with HLHS after staged Norwood operation - data about quality of life in this group of patients and their families is limited. Our hospital is one of the biggest centers for HLHS treatment in Poland.
Aim: The aim of the study was to assess the quality of life of our HLHS patients and their families.
Methods: Mothers of 57 children with HLHS; 40 boys and 17 girls (age 4–16 years) completed the questionnaire to assess quality of their children's life and impact of child's illness on the family. All children were operated at our institution. In 20 families (35%) child with HLHS was the only child, in 32 families (56%) child with HLHS was the first child. HLHS was diagnosed prenatally in 21cases (37%) and in 18 (32%) the child was born at our institution.
Results: Limited physical activity in HLHS patients was reported in 80%, but only 8 patients (14%) attend to rehabilitation, emotional problems in 25%, educational in 9%. Development estimated as normal was reported in 89%; 79% of patients attend to normal schools or kindergartens. Good tolerance of frequent hospitalizations was reported in 75% of cases. Child illness is connected with strong parental stress (73%), and negative emotions like sadness (41%), fear and helplessness (42%). Own family support, support groups of parents and religious faith were considered most helpful. Only 7 (12%) mothers looked for professional psychological care. 94% responders assessed familial atmosphere as good, in 67% child's illness strengthened parental marriage. Impact of child's illness on family material situation was assessed as significantly negative in 79%. In 59% of families the father is the only working parent.
Conclusions: In majority patients with HLHS are active members of the society, they attend to normal schools and kindergartens although their physical activity is limited. The family functioning is good but child's illness is a reason of strong parental stress and indicates material problems. Increasing number of HLHS survivors indicates the need for continuation studies concerning neurodevelopmental outcome, quality of life and family functioning.

P-122

Infective endocarditis in adults with congenital heart disease

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Infective endocarditis (IE) may adversely impact on long term prognosis of patients with CHD reaching adulthood (ACHD). The aim of this study was to assess features and outcomes of IE in ACHD
Material and methods: Design is a single centre retrospective chart review of IE episodes in patients with CHD and aged >18y at diagnosis. Demographics, past cardiac history, clinical and echocardiographic, therapeutics data and outcomes were reviewed.
Results: From 1980 to 2011, 33 patients were included, mean age 29 years at IE (range 18 to 76). Underlying CHDs were: native VSD in 22%, cyanotic CHD in 35%, AVSD in 6%, aortic valve lesion in 22% and miscellaneous in 10%. CHD was repaired in 19%, palliated in 27% and non-operated in 54%. Thirty percent had received antibiotics prior to IE diagnosis. Heart failure occurred in 22% of cases, septic shock in 11%, neurological complication in 11%, splenomegalia was present in 46% and fever in 100% of cases. Source of infection was dental in 35%,

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

cutaneous in 25%, ENT in 5%. The microbial causal agent was Staphylcoccus in 46% and streptococcus in 32.5%, unknown in 8%. Echocardiographic vegetations were found in 49% of the cases, valves perforation or abscesses occurred in 11% and 24%. Embolic events were frequent (62%). Surgery was performed in 30% of cases, a median of 21 days after onset of IE (1 day to 5 months). Hospital stay was 2 weeks to 6 months. Mortality was 11% and 2 cases recurred.
Conclusion: IE severely impacts on prognosis of ACHD, especially in patients with cyanotic CHD. Embolic events frequently complicate outcomes. Prophylaxis should mainly focus on cutaneous and dental events.

P-123
Limited Value of N- Terminus-Pro-B-Type Natriuretic Peptide Levels in Former Types of Fontan circulation
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Objectives: N-Terminus pro-B-type natriuretic peptide (NT-proBNP) is an important biomarker in congestive heart failure. This has also been confirmed in congenital heart disease. However its clinical value in patients with different types of Fontan circulation remains questionable.
Methods and Results: We prospectively analyzed 124 patients with various types of Fontan surgery between October 2006 and February 2011. We included 49 patients with older Fontan modification (atriopulmonary - and atrioventricular connection [APC/AVC]) and 75 patients with total cavopulmonary connection (TCPC). NT-proBNP levels of patients with APC/AVC were significantly higher than in patients with TCPC ($p = <0.001$). NT-proBNP levels positively correlated with atrioventricular valve regurgitation and ventricular dysfunction only in patients with TCPC, but not in patients with APC or AVC.
Conclusion: NT-pro BNP levels are related to the type of Fontan circulation. The older types (APC/AVC) that involve more atrial tissue in the systemic venous pathway show higher NT-proBNP level. Therefore, NT-proBNP is only an important biomarker to evaluate patients after TCPC. Its clinical value is limited in patients with older Fontan modification.

P-124
Management and Outcome of Patients with Double-Chambered Right Ventricle – Experience from Two Tertiary Adult Congenital Heart Disease Centers
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Background: Double-Chambered Right Ventricle (DCRV) is a rare form of congenital heart disease, in which the RV is progressively separated into a high- and low-pressure chamber. So far, there is no consensus on the optimal management of DCRV in adult patients. Here we report the combined experience with DCRV patients at two tertiary Adult Congenital Heart Disease Centers.
Methods and Results: All patients with DCRV under follow-up at the centers were identified from computerised databases. A retrospective review of medical records was carried out on all 53 identified patients (34 female, age 40.6 ± 15.0 at the time of data analysis, range 17 to 69 years). Almost all patients (96%) had ventricular septal defects (VSD) as an underlying diagnosis; predominantly in the perimembranous portion of the septum (80%). Seven patients had undergone VSD closure during childhood but continued to have a restrictive VSD. Eight patients were completely asymptomatic and were managed conservatively so far (median age 26 years, 34.4 ± 26.2, 19 to 63) with no fatalities. The remaining patients developed symptoms at a median age of 26 years (shortly after birth to 68 years), most commonly shortness of breath and reduced exercise capacity (56%), palpitations (13%) and chest pain (13%). Investigation by echocardiography revealed a mean intra-chamber gradient of 69.4 ± 30.0 mmHg. Surgical relief of the right ventricular obstruction was undertaken in 36 patients (68%, median age at operation 26.7 years, 28.3 ± 20.3, 14 months to 67 years). There was no early or late operative mortality, no patient required reoperation for DCRV during follow-up and 86% of operated patients showed no residual intra-ventricular gradient postoperatively. Symptoms improved significantly after surgery with only 8 patients remaining in NYHA class 2 (mean follow-up time 10.5 ± 9.4 years).
Conclusions: Contemporary DCRV patients, managed at tertiary congenital centers, have good survival prospects and low long-term morbidity. Cardiac surgery is inherently low risk, associated with good long-term haemodynamic and functional results and a proactive treatment approach therefore appears warranted.

P-125
Pregnancy Outcomes and Aortic Dimensions in Repaired Aortic Coarctation
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Introduction: Aortic coarctation has an incidence of 1 per 5000 livebirths and represents 5–10% of all congenital cardiac lesions. With modern management, prognosis is good and most women will survive to childbearing age. There is, however, concern that pregnancy may result in significant morbidity and mortality. Recent studies report hypertension related complications and aortic dissection but little is known about the effect of pregnancy on aortic dimensions.
Methods: All pregnancies in women with repaired coarctation managed by the Newcastle upon Tyne Hospitals Trust over the last decade were identified. Those who had had undergone cardiac magnetic resonance (MR) before and after pregnancy were included in this study. Clinical and obstetric data was obtained and MR scans were reviewed to determine dimensions of the aortic root, ascending aorta and descending aorta before and after pregnancy.
Results: 22 women with repaired coarctation were identified to have been pregnant. Of these, nine women (age 27.5 yrs, all end to end anastomoses) with ten pregnancies underwent serial MR and are the subjects of this study. Three women had bicuspid aortic valves and three had pre-existing dilatation of the ascending aorta (one patient had both). One woman had essential hypertension. Five women were delivered by Caesarean section (three emergency), four had instrumental delivery and two had normal vaginal delivery. One woman was delivered at 35 weeks for intrauterine growth restriction. All others delivered at term.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

The mean duration of the first, second and third stages of labour were 10 hours 54 minutes, 2 hours 28 minutes and 6 minutes respectively. Mean birth weight was 3482 ± 487 grammes. Two women required blood transfusion post-partum. MR performed at 24 months before and 20 months after pregnancy showed no change in dimensions of the aortic root at the sino-tubular junction (30.3 v 33.1 mm $p = 0.41$), ascending aorta (29.0 mm versus 29.5 mm $p = 0.85$) or descending aorta (19.4 mm versus 18.8 mm $p = 0.81$).
Conclusion: In this small cohort, obstetric outcomes were good. A high operative delivery rate was observed which is in keeping with reports in larger series. Aortic dimensions did not significantly change following pregnancy, however, further prospective data collection is warranted.

P-126
Cardiac outcomes in adolescent and adult patients with Down syndrome and congenital heart defect

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Background: Trisomy 21 is the most common single chromosome abnormality at birth and is frequently associated with congenital heart disease (CHD). Advances in diagnosis and management of patients with CHD have also involved patients with Down's syndrome and CHD leading them to survive to adulthood. However, few studies have focused on cardiac outcomes in adult patients with Down's syndrome and CHD.
Aims: to determine cardiac outcomes in adolescent and adult patients with Down's syndrome and CHD.
Methods: We determined the late survival in all patients (≥15 years old) with Down's syndrome and CHD. Then, we examined cardiac outcomes in the subsets of patients with trisomy 21 and 1/with repaired atrioventricular septal defect (AVSD) (n = 33) and 2/with Eisenmenger syndrome (n = 71). We evaluated the impact of Down's syndrome on the results by comparing cardiac outcomes between patients with Down's syndrome and non-Down patients.
Results: Survival of 136 adult and adolescent patients with Down's syndrome and CHD was 58.2% at 40 years. In the 33 patients with repaired AVSD, late complications occurred in 12% and were essentially infective endocarditis. Although actuarial survival after AVSD repair was not different between patients with Down's syndrome and group with normal karyotype (n = 50), clinically significant cardiac lesions were more common in no-Down patients (68% vs 27%, $p < 0.02$), including arrhythmia, anomalies of mitral valve, and heart failure. In Eisenmenger patients, survival was significantly impaired in Down's syndrome compared to patients with normal karyotype (n = 118): survival at 40 years was respectively 51.9% vs 67.5% ($p < 0.0001$, logrank test). Eisenmenger patients with complex anatomy had a significant worse prognosis when compared with those with simple anatomy ($p < 0.0001$, logrank test), independently of karyotype, because Down syndrome patients had more complex heart defect, which is AVSD commonly associated with trisomy 21 (86% vs 61%, $p < 0.001$).
Conclusion: Adults and adolescents patients with Down syndrome have better long-term outcomes after AVSD repair than patients with normal karyotype, but Eisenmenger patients with Down syndrome have a markedly reduced survival, compared with no-Down patients, due to the complex CHD commonly associated with trisomy 21, which is AVSD.

P-127
Longitudinal follow-up study of patients with transposition of great arteries after Senning operation

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Objectives: Longitudinal observational study of hemodynamic and functional parameters of patients after Senning operation of transposition of great arteries (d-TGA).
Methods: 50 patients who underwent surgery between 1984–1997 at a mean age of 5.8 ± 2.3 months were examined (clinical examination, echocardiography, magnetic resonance imaging, 24-hour ECG monitoring, exercise stress testing) at the age of 13.6 ± 3.6 (1st examination) and 22.6 ± 3.6 years (2nd examination).
Results: Ejection fraction (EF) of the systemic right ventricle (RV) was slightly reduced (median 53.2, IQR 45.6–62.2% at 2nd examination) as compared to normal limits for a left ventricle. Maximum oxygen consumption during exercise (VO2max) was significantly decreased. The heart rate (HR) profile was shifted towards bradycardia in 44/50 patients without pacemaker. The degree of tricuspid insufficiency (TI) and functional parameters (NYHA and VO2max) did not deteriorate during longitudinal follow-up. Slight but significant increase of average HR was observed (see Table for data). There was no correlation between VO2max and EF of the systemic RV, tricuspid insufficiency or maximum HR during exercise. A trend towards reduction of VO_2max with increasing BMI ($R = 0.293$, $P = 0.083$) was noted.

Examination	TI grade mean (SD)	VO_{2max} z-score mean (SD)	HR_{min} z-score median (IQR)	$HR_{average}$ z-score median (IQR)	HR_{max} z-score median (IQR)	NYHA class median (IQR)
1.	1.0 (0.7)	−2.5 (1.4)	−1.0 (−1.3/−0.4)	−1.0 (−1.8/−0.4)	−1.2 (−2.2/−0.5)	1.0 (1.0/2.0)
2.	1.0 (0.7)	−2.1 (0.9)	-0.9 (−1.4/−0.5)	−0.9 (−1.4/−0.1)	−1.0 (−2.2/−0.2)	1.0 (1.0/2.0)
P	NS	p = 0.009	NS	p = 0.014	NS	NS

Conclusions: Functional cardiovascular parameters are steadily reduced in longitudinal follow-up of patients after the Senning operation and do not correlate with RV systolic function. Influence of other factors (diastolic dysfunction, BMI) is presumed.
Supported by the project of Internal Grant Agency of Ministry of Health, Czech Republic: NT 13408-4/2012. Supported by the project (Ministry of Health, Czech Republic) for conceptual development of research organization 00064203 (University Hospital Motol, Prague, Czech Republic).

P-128

Prevalence of Arterial Hypertension in Patients with Corrected Aortic Coarctation: Impact of the Concomitant Presence of Complex Congenital Heart Disease

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Introduction: The development of arterial hypertension is a known complication in patients after aortic coarctation (CoA) repair. The current study was designed to evaluate the difference in prevalence of arterial hypertension among patients with previous surgery for CoA. Population was dichotomized according to the presence of isolated CoA (iCoA) vs CoA associated with complex congenital heart disease undergoing biventricular correction (cCoA).
Methods: Patients with signs of kidney disease and/or an arm-to-leg gradient at rest >10 mmHg or/and echocardiographic evidence of recurrent obstruction at the aortic arch, were excluded. We selected 235 children and the data were retrospectively analyzed after dividing them into two groups: 1) 148 iCoA, and 2) 87 cCoA. Patients were defined as hypertensive in the presence of antihypertensive treatment and/or when blood pressure at rest and/or during 24 hour ambulatory blood pressure monitoring (ABPM) was above 95th percentile, respectively for age and height (Pediatrics 2004, 114 (S): 555) and for height (J Pediatrics 1997;130:178).
Results: Patients with iCoA were significantly older than patients with cCoA (22 ± 7 yrs vs 16 ± 7 yrs; $p < 0.001$), with a markedly higher prevalence of arterial hypertension (43% vs 20%). Difference in the prevalence of hypertension remained significant also in analysis of covariance correcting for differences in age among groups ($p < 0.001$), demonstrating a risk of developing hypertension for patients with cCoA reduced by more than a half as compared to iCoA (odds ratio 0.48).
Conclusion: In the presence of CoA the association with complex congenital heart disease results in a significant reduced prevalence of hypertension. Systemic low flow and pressure in patients with cCoA might be associated with a lower rate of arterial hypertension, through the lack of stimulation of baroreceptor reflex in the prestenotic area, as speculated for the iCoA. Further studies are necessary to confirm our findings.

P-129

Normal left ventricular function in long-term follow-up in 90 patients with a history of Kawasaki disease

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Introduction: Kawasaki disease (KD) is an acute pediatric vasculitis that is associated with the development of coronary artery aneurysms (CAAs). Myocarditis is another well-known cardiac manifestation. Although histopathological studies suggest persistent myocardial abnormalities after KD, the long-term effects on cardiac function remain to be revealed. The aim of this study was to evaluate left ventricular (LV) function during KD follow-up using Tissue Doppler Imaging (TDI), and to compare the results with healthy controls.

Table 1. LV parameters in patients with history of KD (n = 90) and controls (n = 63).

	Patients	Controls	p-value*
Systolic			
Fractional shortening (%)	36 ± 5	36 ± 5	.621
TDI IVS S' (cm/s)	7.6 ± 1.0	7.6 ± 1.0	.638
Diastolic			
MV E velocity (m/s)	0.95 ± 0.15	0.97 ± 0.17	.415
MV A velocity (m/s)	0.48 ± 0.11	0.48 ± 0.10	.637
MV E/A ratio	2.09 ± 0.51	2.10 ± 0.52	.956
TDI IVS E' (cm/s)	13.4 ± 1.8	13.7 ± 2.1	.324
TDI IVS A' (cm/s)	5.7 ± 1.0	5.7 ± 1.5	.890
TDI IVS E/E'	7.1 ± 1.3	7.1 ± 1.4	.893

* Differences between patients and controls were analysed using independent sample t-tests.

Methods: Patients with a history of KD (aged 4–18 years) were included. All subjects underwent 2-dimensional transthoracic echocardiography at least 6 months after the disease onset. The examination assessed LV systolic performance (fractional shortening) and diastolic performance (Doppler flow over the mitral valve). Additionally, peak velocities of systolic (S), early diastolic (E), and late diastolic (A) motion of the intraventricular septum (IVS) were measured using TDI in apical 4-chamber view. For control subjects the same measurements were performed.
Results: Ninety patients (mean age 11.4 ± 4.1 years, 68% male) and 63 controls (mean age 10.5 ± 4.1 years, 57% male) were included. The mean interval from KD onset to echocardiography was 7.8 ± 4.6 years. The LV parameters are given in Table 1.
Conclusion: LV function is normal after long term follow-up in patients with a history of KD.

P-130

Normal systolic and diastolic left ventricular function in 27 untreated pediatric Fabry disease patients – preliminary results

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Introduction: Fabry disease (FD) is an X-linked, lysosomal storage disorder that may present itself during childhood with acroparesthesia, hypohidrosis and (micro)albuminuria. At a later age, patients often develop left ventricular hypertrophy (LVH). In young adults without LVH, diastolic dysfunction has been identified as an early sign of cardiac involvement.
Aim: We assessed the cardiac status of paediatric FD patients in order to gain insight in the age of onset of cardiac involvement.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Table 1. Mean values and standard deviations (SD)

	Control	Mean (SD)	FD	Mean (SD)
IVSd (mm)	5.71	(0.98)	6.56	(1.16)
LVPWd (mm)	6.18	(1.13)	6.57	(1.17)
LVED (mm)	44.48	(5.73)	47.53	(4.64)
LVES (mm)	28.59	(4.75)	30.23	(4.50)
Fractional Shortening %	35.87	(5.98)	36.48	(5.96)
E-wave peak velocity (cm/s)	96.48	(17.07)	99.33	(16.79)
E-wave dec. time (ms)	170.32	(28.75)	172.62	(33.38)
A-wave peak velocity (cm/s)	47.16	(9.72)	45.85	(9.16)
A-wave duration (ms)	100.60	(15.48)	95.15	(10.87)
E/A ratio	2.17	(0.55)	2.27	(0.54)
IVS-E (E') (cm/s)	13.79	(2.18)	13.24	(1.82)
E/E' ratio	7.07	(1.35)	7.54	(1.10)

Table 2. Linear regression For FD and Controls

	Mean difference: adjusted for age and gender.			
	Mean difference	95% Confidence interval		p-value
IVSd	0,53	0,07	0,99	0,02
LVPWd	0,04	−0,43	0,51	0,86
LVED	−0,13	−1,62	1,37	0,87
LVES	−0,53	−2,30	1,24	0,55
Fractional Shortening %	0,82	−2,18	3,82	0,60
E-wave peak velocity	2,73	−5,65	11,11	0,52
E-wave dec. time	−2,69	−17,50	12,12	0,72
A-wave peak velocity	−0,79	−5,49	3,92	0,74
A-wave duration	−8,20	−14,84	−1,56	0,02
E/A ratio	0,05	−0,23	0,32	0,73
IVSE	−0,77	−1,77	0,23	0,13
E/E' ratio	0,60	0,00	1,21	0,05

Abbreviations for table1 and 2.
IVSd: intraventricular septum diastole; LVPWd: left ventricular posterior wall thickness diastole; LVED: left ventricular end diastolic dimension; LVES: left ventricular end systolic dimension

Methods: We investigated cardiac function in 27 genetically confirmed FD patients and controls with conventional echocardiography and tissue Doppler imaging (TDI) on the VIVID 7 (GE). All examinations were performed before initiation of enzyme replacement therapy (ERT) and below age 18. All included patients had a genetically confirmed and certain diagnosis of FD. Data were analyzed with linear regression to adjust for confounding factors age and gender, using SPSS (IBM, version 20). Left ventricular mass index (LVM) was calculated using the Devereux formula.
Preliminary results: Twenty-seven (12M, 15F) FD patients (median age 14 years, range 5–17), and 58 (46 M, 33 F) healthy controls (median age 11.5 years, range 5–18) were included. Valve abnormalities were infrequent and only comprised of grade 1–2 mitral or tricuspid insufficiency. One patient had a bicuspid aortic valve. None of the FD patients had LVH, the intraventricular septum (IVSd) was significantly increased in FD patients; left ventricular function was in the normal range. The mean values and standard deviations are presented in Table 1. The regression coefficient, mean differences [95% Confidence intervals] and p-values between the FD patients and controls, for M-Mode, Mitral Valve, Doppler and TDI parameters are presented in Table 2.
Discussion: In this large group of paediatric FD patients, there were no signs of diastolic dysfunction. The IVSd was increased, while there was no LVH according to the calculated LVM. The results of this study may contribute to the discussion on early initiation of ERT and the need for cardiological follow up at a young age.

P-131
The Evaluation of Cardiac Functions by Tissue Doppler Imaging in Childhood Leukemia Survivors in Remission

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Objectives: Improvement in long-term survival in patients with acute childhood leukemia has emerged the need to monitor chemotherapy related morbidity and mortality. The aim of this study was to evaluate cardiovascular status of children with acute leukemia and in remission, by using tissue Doppler imaging (TDI).
Methods: Sixty patients diagnosed with acute leukemia and in remission for at least two years and 30 healthy children were evaluated by conventional echocardiography and TDI.
Results: The median age of patients was 11.7 (10–14.9) and remission time was 4 (2.5–5) years. The median EF was found to be significantly reduced in patients than in control group (69 vs 73%), although EF of all patients were in normal range. Myocardial velocities were significantly lower in patients at basal segments of the myocardium. The median Sm velocity was 6 vs 8 cm/s at interventricular septum (IVS), 6 vs 8 cm/s at left ventricle (LV), and 10 vs 12 cm/s at right ventricle (RV). The median Em velocity was 12 vs 15 cm/s at IVS, 15 vs 17 cm/s at LV, and 15 vs 16 cm/s at RV. The median Am velocity was 5 vs 7 cm/s at IVS, 6 vs 7 cm/s at LV, and 7 vs 10 cm/s at RV. TDI also revealed significantly shortened ICT at all basal segments and IRT at basal IVS in patients. The median ICT was 70 vs 80 ms at IVS, 70 vs 92 ms at LV, and 63 vs 87 ms at RV. The median IRT was 59 vs 63 ms at IVS.
Conclusions: The significant decreases in myocardial velocities Sm, Em and Am at all segments are the signs of decreased systolic contraction, delayed relaxation and restriction of the myocardium, respectively. However, the significant shortenings in ICT and IRT do not suggest an obvious systolic and/or diastolic dysfunction. The periodic assessment of leukemia survivors for cardiac disease is mandatory because of the risk of symptomatic cardiac disease years after chemotherapy. TDI seems to give more and early information than conventional methods.

P-132
Myocardial Geometry and Functions in Children on Recombinant Human Growth Hormone Therapy: from Baseline to 12th Month

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Objectives: This study was designed to investigate the effects of recombinant human growth hormone (rhGH) therapy on myocardial geometry and functions in children with idiopathic

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

isolated growth hormone deficiency (GHD) by conventional echocardiography and tissue Doppler imaging (TDI).
Methods: Thirty patients (19 boys and 11 girls) with a diagnosis of idiopathic isolated GHD were enrolled in this study. The mean age of patients were 11.0 ± 2.6 (6.3–15.5) years. At baseline, 3rd, 6th and 12th month of treatment, the structure of left ventricle (LV) was assessed by conventional echocardiography, and myocardial rates and time intervals were assessed by TDI. By using these data; LV mass index (LVMI) by two different methods (LVMI1: g/m^2 and LVMI2: g/m2.7), relative wall thickness [RWT: (IVSd + LVPWd)/LVDd] and myocardial performance index (MPI: ICT+IRT/ET) for LV, IVS and RV were calculated.
Results: LVMI1, LVMI2 and RWT increased at 3rd, 6th and 12th month according to baseline (Table 1), and differences were significant for both LVMI1 and LVMI2 after 6th month, and for RWT at 12th month. There were no significant differences for MPI at LV, IVS and RV at 3rd, 6th and 12th month according to baseline (Table 2).
Conclusions: The results of this study showed that the rhGH therapy changes LV mass after 6th month and LV geometry after 12th month. However it does not affect both systolic and diastolic functions of the myocardium.

P-133
Assesment of Ventricular Functions of Patients with Pulmonary Hypertension by Standard and Pulsed Wave (PW) Tissue Doppler Echocardiography: Before and After Medical Treatment
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Aim: Our aim is to evaluate right and left ventricular myocardial functions by Standard and pulsed wave tissue Doppler echocardiography (TDE) before and after medical treatment of patients with pulmonary arterial hypertension (PAH).
Method: The study included 23 patients with Eisenmenger syndrome associated with congenital heart disease (13 with ventricular septal defect (VSD), 3 with patent ductus arteriosus (PDA) and atrial septal defect (ASD), 2 with atrioventricular septal defect (AVSD), 2 with ASD and VSD, 2 double outlet right ventricle (DORV), 1 with ASD) and 20 healthy control subjects. All patients underwent standard and tissue Doppler echocardiography. Regional parameters were evaluated by the measurement of three different myocard segment (basal lateral wall of right and left ventricle and basal septum) apical four chamber view with PW tissue Doppler. Patients World Health Organization (WHO) functional class, 6-minute walk distance (6MWD), and systemic arterial oxygen saturations (SaO2) and echocardiographic parameters of all patients were measured before treatment and during the treatment course.
Results: Before treatment patients with PAH had significantly greater right ventricular end diastolic diameter, pulmonary artery diameter, mean pulmonary artery pressure, Tricuspid annuler plane sistolic excursion (TAPSE) and right ventricular ejection fraction were significantly decreased when compared with the control group (3.78 ± 0.98 versus 2.78 ± 0.31 cm, 2.9 ± 0.37 versus 2.4 ± 0.15 cm, 63.2 ± 11.6 versus 14.5 ± 2.8 mm Hg, 1.68 ± 0.18 versus 2.0 ± 0.1 cm, % 31.4 ± 9.8 *verssus* % 56.3 ± 4,7 respectively; all $p < 0.05$).
Isovolumetric relaxation time (IVRT) and myocardial performance index (MPI) were higher in patients with PAH compared with controls (79.1 ± 18.7 versus 48.5 ± 7.2 sn, 0.62 ± 0.9 versus 0.32 ± 0.01 respectively; all $p < 0.05$). After treatment functional capacity and 6MWD were significantly higher and MPI were significantly lower at 6th and 9th months (448 ± 92.4 and 445 ± 62.2 to 361 ± 113 m, 0.52 ± 0.08 to 0.49 ± 0.08 respectively; all $p < 0.05$).
Conclusions: Right and left ventricular myocardial diastolic function was significantly decreased in patients with severe PAH. Short time after treatment functional capacity, 6MWD and MPI of both ventricles improved significantly. This study suggests that TDE may be useful for estimating prognosis and subclinic changes according to severity of PAH.

P-134
Evaluation of left ventricular function by echocardiography, tissue Doppler imaging and carotid intima-media thickness in obese adolescents with non-alcoholic fatty liver disease
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We aimed to evaluate left ventricular (LV) systolic/diastolic functions in obese adolescents with nonalcoholic fatty liver disease (NAFLD) by conventional echocardiography and pulsed-wave tissue Doppler imaging (PW-TDI) and investigate relationships between carotid intima-media thickness (CIMT) and LV function. 181 obese adolescents and 68 healthy controls were enrolled for the study. LV enddiastolic, end-systolic, left atrial (LA) diameters, left ventricular mass (LVM) were higher in both obese groups when compared with controls. By PW-Doppler echocardiography and PW-TDI, NAFLD group had normal LV systolic function, impaired diastolic functions and decreased global systolic and diastolic myocardial performance. In NAFLD patients, LVM were positively correlated with HOMA-IR and serum alanine aminotransferase (ALT). CIMT were positively correlated with HOMA-IR, ALT and LVM. By multiple stepwise regression analysis, ALT (β: 0.124; $p = 0.026$), HOMA-IR (β: 0.243; $p = 0.0001$), LVM (β: 0.874; $p = 0.0001$) were found as independent parameters associated with increased CIMT. Multivariate regression analysis revealed HOMA-IR as an independent predictor of increased LVM with an odds ratio of 1.4 and BMI as an independent predictor of increased CIMT with an odds ratio of 10. In conclusion, we suggest to use PW-TDI to detect early LV dysfunction at an earlier stage in obese adolescents with NAFLD for careful monitoring of cardiovascular risk.
We showed that insulin resistance have a significant independent impact on both CIMT and on LV remodelling in the absence of diabetes in NAFLD patients.

P-135
New MRI-Scanner – New Reference Values? Can We Transfer 1.5 T Reference Values To The 3 T Era?
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Introduction: Cardiac MRI is important in the treatment of children with congenital heart disease. It is reference standard for

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

the assessment of ventricular dimensions and function. Stronger magnetic fields increase signal to noise ratio, allowing measurements with high resolution. Field inhomogeneity leads to poor quality pictures. Most published reference values were obtained by 1.5 T MRI scanner.
Methods: Quantitative volumetric cardiac MR measurements were performed on a 3T TX MRT (Philips ACHIEVA) and a 1,5T MRT (Philips Intera) using a multi slice multi-phase steady-state free precision gradient-echo acquisition in breath hold (TR/TE/flip = 2.9 ms/1,45 ms/40°; Matrix = 1.4–1.5×1.5–1.7 mm^2; 22–30 phases, 5 mm slice thickness). Patient sample included 17 healthy Persons (7 male, 10 female, mean age 13.5 y ± 4.3a; range 6–20a). Calculated stroke volume was controlled by flow derived stroke volumes using phase-contrast MRI. Data were quantified by single expert.
F-Test and unpaired T-Test was performed.
Results: There were no significant differences for both, left end right ventricle.

	LV-EDV	LV-ESV	RV-EDV	RV-ESV
3 T	79,2 ± 14,6	29,0 ± 5,4	78,7 ± 16,1	30,5 ± 7,4
1.5 T	82 ± 12,4	29,8 ± 6,9	80,9 ± 13,6	30,9 ± 8,1
P-Value	0,52	0,7	0,66	0,88

All volumes indexed for BSA [ml/m^2]. RV = right ventricular, LV = left ventricular, EDV = end-diastolic volume, ESV = end-systolic volume

Conclusions: There is no relevant difference in ventricular size and calculated stroke volume by using a 1.5 T or 3 T MR scanner.

P-136
Quantification of tissue Doppler, Strain and Strain rate normal values in a population of healthy newborns at term
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Background: The first aim of the study was to quantify the normal values of tissue Doppler echocardiography (TDI), strain (S) and strain rate (SR) parameters to evaluate systolic and diastolic function in a sufficiently representative population of healthy infants born at term. The second aim was to standardize a protocol to obtain and analyze off line measurements from new advanced imaging techniques (TDI, S and SR derived from speckle tracking).
Methods: We prospectively enrolled 68 healthy infants (41 male) admitted to the neonatal center of Bergamo Ospedali Riuniti. The echocardiographic evaluation was performed before the first 72 hour of life. The TDI was acquired in the apical 4 rooms view placing the right ventricle and then the left ventricle in the center of the screen. The frame rates were always kept more than 200 FPR. Three cardiac cycles were recorded for offline analisis. The strain and strain rate were obtained with speckle tracking method (2DSTE). The grayscale images were acquired at frame rates of 65 to 85 FPR, optimizing them to precisely define the endocardium.
Results: The systolic function evaluated through systolic peaks waves is inferior in the left ventricle (LV) compared to the right (RV). The pulsed wave transmitral flow descrive a diastolic dysfunction of first degree. We obtained lower S and SR normal value in the LV than in the RV. The data showed a significant regional variability in the same ventricle and between the two: the longitudinal S was higher in the apical segments than in the basal of LV; higher longitudinal S and SR in the RV than in the LV.
Conclusion: This study was the first that prospectively evacuate TDI, S, SR in a healthy population of newborns using color TDI and 2DSTE tecniques. Moreover, 2DSTE itself was used in an extensive way on newborns for the first time and we standardized a protocol for the echocardiographic laboratory

P-137
Safety, feasibility, and diagnostic value of cardiac magnetic resonance imaging in patients with congenital heart disease and MRI-conditional pacemaker systems
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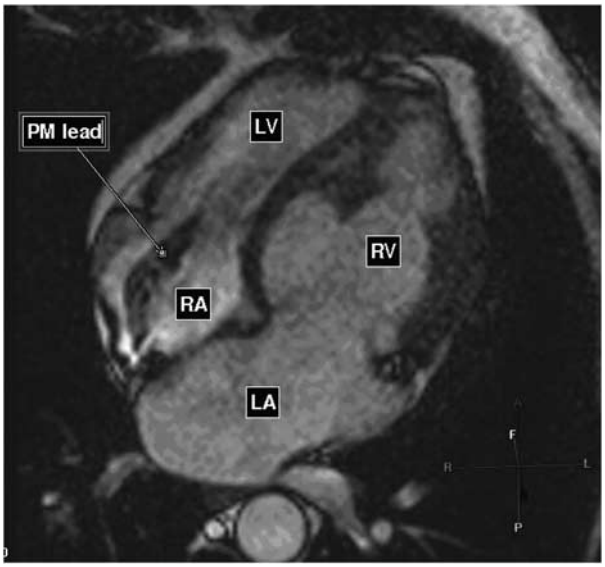
Introduction: In CHD patients, cardiac MRI (cMRI) is increasingly becoming the modality of choice for functional and anatomical imaging. With the recent introduction of MRI-conditional pacemaker systems (PMs), cMRI could also become accessible for CHD patients with PMs. Our aim was to analyse safety, feasibility and diagnostic value of cMRI in patients with CHD and MRI-conditional PMs.
Methods: CHD patients with MRI-conditional PMs and a clinical need for cMRI were examined with a 1.5 T MRI system (Philips Medical Systems, Best, the Netherlands). Specific absorption rate (SAR) was kept below 2 W/kg. Lead function was tested before and after cMRI. Image quality (IQ), relating to artefacts caused by device and PM leads, and diagnostic value (DV) were evaluated for each examination according to Naehle et al.[1] by 2 experienced investigators in consensus using a 4-point grading scale.



Results: Six patients (mean age 32.9 years, range 19.5–51.3 years) underwent a total of eight cMRI examinations. Diagnoses were dTGA after atrial redirection (n = 2), TOF (n = 1), DORV with PS (n = 1), ccTGA (n = 1) and TAPVC after anatomical repair (n = 1). Pacing indications were sinus node dysfunction (n = 4) and complete AV-block (n = 2). Patients did not note any PM-related sensations during cMRI, and there was no PM dysfunction. Device parameters did not change significantly compared to pre-cMRI. IQ was sufficient for evaluation of clinical questions in all cases (Figure 1, 4-chamber view, ccTGA).

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

In patients with systemic RV, ventricular function could be calculated accurately. In the TOF patient, cMRI results led to consecutive RPA stenting due to peripheral branch stenosis. Comparing the results of two cMRI examinations in the DORV patient, planned percutaneous pulmonary valve implantation could be postponed due to stable RV volumes.
Conclusions: CMRI can be safely performed with relatively good IQ and DV in CHD patients with MRI- conditional PMs. In patients who need implantation or revision of transvenous PMs, switching to MRI-conditional systems allows for application of cMRI for non-invasive monitoring of CHD.
Reference: 1. Naehle et al. Am Heart J 2011;161:1096-105.

P-138
Sports Related Alteration In Left Ventricular Geometric Patterns Among Children And Adolescent Athletes
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Introduction: Aim of this study is to determine left ventricular geometry of athletes by echocardiography in various types of sports.
Methods: Athletes from 5 different sports clubs (basketball, swimming, football, wrestling, and tennis) who regularly practice at least 3 hours a week for at least 2 years were included in this study. 98 (79%) were male and 26 (21%) were female. The control group consisted of sedentary children and adolescents (n: 25). Athletes were also grouped according to the type of exercise; dynamic (football, tennis), static (wrestling), combined (basketball, swimming). Athletes who had systemic hypertension, diabetes mellitus, chronic renal failure, chronic respiratory disease and a history of anabolic steroid use were also excluded. Left ventricular (LV) hypertrophy was considered if the LV mass index (LVMI) was over 95.th percentile by age and sex. LV geometry was classified according to LVMI and relative wall thickness (RWT).
Normal: LVMI $\leq$95. persentil + RWT$\leq$0,42, concentric LV remodelling: LVMI $\leq$95. persentil + RWT $>$0,42; concentric LV hypertrophy: LVMI $>$95. persentil + RWT $>$ 0,42; eccentric LV hypertrophy: LVMI $>$95. persentil+RWT $\leq$0,42

	Left ventricular geometry			
Sports type	Normal (n)(%)	Remodelling (n)(%)	Concentric hypertrophy (n)(%)	Eccentric hypertrophy (n)(%)
basketball	12(48)	7(28)	1(4)	5(20)
football	15(48.4)	5(16.1)	5(16.1)	6(19.4)
swimming	11(24.4)	5(11.1)	13(28.9)	16(35.6)
wrestling	10(43.5)	3(13)	1(4.3)	9(39.1)
total	48(38.7)	20(16.1)	20(16.1)	36(29)

p = 0,009(basketball vs swimming)

Results: Forty eight (38.7%) athletes had normal left ventricular geometry, 20(16.1%) had concentric left ventricular remodelling, 20(16.1%) had concentric hypertrophy, 36(29%) had eccentric hypertrophy (p = 0.001). LVDd did not differ significantly between the sports groups. IVSd and LPWd of both combined and dynamic group were significantly higher than control group. RWT of combined group was higher than static and control group. LVDd index was normal in 108 athletes, more than +2SD in 12 athletes (6 wrestler, 3 football and 3 swimmer) and lower than −2SD in 4 athletes. IVSd and LPWd were highest in swimmers. IVSd, LPWd and RWT were positively correlated with weekly exercise duration.
Conclusions: The majority of athletes had abnormal left ventricular geometry predominantly eccentric hypertrophy. Left ventricular hypertrophy is prominent in combined and dynamic sports.

P-139
Diastolic and systolic right ventricular function in children and young adults with complete heart block and chronic right ventricular pacing: a two-dimensional speckle tracking study
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Introduction: The effect of right ventricular apical (RVA) pacing on systolic and diastolic RV function is not known. The present study assessed RV function in children and young adults with complete heart block and RVA pacing using two-dimensional speckle tracking echocardiography (2D-STE).
Methods: Twenty-eight subjects (14 patients, and 14 controls, mean age 13, 37 ± 5, 32 yrs; 43% male) were prospectively studied. Global LV longitudinal peak systolic strain (LV LS), LV early diastolic strain rate (LV SRe), and RV function was assessed in the apical 4-chamber view using 2D-STE. The weighted average of the RV free wall segments (base, mid, apex) provided the value of global longitudinal RV strain (FW RV) and early diastolic strain rate (RV SRe).
Results: LV deformation parameters were significantly impaired in patients with RVA pacing compared with controls (LV LS: –19.39 ± 2.74% vs –23.37 ± 2.35%, P < 0.001; LV SRe: 1.61 ± 0.39 s-1 vs 2.15 ± 0.39 s-1, P < 0.002). Global systolic and diastolic RV function was preserved in patients (FW RV: − 27.28 ± 5.53% vs –30.32 ± 4.42%, P = 0.151; and RV SRe: 2.27 ± 0.59 s-1 vs 2.32 ± 0.42 s-1 P = 0.822). However, regional apical RV longitudinal peak systolic strain was significantly decreased in patients (RV apical strain: − 24.14 ± 6.09% vs − 29.86 ± 2.95%, P = 0.006).
Conclusions: Although global systolic and diastolic longitudinal RV strain are preserved, regional apical RV function seems impaired in patients with complete heart block and chronic RVA pacing.

P-140
Left ventricular efficiency after ligation of patent ductus arteriosus for premature infants
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Objectives: The purpose of this study was to evaluate the hemodynamic changes in the left ventricular function before and after patent ductus arteriosus ligation in premature infants with regard to the energetic efficiency of left ventricular pumping.
Methods: Thirty-five premature infants who underwent patent ductus arteriosus ligation were enrolled in this study. The left ventricular efficiency was evaluated at four points: within 24 hours before patent ductus arteriosus ligation, within 24 hour after patent ductus arteriosus ligation, between postoperative days 2 and 4, and

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

on postoperative day 7. The indices of contractility (Ees) and afterload (Ea) were approximated on the basis of the systemic blood pressure and systolic or diastolic left ventricular volume. The ratio of stroke work and pressure-volume area, representing the ventricular efficiency, was estimated using the theoretical formula: The ratio of stroke work and pressure-volume area = 1/(1 + 0.5 Ea/Ees).
Results: The left ventricular efficiency was transiently deteriorated within 24 hours after patent ductus arteriosus ligation due to the marked increase of the afterload and the slight increase of contraction, and then recovered to the pre-operation levels by 2–4 days after patent ductus arteriosus ligation.
Conclusions: The analysis of indices representing the afterload, contractility and energetic efficiency of left ventricle could provide practical information for the management of premature infants during the postoperative period after patent ductus arteriosus ligation.

P-141
Detection of right ventricular dysfunction by Pulsed Tissue Doppler Imaging in asymptomatic iron loaded beta thalassemia patients
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Introduction: Iron overload contributes to cardiac dysfunction in patients with beta thalassemia. MRI can detect cardiac iron overload early by parameter termed T2*.
Objective: to clarify the value of Tissue Doppler imaging (TDI) in early detection of global myocardial dysfunction in iron loaded thalassemia patients diagnosed by Cardiac MRI.
Patients and Methods: Two groups were included in the study; group I: 69 asymptomatic thalassemic patients (28 females, 41 males) with a mean age 18.1 ± 7.03 years (range 6 to 39 years). Group II of 41 healthy controls with normal hearts matched for age and sex was selected. Serum Ferritin and cardiac MRI were performed for thalassemic patients to assess the degree of iron overload. Group I was further subdivided into two subgroups; Group Ia (n = 26) T2*value < 20msec and Group Ib(n = 43) T2* > 20msec. Conventional echocardiographic measures of LV dimensions and functions were obtained as well as conventional Doppler measures of the LV and RV, and pulmonary artery pressure.TDI measures included systolic and diastolic myocardial velocities (S', E', A' and E'/A') of the basal segments of septal wall, lateral LV and RV free walls.
Results: Tricuspid annular A' was significantly higher in Group Ia compared to group Ib (16.69 ± 5.16 cm/sec versus 12.07 ± 4.0 cm/sec, P = 0.0001). Tricuspid E'/A' was significantly low in group Ia compared to Ib and group II (1.05 ± 0.42, 1.47 ± 0.43, 1.6 ± 0.33, P = 0.0001). Group Ia (T2* < 20msec) had high serum ferritin level compared to group Ib (T2* > 20msec) (6357 ± 2478 microgram/L versus 2965 ± 2289 microgram/L (P = 0.0001). By univariate and multivariate analysis, Tricuspid E'/A'< 1 was significantly correlating with Ferritin and T2* level.
Conclusion: Right ventricular diastolic dysfunction was evident by Tissue Doppler imaging in cardiac iron loaded asymptomatic β thalassemia patients.

P-142
A critical comparison of 3D echocardiography versus M-mode for the estimation of LV mass
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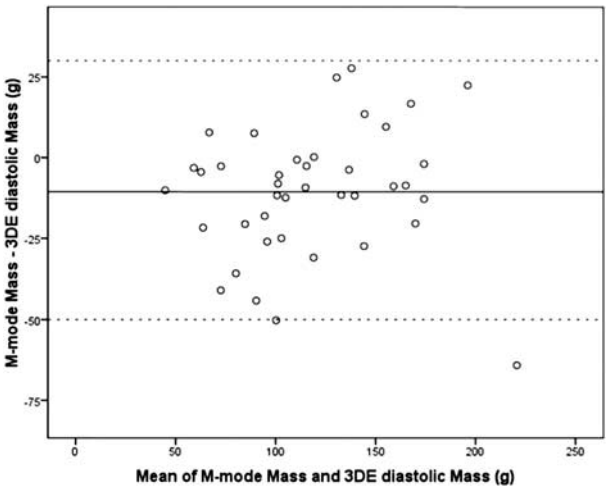
Left ventricular mass (LVM) is an important clinical variable which has prognostic value in many cardiovascular disease states. The most established technique for measurement of LVM is M-mode echocardiography but this is limited by geometrical assumptions of ventricular shape. Three dimensional echocardiography (3DE) has the attraction of semi-automation and avoidance of geometric assumptions. We report the comparison of LV mass measured by 3DE versus M-mode echocardiography.
Methods: 3DE measurements were made by subtraction of endocardial volume from epicardial volume X 1.05. 3DE estimates made at both end-diastole (D) and end-systole (S). M-mode measurements made at end-diastole using Devereux formula. Propsective observational study of 20 patients with disease states (chronic renal disease or neuroblastoma) and 20 healthy patients. Median age 16 years (range 6–29 y), median height 162 cm (range 110–180), median weight 56 kg (21–102), median BSA 1.6 m^2 (range 0.8–2.3).

Comparison of mass	Mean diff (%)	95% CI of mean difference	Limits of agreement
M mode – 3D Diast	−12%	−6 to −18%	−50% to +20%
M mode – 3D Sys	−0.5%	−6 to +6%	−50% to +30%
3D Sys – 3D Diast	−12%	−8 to −14%	+9% to −30%



Results: The median 3DE diastolic LV mass was 117 g (range 50–252 g) and median 3DE systolic LV mass was 105 g (range 40–195 g). 3DE diastolic mass intraclass correlation (ICC) was good for 3DE systolic mass (ICC 0.97) and for M-mode (ICC 0.94). Intra- and interobserver repeatability were good for 3DE diastolic mass (ICC 0.99, 0.93), 3DE systolic mass (0.99, 0.87) and M-mode mass (ICC 0.88, 0.93).
Conclusions: 3DE estimation of LV mass at end-diastole has a bias to produce higher values of LVM than those from M-mode. There is no significant bias for 3DE LVM at end systole versus M-mode. For all comparisons, limits of agreement at relatively wide, thus different techniques cannot be used interchangeably.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

P-143

Assessment of inter-atrial, inter-ventricular and atrio-ventricular interactions in Tetralogy of Fallot patients after surgical correction. Insights from 2-Dimensional Speckle tracking and 3-Dimensional Echocardiography

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Introduction: The atria play a crucial role in patients evolving heart failure. We aimed to assess bi-atrial size and function, presence of inter-actions on atrial and ventricular levels as well as atrio-ventricular coupling, in patients after TOF repair.
Methods: Thirty-four patients with a mean age of 20.9 ± 9 years and 25 healthy controls were included and underwent two-dimensional speckle tracking echocardiography (2D STE) and real time three-dimensional echocardiography (RT3DE) to evaluate ventricular and atrial function. Using 2D STE the mean peak atrial longitudinal strain (PALS) reflecting the passive atrial deformation and the mean peak atrial contraction strain (PACS) representing the active atrial deformation were assessed. Using RT3DE atrial and ventricular end-diastolic (ED), end-systolic (ES) volumes were measured and the ventricular as well as atrial ejection fraction (EF) was calculated. All volumes were normalized to the body surface area (BSA).
Results: When compared to controls TOF patients had significantly reduced right atrial (RA) PALS ($p = 0.0001$), RA PACS ($p = 0.0001$), RA EF $p = 0.0001$), left atrial (LA) PALS ($p = 0.003$), LA PACS ($p = 0.021$), LA EF ($p = 0.005$).
In the TOF group, left ventricular (LV) EF significantly related to the right ventricular (RV) EDV/BSA ($r = -0.66$, $p = 0.0001$), RV EF ($r = 0.63$, $p = 0.0001$). The RA EDV/BSA correlated significantly with LA PALS ($r = 0.66$, $p = 0.0001$) and LA PACS ($r = 0.67$, $p = 0.0001$) among the TOF patients. At the atrio-ventricular level, the RA EF among the TOF patients correlated negatively with RV ESV/BSA ($r = 0.64$, $p = 0.0001$) and positively with RV EF ($r = 0.633$, $p = 0.0001$). Such correlations were not present on the left side.
Conclusions: In TOF patients with moderately enlarged RV and mildly impaired biventricular systolic function bi-atrial dysfunction exists and can be quantified via 2D-speckle tacking as well as RT3DSTE. Robust right atrio-ventricular interaction, altered inter-aterial and inter-ventricular interactions are evident among such cohort.

P-144

Diagnostic problems of cardiac involvement in newborn of diabetic mother

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Cardiac involvement is common in the newborn of diabetic mother and requires early diagnosis and monitoring of evolution.
Objectives: To present the main aspects of the diagnosis of cardiac involvement in newborn of diabetic mothers (IDMs).
Methods: Patients: 89 IDMs newborns investigated in the first week of life and 53 of them re evaluated at 6–12 month, by clinical exam, ECG, cardiothoracic radiography (Rx.CT), Doppler echocardiography (Echo) for congenital and/or acquired cardiac diseases. Fetal echo:in 38 cases after 28 weeks of gestation.
Results: Fetal Echo: cardiomegaly and myocardial hypertrophy of left ventricle (LV)(11 cases), confirmed by postnatal Echo. Clinical exam in newborn: macrosomia (40%), systolic murmur (32), only 3 cases with signs of heart failure, the other being asymptomatic or presenting signs for other pathology than cardiac. ECG: LV hypertrophy (14 cases) and disturbed ventricular repolarization (30 cases). Rx.CT: cardiomegaly (12 cases). Echo: non obstructive hypertrophic cardiomyopathy (HCMP) with asymmetric
IVS hypertrophy (34 cases: 42%), arterial pulmonary hypertension (6 cases), LV diastolic dysfunction with normal systolic function (52% of cases) and congenital cardiac anomalies: PDA (6 cases), VSD (3 cases), coarctation of aorta (1 case), ASD (4 cases). LV myocardial hypertrophy was not significantly correlated with the type of mother's diabetes, before pregnancy or gestational, but rather to an inadequate control of disease. Control performed at 6–12 months (27 cases) revealed a normal morphological cardiac aspect (15 cases) or significant reduction of HCMP (11 cases), all of them showing normal diastolic and systolic LV function.
Conclusions: Newborn of diabetic mother presents a high incidence for cardiac involvement: cardiac congenital malformations (16% of cases) or acquired cardiac pathology: hypertrophic cardiomyopathy (42% of cases) and disturbances of diastolic function of LV (54% of cases), which may be symptomatic or asymptomatic. Fetal echo provides useful data for diabetic pregnant women and should be made mandatory to all these patients. Early cardiologic screening for all of these newborns with or without of cardiac suffering symptoms, especially by Doppler echocardiography that is the most sensitive, noninvasive method for initial diagnostic as well as for follow up.

P-145

The involvement of vasa vasorum in development of vasculitis in the Kawasaki disease animal model

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Introduction: Kawasaki Disease (KD) involves a diffuse and systemic vasculitis of unknown etiology that mainly affects infants and children.
Although a lot of analyses had already been done clinically, histopathologically or molecular-biologically about the mechanism of coronary arterial lesions, it is still not well elucidated.
The objective of this study is to analyse the process of the formation of aneurysms or the change of coronary arteries themselves using animal model.
We investigated the involvement of the invasion from the adventitia in the mechanism of vascular involvement and the development of disease state by scanning electron microscope (SEM), micro CT, and sequential histopathology using murine model of vasculitis induced with Candida albicans water-soluble fraction (CAWS), because KD is associated with a very low mortality recently, and we seldom have a chance to get autopsied heart
Materials And Methods: To prepare the animal model for KD, CAWS was intraperitoneally injected to C57BL/6N mice for 5 days as reported by Ohno et al.
We observed the changes of vasa vasorum at aorta and the orifice of coronary arteries by SEM and micro CT, and also compared neovascularization and distribution at the media and the adventitia quantatively by immunohistochemical analysis.
Results: As previously reported, obvious inflammation were detected 2 weeks after the injection of CAWS, and also

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

subsequent aneurysmal formation, and intimal thickening 3 weeks after it.
In model mice, we found micro vessels verging on the adventitia of Aorta (vasa vasorum), and they increased in model mice. We observed each 1w, 2w, 3w model mice, and found they started increasing 1w after the injection of CAWS, before obvious vasculitis was microscopically detected.
Conclusion: This result indicates that the vasculitis starts by the disorder of vasa vasorum in Kawasaki disease.

P-146

Bicycle Stress echocardiography in children: feasibility, safety and determination of interobserver variability

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Background: Exercise stress echocardiography (ESE) is a well validated technique able to provide a dynamic evaluation of myocardial structure and function in adult population. However its use in children is quite limited, mainly because of the precipitous rapid drop in peak heart rate after exercise. Therefore we aimed to assess the feasibility, the safety and the reproducibility of ESE, using on-line scanning in semi–supine cycloergometer protocol in a large paediatric population.
Methods: Between July 2008 and January 2013, 47 patients (mean age 14 ± 3.1) were evaluated with a bicycle ESE. Two independent observers without knowledge of any patient data interpreted 42 stress studies, grading quality of each acquired image and presence of regional wall motion abnormalities (RWMA).
Results: ESE was successfully performed and well tolerated by all patients. No patient had arrhythmia or complications from stress-induced ischemia. HR was 82 ± 14.3 at rest, and 152.3 ± 19.1 during peak exercise (80.2 ± 15 to 161.4 ± 18 excluding heart transplant patients and patients on beta-blockers). Among 544 views analyzed for grading of image quality, the visualization was optimal in 473 (87%), suboptimal in 39 (7%) and inadequate in 32 (6%). Among 10 patients with hypertrophic cardiomyopathy we were able to assess a significant increase (>25 mmHg) of the left ventricular outflow tract gradient during exercise in 3 patients (33%). ESE was performed in 34 patients with congenital or acquired coronary abnormality (Kawasaki disease, heart transplant recipients, congenital coronary abnormalities, transposition of the great arteries after arterial switch operation). In this group, the RWMA were revealed in 8 patients (24%). The agreement between the two different observers showed a K index of 0.7276 (95% CI = 0.6497 to 0.8055) for the image quality and a K index of 0.5125 (95%CI = 0.4782 to 0.5468) for the RWMA analysis.
Conclusions: Bicycle stress echocardiography performed by on-line scanning during exercise is feasible, safe and reproducible modality in children. Further data to assess its diagnostic accuracy are however needed.

P-147

Myocardial function following repair of anomalous origin of left coronary artery from the pulmonary artery (ALCAPA) in children - a speckle tracking study

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Introduction: We hypothesised that speckle tracking may improve detection of myocardial dysfunction and provide new pathophysiological insights in anomalous coronary artery from the pulmonary artery (ALCAPA).
Method: Echocardiography including speckle tracking was performed in 22 children with ALCAPA (8 male, median age at surgery 0.4 years; IQR: 0.21–1.05) pre and postoperatively and in 22 healthy controls. Measurements included global and segmental longitudinal, radial and circumferential peak deformation (strain) and synchronicity index (SI) defined as agreement of time to peak strain measurements between segments per subject summarised using intraclass correlation coefficient.
Results: Global strains were lower in unoperated patients than controls (longitudinal: −46 vs. −123; $p < 0.001$; circumferential: 48 vs. −118; $p < 0.001$, radial: 110 vs.357; $p < 0.001$) and improved postoperatively (longitudinal:-46pre vs.-82post;$p = 0.002$, circumferential: 48 vs.96; $p = 0.012$, radial: 110 vs.317; $p = 0.001$). Unoperated patients with normal 2D function ($n = 8$) had significantly impaired strain. Global dyssynchrony significantly improved postoperatively (longitudinal SI 0.93 pre vs. 0.94 post, circumferential 0.85 vs. 0.9, radial 0.71 vs. 0.88). Global time to peak shortened (longitudinal 2236 pre vs. 1589 post; $p < 0.001$, circumferential 2037 vs. 1447;$p = 0.005$, radial 2169 vs. 1602; $p = 0.01$, ms). Despite overall global improvement some abnormalities remained. Strain improved in the majority of segments but apical septal and anterolateral segments remained abnormal. Post systolic index improved in some segments but presystolic stretch persisted.
Conclusions: Both global contractility (strain) and global synchrony (coordinated contraction) improved after repair of ALCAPA suggesting recovery of hibernating myocardium. Contractility in some segments supplied by the anomalous left coronary artery failed to improve following ALCAPA repair suggesting a degree of irreversible myocardial damage. 2D speckle tracking identified impairment of function not revealed by standard echocardiography.

P-148

Quantification of right ventricle volume in postoperated Tetralogy of Fallot children. Validation of a new 2D echocardiographic method compared with cardiac magnetic resonance and establishment of cutoff values for decision making

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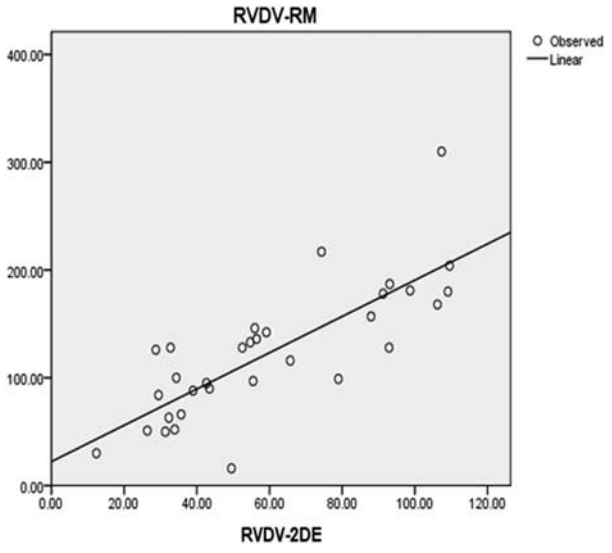
Introduction: Quantification of right ventricular end-diastolic volume (RVEDV) is an essential parameter in the long term follow up of repaired Tetralogy of Fallot (TOF) patients. We have validated a new 2D-echocardiographic method based in RV geometry that reliably correlates with Cardiac Magnetic Resonance (CMR) adding new patients to a previous sample. The aim of this study is a prospective demonstration of the validity of our method and to propose a cutoff value to establish a threshold for pulmonary valve replacement (PVR).
Methods: prospective study of 35 TOF repaired patients. Analysis of the relationship between RVEDV estimated by 2DE (RVEDV-2DE) and later obtained by CMR (RVEDV-CMR). Total RVEDV-2DE was obtained by the sum of two volumes: in apical four chamber view using Simpson formula for an ellipsoid and parasternal short axis view of the right ventricular outflow tract using truncated cone formula. Statistical analysis was

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

performed applying Pearson coefficient method and logistic regression.
Results: 35 new repaired TOF patients were included, mean age 10.6 years (ranging from 1 to 21). Mean RVEDV-2DE versus RVEDV-CMR were 58.36 versus 118.7 ml (ranging from 12.35 versus 16 ml and 109.55 versus 204 ml). RVEDV-2DE had significant correlation ($p < 0.0001$) with the RVEDV-CMR, with Pearson Correlation Coefficient of 0.794. Linear regression model resulting in the following: RVDV-CMR = 22.26 + (1.68 × RVDV-2DE). From the lineal model formula we can establish the following cutoff values for RVDV-2DE measurements to define a large RV and establish indication of PVR: 76 ml/m^2, equivalent value to cutoff established by Tal Geva1 of 150 ml/m^2 by CMR.
Conclusions: we confirm that quantitative assessment of RVEDV-2DE using our method is feasible and correlates with RVEDV-CMR. We establish cutoff values of remarkable importance for decision making in this group of patients using easy, helpful and available 2D echocardiography. This method could also be useful for patients with contraindications for CMR performance.



P-149
Echocardiographic right ventricular function variables: external validity study in a normal pediatric population
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Introduction: Regular echocardiographic assessment of right ventricular (RV) function is required in patients with congenital heart disease (CHD). Although normal values are validated in the adult population, less RV studies have been published in normal pediatric population. The aim of this study is to define in real clinical situation normal values for RV variables in children and to compare the results to published studies.
Methods: We conducted a prospective multicenter study from 2010 to 2012 in our pediatric cardiology tertiary care center. 314 normal children (normal physical examination, electrocardiogram and echocardiography and no CHD, chronic disease or treatment) aged 2 days to 18 years were included (46% female, 88 infants under 1 year old, 26 neonates, 226 children). Following RV variables were collected by 4 pediatric cardiologists using 2 ultrasound systems (Philips IE33, Aloka Alpha 10): tissue Doppler imaging (TDI) E', A' and S waves at the tricuspid valve, TDI Tei Index and tricuspid annular plane systolic excursion (TAPSE). The impossibility to measure a variable was informed. A univariate polynomial linear regression was used to assess the relationship between these indexes and the anthropometric factors (age, weight, height, BSA and gender) via the coefficient of determination (R^2).
Results: More than 90% variables could be easily collected. Mean values for E', A' and S waves were: 13, 7 ± 3, 8 cm/s, 10, 1 ± 3, 7 cm/s, and 12 ± 2, 2 cm/s; mean values for TAPSE were 18, 7 ± 4, 9 mm and 0, 41 ± 0, 11 for DTI Tei index. In infant, E', A' and S waves were best correlated with BSA, and TAPSE with height. In children S wave and TAPSE were best correlated with weight, and A' with age. No significant correlation was found for E' wave in children, and for DTI Tei index. No significant differences were found between genders, ultrasound systems. Values were mostly well correlated to published studies. The experience of the pediatric cardiologist had an impact on the results.
Conclusion: We established echocardiographic RV function reference values in children with good feasibility and correlation to published reference studies. Routine application to children with CHD must be defined.

P-150
Early cardiovascular prevention: Does physical fitness and activity affect arterial structure and function in children?
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Objectives: The main objectives of this study were to investigate, if physical fitness and physical activity are associated to arterial structure and function in children.
Methods: 119 children (53 girls) aged in the median 12.3 years, IQR 11.9–12.9 years and median body mass index standard deviation score (BMI-SDS) of 0.06 (IQR -0.93–1.18). All children had ultrasound sonography (ProSound alpha7, Hitachi/Aloka) on carotid intima-media thickness (cIMT), arterial compliance and stiffness. Brachial flow-mediated dilatation (FMD) was measured using a continuous eTRACKING mode, analyzing changes in blood flow velocity and arterial diameter at baseline, ischemia and vasodilatation (Prosound alpha 6, Hitachi/Aloka). Physical fitness was tested via a symptom limited pulmonary exercise test on a bicycle ergometer (Geratherm Respiratory, Ganshorn Medical, Germany). Physical activity was assessed using GT3x accelerometers (Actigraph, USA).
Results: No significant relations between cIMT and physical fitness as well as physical activity revealed. However, physical fitness was inversely correlated to carotid ($r = -0.246$; $p = 0.012$) and brachial ($r = -0.208$; $p = 0.036$) stiffness indices. Time in sedentary lifestyle was inversely correlated to carotid arterial compliance ($r = -0.210$; $p = 0.034$), whereas time in high intensive activity level was negatively correlated with the brachialis stiffness ($r = -0.286$; $p = 0.004$).
Furthermore sex differences revealed. Girls demonstrated higher arterial stiffness and lower arterial compliance than boys.
Additionally, girls spent less time ($p = 0.001$) in moderate activity than boys. No sex-differences in systolic FMD (%) and diastolic FMD (%), anthropometric data were assessed.
Conclusions: This research is suggestive of the fact that physical fitness as well as physical activity have an affect on arterial

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

function but not on arterial structure. Girls seem to be at higher atherosclerotic risk than boys due to less physical activity, demonstrating higher arterial stiffness and lower compliance. Further research need to clarify the mechanisms of early endothelial (dys)-function and the impact of physical fitness on them.

P-151

Unusual presentation of atrioventricular (AV) univentricular connection: Absent left AV connection with a dominant ventricle of left morphology. Anatomical and echocardiographic findings in four cases

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Introduction: Despite of existing a systematized nomenclature for all types of univentricular AV connection, there are still some cases challenging pediatric cardiologists/echocardiographers. Usually, absence of one AV connection is associated with the presence of a rudimentary ventricular chamber at the same side, as the absent connection. Very rarely, the hypoplastic chamber is located at the contralateral side of the absent connection, with the dominant ventricle of unexpected morphology for the side of absent connection. We sought to clarify this unusual feature by reviewing a series of four cases, including one fetal diagnosis, with this morphological arrangement.
Methods: From the hospital database/char, data were obtained. Echocardiogram studies were reviewed and correlation with anatomical features from pathological study was done.
Four cases were studied with absence of left AV connection with a dominant left ventricle and rudimentary right ventricular chamber located at the right side. Three specimens were examined from patients who died. The remaining patient has been followed after a palliative surgery.
Results: There was agreement between the initial echocardiogram report and pathological features in two cases. In the two remaining, the initial diagnosis was "Tricuspid atresia", including one fetal diagnosis.

Patient	ASD	VSD	Ventriculo-arterial connections	Location of rudimentary chamber	RVOTO	TAPVD
1	PFO	Muscular restrictive	Concordant	Right	Yes	No
2	Restrictive PFO	Muscular restrictive	Discordant	Right	No	No
3	Secundun ASD	Non restrictive muscular	Concordant	Right	No	No
4	PFO	Non restrictive muscular	Concordant	Right	Yes	Yes

In all cases there was an early clinical presentation with hypoxia and low cardiac output. Right outflow tract obstruction was detected in two patients. Two patients required atrial septostomy for a restrictive PFO. In the remaining patient there was a small PFO, but there was total anomalous pulmonary venous return to the coronary sinus. (table 1)
Conclusions: Echocardiographers must be aware about unusual forms of AV connection. The correct interpretation of absent AV connection by standard views needs a dynamic evaluation and the combination of different views to establish the correct spatial orientation of cardiac chambers and determination of ventricular morphology. Fetal diagnosis requires more views and careful interpretation as well. The sequential segmental analysis is fundamental to achieve the correct diagnosis, both by echocardiography and other morphological methods.

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Echocardiography by Neonatologists: "Reliable or risk"?

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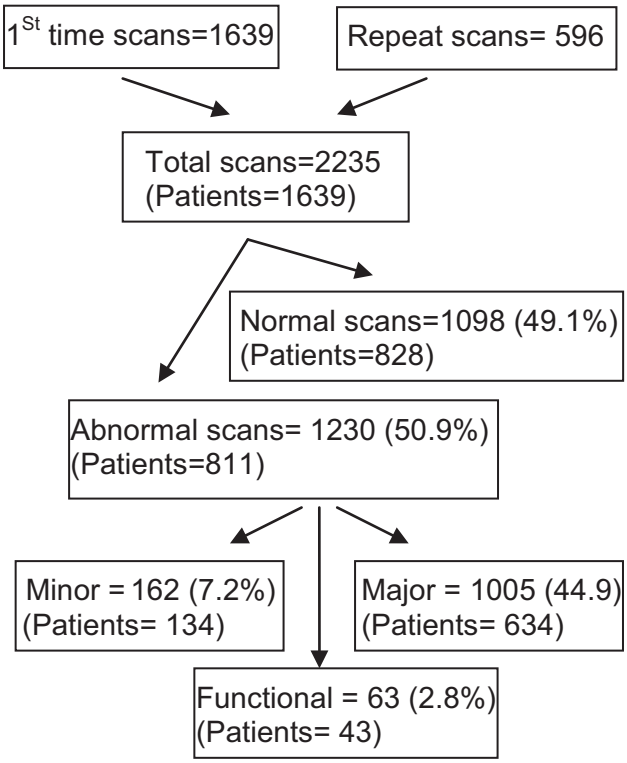
Background: Echocardiography is an important investigation for the management of sick neonate and is also essential for diagnosis of congenital heart disease. Not all Neonatal Intensive Care Units have access to Paediatric Cardiology services. Many units have neonatologists doing echocardiograms. This study aims to assess the validity of echocardiograms performed by the Neonatologists in a level 3 Neonatal Intensive Care Unit.

Chart 1: Flow diagram of Echocardiograms



Methods: This is a retrospective study over a $7\frac{1}{2}$ year period. Echocardiograms were performed either on NICU or outpatient murmur clinics. Selected abnormal examinations were referred to regional Cardiology services. Database of all the scans were maintained electronically. This neonatal database was compared with regional Paediatric Cardiology database called Cardiobase® to assess concordance and discordance of the findings.

Table 1. Concordance Rates

Cardiobase® Data (Total)	454 scans	100%
Concordance	417 scans	91.8%
Partial concordance	20 scans	4.4%
Discordance	17 scans	3.7%

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Table 2. Sensitivity and specificity rates

Sensitivity	98.4%
Specificity	98.9%
Positive predictive value	97.2%
Negative predictive value	99.4%

Results: Total of 2322 scans were performed on 1639 neonates from 1st September 2003 till 31st December 2011. Cardiobase® data was available for 454 scans. Complete concordance was found in 91.8% and partial concordance was found in 4.4% of scans. This was statistically significant with p value of 0.0001 on comparing the proportions of concordant and discordant scans. 31 infants (1.6% of total infants scanned) had critical structural abnormalities, all of which were identified correctly. 3.7% of the scans had discordant findings, most of these were false positives referred to exclude critical events. Sensitivity and specificity was 98%.
Conclusion: Neonatal echocardiography by neonatologists has high concordance rates and has a high sensitivity and specificity in detecting congenital heart disease. With appropriate Paediatric Cardiology support Neonatal Echocardiography by neonatologists can be a safe and reliable tool.

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Non-invasive measurements of hemodynamic transition directly after birth

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Objective: To assess the hemodynamic changes in newborns delivered by caesarian section in the first 10 minutes after birth using non-invasive techniques.
Study design: In 24 term newborns, heart rate (HR) and preductal blood pressure (BP) was measured non-invasively at 2, 5 and 10 minutes after birth. An echocardiographic examination was performed at the same intervals to monitor left ventricular dimensions and function.
Results: Mean (SD) HR and mean BP did not change in time (HR: 157 (21) bpm at 2 minutes, 154 (17) bpm at 5 minutes and 155 (14) bpm at 10 minutes; mean BP: 51.2 (15.4) mmHg at 2 minutes, 50.5 (11.7) mmHg at 5 minutes and 49.6 (9.5) mmHg at 10 minutes; ns). Left ventricular end diastolic diameter (LVedd) increased from 2 to 5 minutes (14.3 (1.3) mm vs. 16.3 (1.7) mm) ($p < 0.001$) and remained stable with 16.7 (1.4) mm at 10 minutes. Left ventricular output (LVO) increased between 2- and 5 minutes (151 (47) vs. 203 (55) mL/kg/min ($p < 0.001$)) and remained stable at 201 (45) mL/kg/min at 10 minutes. The increase in LVO was significantly correlated with left ventricular stroke volume (LVSV; $r = 0.94$, $p = 0.01$), but not with HR ($r = 0.37$, ns).
Conclusion: The most significant hemodynamic change occurs within the first 5 minutes after birth with an increase in LVedd and LVSV causing a significant increase in LVO, whereas blood pressure is stable during the 10 minutes after birth.

P-154

A four year follow up of the parameters of deformation by means of the strain 2D speckle tracking method in patients with systemic right ventricle

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Introduction/Objectives: The purpose of this study has been to assess the evolution in time of the deformation parameters in the systemic right ventricle in adult patients with D-transposition of the great arteries (TGA) who have been treated with the Senning technique.
This study has been carried out using the 2D speckle tracking strain quantitative technique independent of ventricular geometry.
Material and Methods: Ten patients with D-TGA who had been treated with the Senning technique were studied. These patients (except for one that was lost during the study) were followed for four years.The average age was 25.7 ± 2.7. All patients have emained in functional class I and no complications occurred. Ten other patients with D-TGA corrected by means of the Jatene technique were included, average age 9.9 ± 5.4 and also a normal control group average age 21.8 ± 7.7. For this study we used a GE VIVID 9 and some equipment for off-line analysis (software 2D EchoPac strain).
Results and Conclusions: 1.No significant changes were observed in the parameters of systemic right ventricle strain after four years from the initial study in these patients. 2. No changes were obtained in the parameters of left ventricular deformation in such patients. 3.The patients who underwent the Jatene technique have shown no difference from the normal group 4. The parameters of the systemic right ventricular strain obtained by 2D strain made it possible to provide information on the prognosis of such patients.

	Strain Global RV	Strain Rate RV	Strain Global LV
TGA2007	$-14,4 \pm 3$	$-1,3 \pm 0,3$	$-17,85 \pm 2$
TGA2011	$-16,68 \pm 3,3$	$-1,35 \pm 0,4$	$-18,64 \pm 2$
NORMAL	$-22,97 \pm 3$	$-1,98 \pm 0,7$	$-17,1 \pm 4$
JATENE	$-20,67 \pm 2,8$	$-1,95 \pm 0,6$	$-17,46 \pm 2$

P-155

The role of cardiac magnetic resonance to address the treatment of choice for pulmonary valve replacement late after repair of Tetralogy of Fallot

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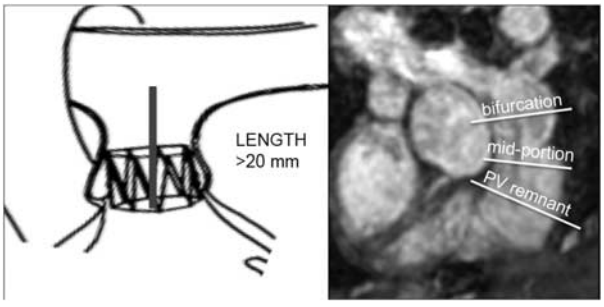
Introduction: Severe pulmonary regurgitation (PR) with progressive right ventricular (RV) dilatation and dysfunction occurs late after repair of Tetralogy of Fallot (TOF) and may be strongly associated with ventricular tachycardia and sudden death. Cardiac magnetic resonance (CMR) is the gold standard to evaluate the pathophysiology in repaired TOF and the main tool to support decision for pulmonary valve replacement (PVR) in asymptomatic patients. Given the various options available for PVR, we sought to evaluate the usefulness of CRM to address patients towards either a surgical or interventional procedure.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Methods: Between 2008 and 2012, 69 patients with repaired TOF underwent CMR study to address PVR. Indications for PVR were RV end-diastolic volume >150 ml/m^2, RV end-systolic volume index >80 ml/m^2 and a RV ejection fraction < 47%. Pulmonary trunk (PT) morphology, length, and dimensions measured at three levels (PV remnant, mid-portion, bifurcation) and coronary anatomy were evaluated, using a 3D SSFP navigator sequence. PT expansion during the cardiac cycle was assessed by cine sequences in three planes.
Results: Right ventricular outflow tract (RVOT) and coronary examination was possible in all patients. Suitability for percutaneous treatment included maximum PT diameter between 19 and 27 mm and PT length greater than 20 mm, whereas RVOT aneurisms, shape irregularity and/or significant systolic PT expansion were exclusion criteria. Associated pulmonary branch stenosis and/or hypoplasia were indication for homograft RVOT replacement. Seven patients (10%) were addressed to transcatheter PV, which was successful in all cases. All other patients (90%) underwent surgery by means of bioprostethic (37), homograft (17), perventricular injectable valve (8). There were no deaths or major complications.
Conclusions: CMR is a fundamental tool after TOF repair, not only in assessing RV volumes and function, but also in evaluating RV outflow tract measures and morphology during the cardiac cycle in order to address the best choice for PVR.



P-156
Aortic root dilatation in adult patients with repaired tetralogy of Fallot
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Introduction: Aortic root dilatation is commonly observed among patients with repaired tetralogy of Fallot. The aortic root is mostly involved, at all levels. Nevertheless, the prevalence of aortic root dilation and its rate of growth remain to be defined.
Methods: We conducted a retrospective study, reviewing aortic MRI measurements at different levels (sinuses of Vasalva, sinotubular junction, ascending aorta, horizontal aorta, isthmus, and descending aorta) from 51 consecutive adult patients with repaired tetralogy of Fallot. Matched controls for age and sex were recruited in a healthy population of patients undergoing a cardiac MRI study for other reason. Moreover, the annual rate of aortic growth was determined by MRI for 28 patients with repaired tetralogy of Fallot.
Results: 59% of patients with repaired tetralogy of Fallot suffered from an aortic dilation located at the level of the sinuses of Vasalva, versus 6% in the control group, according to the Roman criteria ($p < 0.001$). Compared to the control population, aortic segments are significantly larger at all ascending levels, including the horizontal segment in patients with repaired tetralogy of Fallot: at the sinuses of Valsalva, mean aortic diameter is 20,4 mm in the tetralogy of Fallot group, versus 15,6 mm in the control group ($p < 0.001$). There is no difference between the two groups at the descending level of the aorta (9.9 mm in patients with repaired tetralogy of Fallot, versus 9.8 mm in control patients, $p = 0.267$). Among patients with repaired tetralogy of Fallot, the rate of aortic growth is 0.697 ± 1.6 mm/year at the sinuses of Vasalva and 0, 236 ± 1, 29 mm/year in the ascending aorta.
Conclusions: Aortic root dilatation is frequent among patients with repaired tetralogy of Fallot and mostly concerns the aortic root, compared to a control group of healthy patients. Horizontal and descending aortas do not seem to be involved in the dilation. Aortic root dilation needs to be carefully and regularly controlled, as it appears to be a dynamic and progressive phenomenon, although rather slow.

P-157
Echo and psycho status of TGA postsurgery patients from Sarajevo
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Introduction: Postsurgical follow up of TGA pts in relation to type of surgical correction and type of TGA, demands a multidisciplinary approach with the aim of morphohaemodinamic evaluation as well as the child's adjustment, baring in mind a degree of hypoxia in these pts.
Methods and pts: During 1996–2013 at Paediatric clinic of CCU Sarajevo 56 pts were diagnosed as TGA by clinical examination, ECG, X-ray and confirmed echocardiographically. This study is hospital based and includes 17 pts age 6–18 years with the aim of echo and physcho status evaluation compared with control Group of healthy children in relation to BW. Psycho tests were done using REWISC, Bine-Simon Scale, Beck Youth Inventory (Self-Concept, Anxiety, Depression, Anger and Disruptive Behavior) and Achenbach's Child Behaviour tests.
Results: Study formed 2 Groups of pts: I Group 13 pts with D TGA (9 boys) and II Group 4 pts (4 boys) with complex TGA. Mean age at the time of diagnosis in I Group was 18.3 days/1–43 days/, in II Group 88.5 days/24–180 days/. Oxygen saturation in I Group at admission 78.7%/65%–87%/, and in II 75.2%/67%–85%/. Surgery was performed in I Group mean age 67.2 days of life, in II Group mean age 105 days of life. Post surgical follow up period in I Group was 12.2 years/4–18years/, II Group 14.5 years/12–16years/. Pts echo parameters post surgery from Group I and II, in relation to control group of healthy children had statistically significant changes in: LVEDD ($p = 0.025$), LVESD ($p = 0.016$), IVS ($p = 0.039$), with no statistically significant changes in coronary arteries, FS LV, MV E/A ratio, TV, RVOTO, LVOTO, presence of thrombus or vegetation. Intellectual abillities in a normal range had 75% pts, below-average intelligence was found 12.5%, 46.2% had above average self-concept, 25.0% had a mild anxiety, mild depression in 18.8% and 12.5% pts had above average high frequency of anger.
Conclusion: Postsurgical echo results in TGA pts had satisfactory morphohaemodinamical outcome except LV dimensions. The most of the children in our study had intellectual abilities in a normal range, and normal psychological functioning. There is no significant differences between two groupes in tested psycho variables

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

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Echocardiographic Nomograms for Ventricular, Valvular and Arterial Dimensions in Neonates and Infants

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Background: A quantitative assessment is essential during the performance of a paediatric echocardiography. Despite this actual nomograms are limited and heterogeneous, particularly for the neonatal age. Aim of the present study is to establish reliable echocardiographic nomograms in a population of healthy neonates, infants and children.
Methods: Two-dimensional and M-mode measurements were made of 22 individual cardiac structures in 368 healthy children (mean age 4,99 Sd 8,47 months; 186 F, 182 M) with a body surface area (BSA) $<0{,}6\,m^2$. Models using linear ($y = a + b^{\star}x$), logarithmic ($y = a + b^{\star}\ln[x]$; $\ln[y] = a + b^{\star}x$), exponential ($\ln[y] = a + b^{\star}\ln[x]$), and square root ($y = a + b^{\star}x^{1/2}$; $y^{1/2} = a + b^{\star}x$) relationships were tested. The presence or absence of heteroscedasticity was tested by White test and Breusch-Pagan test and observing the graphical analysis of standardized residuals. Age, weight, height and BSA (calculated using the formulas by Du Bois and by Haycock) were used as the independent variables in four different linear regression analyses to predict the mean values of each echocardiographic measurement. Between the models that satisfy the assumption of homoscedasticity, the R^2 score were used to determine which model best fitted the data. We computed structures Z score by dividing the residual value by the modeled standard error of the residual value.
Results: The use of BSA calculated by Du Bois formula provided the best results, and these are presented in Table 1. According to these data, different equations should be employed for various parameters.
Intra and inter-observer variability was within the 95% tolerance limit.
Conclusions: We present echocardiographic nomograms calculated on the widest population of healthy neonates and infants reported so far. Our data will allow for a more reliable calculation of cardiac z scores for some essential cardiac and vessels dimensions in neonates and infants. Further studies however are required to reinforce these data, as well as to evaluate other parameters.

	Linear $Y = a + bx$		Logarithmic $Y = a + b^{*}\ln[x]$		Exponential $\ln[y] = a + b^{*}\ln[x]$	
Measurement	BP	W	BP	W	BP	W
IVC	0.000	0.000	0.000	0.000	0.501	0.630
LVEC M-mode	0.051	0.127	0.006	0.024	0.013	0.046
LVES M-mode	0.029	0.030	0.0007	0.0006	0.364	0.467
IVSd M-mode	0.014	0.140	0.014	0.137	0.068	0.113
LVPWd M-mode	0.000	0.002	0.000	0.003	0.051	0.347
Tricuspid annulus	0.000	0.0006	0.000	0.0001	0.482	0.489
Mitral annulus	0.0003	0.0001	0.001	0.005	0.029	0.031
Mitral long axis	0.003	0.015	0.001	0.009	0.124	0.306
Aortic annulus	0.000	0.000	0.000	0.000	0.022	0.035
Sinuses of Valsalva	0.000	0.000	0.000	0.000	0.049	0.144
Junction	0.000	0.000	0.000	0.000	0.747	0.132
Asc AO	0.000	0.0003	0.000	0.0001	0.230	0.298
Arch lA LCA	0.000	0.000	0.000	0.000	0.642	0.273
Arch LCA-LSA	0.000	0.000	0.000	0.000	0.240	0.049
Arch After. LSA	0.000	0.000	0.000	0.000	0.774	0.007
lsthmjs	0.000	0.000	0.000	0.000	0.764	0.004
Thorac AO	0.000	0.000	0.000	0.000	0.913	0.211
Abdominal Ao	0.995	0.999	0.434	0.785	0.002	0.019
Pulomary annulus	0.000	0.000	0.000	0.000	0.894	0.954
MPA	0.0001	0.0005	0.000	0.0001	0.220	0.358
LPA	0.0001	0.007	0.001	0.030	0.003	0.010
RPA	0.000	0.000	0.000	0.001	0.218	0.518
	Square root $Y = a + bx^{1/2}$		Logarithmic 1 $Ln[y] = a + bx$		Square root 1 $Y^{1/2} = a + bx$	
	BP	W	BP	W	BP	W
IVC	0.000	0.000	C.773	0.528	0.0001	0.0002
LVEC M-mode	0.029	0.093	C.024	0.004	0.719	0.339
LVES M-mode	0.006	0.007	C 073	0.159	0.958	0.723
IVSd M-mode	0.018	0.159	C 146	0.020	0.643	0.481
LVPWd M-mode	0.000	0.003	0.071	0.374	0.000	0.015
Tricuspid annulus	0.000	0.0004	C 826	0.345	0.022	0.091
Mitral annulus	0.0007	0.002	C 100	0.000	0.311	0.0007
Mitral long axis	0.002	0.023	C.229	0.071	0.466	0.346
Aortic annulus	0.000	0.000	0.0003	0.001	0.000	0.000
Sinuses of Valsalva	0.000	0.000	C.010	0.006	0.000	0.000
Junction	0.000	0.000	C 804	0.028	0.002	0.001
Asc AO	0.000	0.0002	C.274	0.345	0.0001	0.010
Arch lA LCA	0.000	0.000	0.171	0.030	0.0002	0.001
Arch LCA-LSA	0.000	0.000	C.009	0.002	0.000	0.000
Arch After. LSA	0.000	0.000	C 104	0.002	0.000	0.000
lsthmjs	0.000	0.000	C.0G2	0.3002	0.000	0.000
Thorac AO	0.000	0.000	C 739	0.007	0.003	0.005
Abdominal Ao	0.790	0.925	C.001	0.010	0.058	0.289
Pulomary annulus	0.000	0.000	C 761	0.587	0.043	0.107
MPA	0.000	0.0002	C.204	0.002	0.226	0.058
LPA	0.0003	0.022	C 069	0.005	0.323	0.370
RPA	0.000	0.0001	C 910	0.020	0.006	0.019

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Exercise Stress Echocardiography in Children with Hypertrophic Cardiomyopathy

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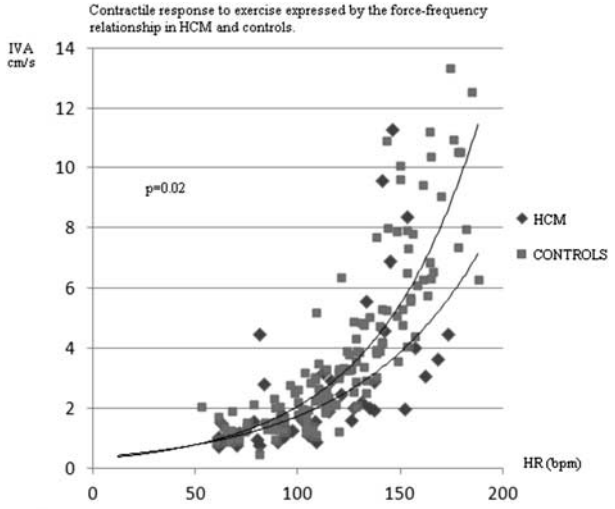
Background: Hypertrophic Cardiomyopathy (HCM) is the most common genetic cardiomyopathy and the leading cause of sudden death in young athletes.The disease commonly causes left ventricular (LV) diastolic dysfunction, and late LV systolic dysfunction. Diastolic dysfunction has been described limiting exercise capacity but less information is available on systolic contractile reserve in pediatric patients. The aim of the current study was to evaluate myocardial response to exercise in children with HCM using semi-supine cycle ergometry stress echocardiography (SSCE).
Materials and Methods: Seventeen children with HCM and 24 controls were included. A stepwise SSCE stress echocardiography protocol was used. Tissue Doppler Imaging (TDI) peak systolic and early diastolic velocities were measured in the LV lateral wall and basal septum during exercise and the changes in E' and S' values from baseline to peak were compared between HCM and controls. LV myocardial acceleration during isovolumic contraction was measured in all the subjects at incremental HR to evaluate the force-frequency relationship (FFR).
Results: The change in E' and S' from baseline to peak exercise was lower in HCM patients than in controls in LV later wall (ΔE'5.7 ± 1.7 vs. 7.5 ± 2.9 cm/s, $p = 0.03$; ΔS' 3.4 ± 4 vs.7.4 ± 3.4 cm/s, $p = 0.001$) and basal septum (ΔE'5.1 ± 2.9 vs. 6.9 ± 1.8 cm/s, $p = 0.02$; ΔS' 3.7 ± 2.3 vs. 5.3 ± 1.6 cm/s, $p = 0.01$). The contractile response as studied by the FFR, was significantly blunted in HCM compared with controls, $p = 0.02$ (see figure).
Conclusions: Our data suggest a significantly blunted myocardial response exercise in patients with HCM. Apart from a previously described reduced diastolic response suggested by the blunted

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

increase in E'-velocities, we also demonstrate a reduced systolic response as suggested by the reduced S' response and particularly by the blunted force-frequency response.



P-160

Can Echocardiography Be Used To Delineate Changes in Pulmonary Hemodynamics in First two Days of Life in Human Neonates?

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Can Echocardiography Be Used To Delineate Changes in Pulmonary Hemodynamics in First two Days of Life in Human Neonates?

Echocardiography (echo) is used to diagnose acute pulmonary hypertension (PH) in neonates; but, echo- derived indices (EDIs) of PH have not been well described during early postnatal period.
Objective: To test the reliability of EDIs to evaluate pulmonary hemodynamics in newborns and determine changes occurring between day 1 and 2 of life, when PVR normally falls.
Design/Methods: 50 healthy term neonates had serial echo evaluations at 15 ± 2 & 35 ± 2 hours of life. EDIs relating to pulmonary vascular resistance [pulmonary artery acceleration time (PAAT), right ventricular ejection time (RVET), pulmonary vascular resistance index (PVRI = RVET/PAAT)] and pulmonary blood flow [right ventricular stroke volume (RSV) and output (RVO)] were measured. Inter-rater agreement was analysed using Bland-Altman analysis and interclass correlation coefficient (ICC) was calculated.
Results: Pre-defined EDIs were successfully measured in 98 out of 100 scans. Changes from D1 to D2 included [mean ± SD; *p*) decreased PVRI [4.8 ± 1.3 vs. 3.8 ± 0.8; p < *.001*) and increase in PAAT [49 ± 17 vs. 59 ± 15 msec; p < *.001)*, RSV [2.1 ± 0.6 vs. 2.3 ± 0.7 mls; *p = 0.009*; and RVO (246 ± 66 vs. 294 ± 108 mls/min/kg; *.p = .002*.
Conclusions: EDIs to evaluate pulmonary circulation can be reliably obtained in newborns. Serial measurements may provide a semi-quantitative method to study changes in PVR over time.

Table. Sequential measurements of echocardiography-derived indices during the first 24 hours

Age (hrs)	<0.5	2.5	8	22	*p*
PAAT (msec)	33 ± 1	37 ± 1	43 ± 1*	55 ± 2*	<.001
RVET (msec)	166 ± 9	186 ± 10	208 ± 6*	216 ± 6*	<.001
PVRI (RVET/PAAT)	5.1 ± 0.3	5.0 ± 0.2	4.9 ± 0.2	4.0 ± 0.2*	<.001
PA VTI (cms)	8.2 ± 0.5	9.9 ± 0.9	10.3 ± 0.4*	12.2 ± 0.5*	<.001
RVSV (mls)	5.0 ± 0.6	6.0 ± 0.7*	6.3 ± 0.4*	7.4 ± 0.5*	<.001
RVSV/kg	1.4 ± 0.2	1.7 ± 0.2	1.8 ± 0.2*	2.1 ± 0.2*	<.001
RVO (mls/kg/min)	210 ± 20	206 ± 28	217 ± 28	248 ± 27	.059
Ao VTI (cms)	7.5 ± 0.5	9.5 ± 0.6*	10.3 ± 0.7*	10.9 ± 0.5*	<.001
HR (beats/min)	144 (135–173)	124 (110–133)	122 (104–130)	113 (107–140)	.09

P-161

Prospective Study to Establish Quantitative Indices of Normal Right Ventricular Function in Newborn Infants Using Echocardiography

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Background: Systematically established normal reference ranges may help to investigate right ventricular (RV) dysfunction in neonates.
Objectives: To test the feasibility of echocardiography to quantify right ventricular function in neonates, establish normal reference ranges and describe changes during first two days of postnatal transition.
Methods: In a prospective observational study 50 well neonates (17 males) with a mean (±SD) gestational age of 39.7 (±1) weeks and birth weight of 3.5 (±0.4) Kg underwent 2 serial echocardiograms (mean age 15.1 (±2) & 34.9 (±2) hours at 1st and 2nd scan respectively). Images were acquired for blinded offline calculation of RV functional parameters as per the American Society of Echocardiography guidelines. In addition a novel image of RV-focused apical short axis (RV 3 chamber view) was taken to visualize inflow & outflow tract simultaneously. Following RV functional indices were calculated – Fractional area change in 4 chamber (FAC-4C), 3 chamber (FAC-3C) and global FAC (average of 4C & 3C); TAPSE; MPI; tissue Doppler

Variable	Day 1	Day 2	*p* value	ICC—intra	ICC—inter
FAC – 4C (%)	26.4 ± 7	27.1 ± 8	0.68	.82	.51
FAC – 3C (%)	38.7 ± 7	38.4 ± 6	0.74	.67	.65
Global FAC (%)	32.6 ± 5	32.8 ± 4	0.89	.72	.76
TAPSE (cms)	0.92 ± 0.14	0.91 ± 0.14	0.69	.94	.77
TVI (peak systolic)	6.5 ± 1.2	6.5 ± 1.1	0.95	.96	.94
2D Strain (lateral wall) %	21.2 ± 5.2	21.3 ± 5.3	0.79	.93	.96
2D Strain (inferior wall) %	21.4 ± 4.3	20.6 ± 4.1	0.40	.77	.84
Global strain %	21.2 ± 3.9	21.2 ± 4.2	0.72	.89	.87
MPI	.37(.31—.49)	.41 (.37—.50)	0.66	.97	.80

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

velocities; peak longitudinal strain in 4C (RV lateral wall), 3C (RV inferior wall) and global (average of 4C & 3C). Two scans were compared using paired t-test & Interclass correlation coefficient was used to test inter-rater reliability.
Results: All pre-defined RV functional indices were successfully measured demonstrating a high inter-rater reliability. Longitudinal strain could be successfully measured in 94% for both lateral & inferior walls. No differences were observed between days 1 vs. 2.
Conclusion: The normative data established in this study can pave the way for further research to investigate the incidence & relevance of RV dysfunction in neonatal diseased states.

P-162
Transesophageal echocardiography with Blood Flow Imaging during Atrial Septal Defect closure: A comparison with the current references

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Introduction: Flow visualisation before transcatheter atrial septal defect (ASD) closure is essential to identify the number and size of ASDs and to map the pulmonary veins. Previous reports have shown improved transthoracic visualisation of ASD and pulmonary veins using Blood Flow Imaging (BFI), which supplements Colour Doppler Imaging (CDI) with angle-independent information of flow direction. This study aimed to compare transesophageal BFI with the current references in ASD sizing (Balloon stretched diameter, BSD) and pulmonary vein imaging (pulmonary angiography).
Methods: 28 children referred to interventional ASD-closure were investigated with transoesophageal echocardiography including BFI scanning of the ASD and the pulmonary veins before the closing procedure. BFI and CDI cineloops were prepared offline and presented to four observers (senior cardiologists) who were blinded to the patient data. They measured the ASD in two planes twice in each patient with BFI and CDI. The maximum ASD-diameter measured with BFI by each observer was compared to the corresponding BSD- and CDI-measurements. Repeatability of the BFI-measurements was calculated as the residual coefficient of variation when accounting for patient and interobserver variance.
The pulmonary veins were evaluated during the procedure. BFI investigations were completed and documented before the pulmonary veins were examined by routine angiography.
Results: The mean maximum diameter measured by BFI was 12.1 mm (SD 2.4 mm). The corresponding BSD- and CDI-measurements were 15.9 mm (SD 3.0 mm) and 11.8 mm (SD 2.5 mm) respectively. The residual standard deviation was 1.2 mm and the corresponding residual coefficient of variance was 9.6%. Compared to pulmonary vein angiography the sensitivity of BFI in detecting the entry of the pulmonary veins was 0.96 (95% CI: 0.82–1.0).
Conclusions: Transesophageal echocardiography with Blood Flow Imaging of the pulmonary veins and BFI-ASD sizing agreed well with the references pulmonary vein angiography and balloon stretched diameter, respectively. The repeatability of the BFI-measurements was close to the inherent measurement error of the ultrasound measurement itself.
Improved echocardiographic imaging in connection with transcatheter ASD-closure may reduce radiation exposure and possible also the need for balloon sizing. BFI may be a suitable supplement in this complete echocardiographic ASD evaluation.

P-163
Percutaneous Pulmonary valvuloplasty in patients with severe right ventricular dysfunction

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Introduction: Percutaneous pulmonary Valvuloplasty has become the standard treatment for congenital pulmonary stenosis. There are enough data regarding its safety and efficacy of this mode of treatment. However, there is less data about its safety in patients with severe right ventricular dysfunction and pericardial effusion.
Objectives: To see the outcome of patients with severe right ventricular dysfunction and pericardial effusion who have undergone percutaneous balloon pulmonary valvotomy in a sub-Saharan center.
Methods: We compared echocardiographically measured peak transvalvular gradient measured before intervention and 3 months after intervention between patients with and without severe right ventricular dysfunction. Assessment of RV function was mainly based on visual judgment of RV contractility on 2D echocardiography and Tricuspid Annular Plane Systolic Excursion (TAPSE).
Results: from Jan 2009 to November 2012, a total of 49 patients have undergone the procedure. All patients had severe pulmonary stenosis defined by echocardiographic peak transvalvular gradient of ≥ 70 mmHg. Of the 49 patients, 20 had severe right ventricular dysfunction. Thirteen patients had mild to severe pericardial effusion. Mean age at intervention for those without RV dysfunction was 8.10 ± 5.75 years while for those with RV dysfunction was 11.28 ± 6.68 years. Peak systolic pressure gradient was 109.20 ± 30.08 mmHg versus 141.73 ± 38.01 (p value .005). Oxygen saturation was $95.12 \pm 6.74\%$ in those without RV dysfunction versus $77.13 \pm 13.13\%$ in group with RV dysfunction (p value .000). There were 3 mortalities, from the RV dysfunction group. The deaths were due to reperfusion injury resulting in pulmonary edema. Prolonged intubation or stepwise dilation has been employed to decrease mortality in patients with severe RV dysfunction. At 3 months following the procedure, echocardiographically measured peak gradient for those without RV dysfunction was 30.05 ± 15.10 mmHg and for group with RV dysfunction, was 36.73 ± 20.81 mmHg (p value .307).
Conclusion: the outcome of percutaneous intervention in those patients with severe RV dysfunction and pericardial effusion was as good as those without, if care is taken to avoid the immediate post procedure mortality due to reperfusion injury. Prolonged intubation and stepwise dilation strategies appear to help overcome mortality due to reperfusion injury in these patients.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

P-164

Ten Year Experience with Transcatheter Closure of Perimembranous Ventricular Septal Defects Using the Amplatzer Asymmetric Perimembranous Ventricular Septal Defect Occluder in Children: A Multicenter study

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Introduction: We present 10 year experience with 78 patients (pts) with perimembranous ventricular septal defects (PMVSDs) who underwent transcatheter closure at 4 different Institutions with the Amplatzer asymmetric PMVSD occluder.
Methods: The age of the pts ranged from 0.3 to 15 years. During the study period 31 other patients were excluded from transcatheter closure because they did not fulfil the patient selection criteria (distance less than 2 mm from the PMVSD to the aortic valve, size of VSD in relation to patients age).
Results: The devise was permanently implanted in 72/78 patients. Complete occlusion of the communication at six month, one-year, and 2-year follow-up was observed in 93%, 97%, and 97% patients, respectively. Main complications included: Early. Were observed in patients less than one year (body weight < 8 Kg) and included: a. Device embolization (2 patients-catheter and surgical removal, respectively), b. severe procedural bradycardia (5 pts) and c. Mobitz II and complete heart block heart in 3 and 1 patients respectively. (sinus rhythm after device removal). Late (follow-up 6 months–10 years). Complete heart block was developed in one patient 4-year old with Down syndrome. No other patient developed heart block during the follow-up. Three patients developed mild aortic regurgitation. In one of them the regurgitation was not seen at the 1-year follow-up. No other complications were observed.
Conclusions: Transcatheter closure using the Amplatzer APMVSD occluder is as a safe and effective nonsurgical alternative that should be offered in properly selected patients with PMVSDs. It should be noted, however, that with the current design of the occluder-delivery system the procedure carries an increased risk in small patients less than one-year of age. Finally, due to anatomic reasons, this therapy cannot be offered to significant number of patients with these defects.

P-165

Recanalisation of completely occluded left pulmonary arteries

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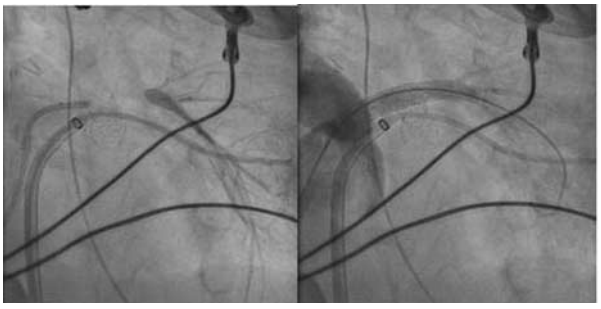
Objective: Total occlusion of pulmonary arteries is a rare and serious complication after congenital heart disease surgery. We sought to assess the technical difficulties, different access approaches and results of left pulmonary artery (LPA) recanalisation in a group of patients with complete occlusion.
Methods: Review of the medical and cardiac catheterisation data of 3 patients with postoperative LPA occlusion. Patient 1 had tetralogy of Fallot with pulmonary atresia and had undergone complete repair with a Contegra conduit. One year post-repair she was found to have complete LPA occlusion. Patient 2 had Fallot with pulmonary atresia and major aortopulmonary collaterals. She underwent unifocalisation and central shunt placement; 8 days later the LPA was found to be occluded. Patient 3 had univentricular heart anatomy and had undergone a Glenn shunt; on the pre-Fontan catheterisation the LPA was found to be completely occluded. Procedural details, complications and mid-term outcome were recorded.
Results: All three patients had successful LPA recanalisation and stenting. Approach was an open hybrid procedure with placement of a sheath in the distal part of the Contegra in patient 1 (age 2 yrs, 12 kg), percutaneously through the left axillary artery in patient 2 (age 4 months, 3.5 kg) and percutaneously through the right internal jugular vein in patient 3 (age 4 yrs, 16 kg). Pulmonary venous wedge angiograms were used in all patients to delineate the distal LPA segment and served to guide the recanalisation process. Creation of a track was performed with small diameter balloons after passage of a 0.014" guide wire and establishment of a vessel lumen was achieved with the placement of premounted Genesis stents. No complications were encountered. All patients are well at 0.5–3 yrs follow-up. Stents are still in situ and functioning well in two patients. In patient 2, the stent was removed at complete repair and the LPA was reconstructed with a Goretex tube.
Conclusion: Postoperative branch pulmonary artery interruption is a rare and serious complication. An attempt to recanalise the vessel should be made as soon as it becomes apparent, in order to ensure that the thrombosed lumen can be safely accessed and stented.



P-166

The use of peripheral premounted Palmaz-Genesis stents in patients with congenital heart disease

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Background: The use of peripheral premounted Palmaz-Genesis stents in patients with congenital heart disease has increased rapidly in the past few years. They are available in diameters of 3–10 mm and different lengths up to 8 cm. There are no available follow-up data following their implantation in cardiac chambers or vessels. The aim of this study was to assess the utility of those peripheral stents, complications and patient mid-term follow-up.
Methods: Retrospective review of the medical records and catheterizations of all patients who underwent stenting of cardiac vessels or the atrial septum at the Evelina Children's Hospital, London between Dec 2005-Jan 2010 and the Mitera Children's

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Hospital, Athens, Greece, between Nov 2010–Dec 2012. Patients in whom stents were implanted in the arterial duct during a hybrid procedure for hypoplastic left heart syndrome were excluded.
Results: 26 patients with median age of 10 ± 3 months and weight 6.5 ± 2.9 kg underwent implantation of 34 premounted Palmaz-Genesis stents (PMS) during 26 cardiac catheterization procedures. The stents were implanted in branch pulmonary arteries (n = 20), atrial septum (n = 5), SVC (n = 3), patent arterial ducts (n = 2), pulmonary valves (n = 2), Fontan fenestration (n = 1) and Blalock-Taussig shunt (n = 1). One patient developed pulmonary haemorrhage due to pulmonary trauma from the edge of the wire. All atrial septum stents have remained in situ. No embolisation or stent migration has been recorded. There have been no procedure-related deaths. The patients have been followed-up for a median period of 12 ± 21 months following stent implantation. Intimal proliferation or severe re-stenosis have not been recorded. Two patients have undergone stent re-dilation to match somatic growth. One patient with stent implantation in a branch pulmonary artery has had the stent excised during subsequent surgery.
Conclusions: The use of premounted Palmaz-Genesis stents in patients with congenital heart disease is feasible and effective, avoiding the need for re-operation in patients with branch pulmonary artery stenosis and securing a good size intracardiac or extracardiac communication when mixing between venous and arterial blood is required. The long-term results depend on the patient's individual anatomy and need for re-operations.

P-167
Progressive Aneurysmal Dilation of CAF after Transcatheter Closure: Successfully Treated by Second Occlusion
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A 3-year-old boy presented continuous murmur and heart failure from a large coronary artery fistula (CAF) of the right coronary artery (RCA) to the right ventricle (RV). (Figure 1) The fistulous tract was significantly dilated at its distal segment before entering into the RV. The diameter of aneurysm was 25 mm and RV orifice was 4 mm. As for concerns of significant heart failure and risk of coronary insufficiency, the CAF was percuteneously closed by Amplatzer duct occluder I 8/6 at its RV orifice. (Figure 2) No residual shunt after the procedure.
However, he was lost to follow-up. Three years later, he was referred from local pediatric cardiologist because of a huge vascular structure compressed the RV free wall. (Figure 3).
CT angiography showed a huge blind pouch fistulous track (31×42 mm) with RV compression. (Figure 4) Compared to previous measurement of the aneurysmal part prior to TCC, the size of the pouch was significantly increased. In addition, there was evidence of reconstitution posterior descending artery (PDA), distal RCA and part of middle RCA by the distal left anterior descending artery (LAD).
RCA angiogram showed device was nicely seated at the RV exit point without residual shunt. Small native RCA branch was demonstrated just proximal to fistulous waist. LCA angiogram showed normal branch with good perfusion supplied to the right coronary territory. Test occlusion, by a balloon wedge catheter, showed no ST-T change. The aneurysm was successfully closed by Amplatzer vascular plug (AVP) II 12 mm with tiny residual shunt. (Figure 5) Normal ECG was demonstrated after the procedure.
Two weeks later, the echocardiogram revealed clot formation in the aneurysm. The plug was well seated without residual shunt. There was good LV and RV contraction without abnormal regional wall movement and no evidence of myocardial infarction via the EKG. (Figure 7-8)
Conclusion: Long-term follow up after CAF closure is indicated since progressive dilation of the blind pouch fistula may occur. Proximal and distal occlusion of the track should be performed to prevent future aneurysmal rupture.

P-168
Coronary Thrombo-embolism in Hypoplastic Left Heart Syndrome - A treatable cause of interstage mortality
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Introduction: Mortality rates in hypoplastic left heart syndrome (HLHS) continue to improve since the introduction of staged surgical palliation. However, inter-stage deaths continue to be challenging to prevent. This case presents a child with HLHS post Glenn shunt presenting with acute ST-elevation myocardial infarction (MI). He was treated using a well established procedure in treatment of acute MI in adults. Treatment of coronary thrombo-embolism in the setting of uni-ventricular circulations has not been previously reported. This case demonstrates the importance of any evidence of myocardial ischaemia in these patients and presents a novel and potentially lifesaving therapy for consideration.
Methods: A three-year old boy with HLHS post Glenn shunt presented with sudden onset, classical symptoms and ECG changes consistent with acute ST-elevation MI. Echocardiography showed acute deterioration in right ventricular systolic function. He was initially treated with thrombolysis and inotropic support for 24 hours but inferior ST-elevation persisted with development of Q-waves and inotropic dependence. He was therefore taken to the catheter laboratory. Angiography showed a dominant right coronary artery with almost total occlusion of the distal right main coronary artery. Using an Export AP aspiration catheter (Medtronic, Minneapolis MN), a large thrombus was aspirated and repeat angiography showed complete thrombus resolution with normal vessel patency. There were no procedural complications.
Results: There was an immediate improvement in ventricular function post-procedure. He was commenced on beta-blocker and ACE inhibitor and was discharged home approximately three weeks post-intervention. He has been seen at one and six weeks and 3 months following discharge and his echocardiogram shows ongoing functional improvement.
Conclusions: This case illustrates the novel application of a proven adult coronary intervention in complex paediatric congenital heart disease. This has not been reported previously. The importance of this intervention is because the entire circulation is dependent on an essentially single coronary artery system. The effects of acute occlusion therefore cannot be mitigated by collateralisation leading to more profound circulatory effects. Furthermore, it suggests one possible aetiology of inter-stage

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

deaths. Finally, the case demonstrates the additional benefits of collaboration between adult and congenital cardiologists providing alternative approaches for challenging situations.

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Initial Experience with Valeo Pre-mounted Stents for Treatment of Pulmonary Arterial Stenoses

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Introduction: Pulmonary arterial stenoses are a source of significant morbidity or mortality in post operative congenital heart disease patients. Stent implantation is effective but challenging in young children owing to the large vascular access required and difficulty tracking long sheaths and stiff stent-balloon assemblies. Stents also need to be ultimately post-dilated to adult size. The Bard Valeo peripheral vascular stent is low-profile, premounted, flexible, open cell and can be dilated to 20 mm. Therefore it can potentially overcome these technical difficulties. In this series we assess the performance of this stent in treating pulmonary artery lesions.
Methods: At cardiac catheterisation proximal, minimum and distal pulmonary artery size, and right ventricle:femoral artery (RV:FA) pressure ratio were measured. Pressure gradients were measured across stenoses. Seven stents were implanted in six patients (one required bilateral stent implantation) using standard techniques. All patients were post-operative (3 PA/VSD/MAPCAs, 2 TOF, 1 TGA). Median age was 3 years (3–10 years) and median weight was 22 kg (11.8–30.2 kg). Final measurements included minimum final diameter, RV:FA ratio and residual gradient across stents.
Results: All stents tracked easily and although long sheaths were used, stents were passed bare across the lesions in 71% (5/7) cases. There were good angiographic and haemodynamic results with no procedural complications. Femoral venous access size was median 7.5 French (5–9F). Minimum vessel diameter improved from mean 4.9 mm (3.4–6.8 mm) to 9.4 mm (8.1–10.0 mm). There was no significant stent recoil on balloon deflation. Final stent diameter as a ratio to distal vessel size was mean 1.08 (0.87–1.25). Pressure gradients across stenoses reduced from mean 18.5 mmHg (16–21 mmHg) to 7.5 mmHg (6–9 mmHg). RV:FA ratio reduced from mean 0.73 (0.72–0.75) to 0.54 (0.54–0.55).
Conclusions: Valeo stents require smaller vascular access than other stents that have the potential to be dilated to adult size, do not require placement of a long sheath and have excellent trackability. They have sufficient radial strength to deal with pulmonary artery stenoses. Although the stents can potentially be expanded to 20 mm, long term follow up will be required to assess whether this is achievable by redilation. Valeo stents provide a good option in treating pulmonary artery stenoses in small children.

P-170

Are prophylactic antibiotics indicated during interventional cardiac catherisation where devices are not being implanted?

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Introduction: For 50 years practice has been to use prophylactic antibiotics in children with congenital cardiac disease prior to surgical intervention. With the advent of interventional cardiac catheterisation this practice has continued. The authors were unable to find evidence to support prophylaxis in routine non-device related interventional procedures. Our hypothesis was that in elective interventions under aseptic conditions, there would be no benefit gained from the routine administration of prophylactic antimicrobial agents.
Methods: There is no unifying protocol at our institution for prophylactic antibiotics in interventional procedures. Two of our operators give antibiotics, whilst three do not unless specific circumstances indicate they may be required. A review of the hospital database, patient notes and catheter laboratory records was performed looking for the administration of antibiotics, indication and type of catheter procedure performed and for evidence of subsequent local wound infections or incidences of bacterial endocarditis.
Results: Over a 2 year period there were 436 interventional procedures; with 212 excluded principally due to device placement during the procedure. 142 of the remaining procedures were interventions such as atrial septostomy and balloon dilations. There were also an additional 82 electrophysiological (EP) studies with ablation procedures. 81 (36.3%) of these interventions received routine prophylactic antibiotics with Flucloxacillin (unless penicillin allergic) whilst 143 did not; including all 82 of the EP studies. In our entire cohort of 224 interventions there were no incidences of post-operative infection. There were 4 deaths within 30 days of the procedure, none of which were attributed to the index catheter procedure.
Discussion: There is no evidence to support the use of routine prophylactic antibiotics to date. The results of our study show that there are no adverse outcomes in children undergoing non-device related cardiac interventions. There are many reasons not to administer an antibiotic without good evidence supporting its benefit, principally, potential for allergic reaction, propagation of antibiotic resistant organisms, additional financial costs, and the safety and efficiency benefits associated with streamlining procedures by minimising unnecessary steps.
Conclusion: There is no evidence to support the administration of prophylactic antibiotics in routine non-device related interventional catheterisation.

P-171

Long-term clinical experience with Amplatzer Ductal Occluder II for closure of the persistent arterial duct

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Objectives: To describe the long term clinical experience and follow-up of the Amplatzer Ductal Occluder II (ADO II) in a single tertiary referral centre.
Methods: All patients undergoing attempted transcatheter closure of persistent arterial duct (PDA) with the ADO II were included. Data including demographic, clinical, echocardiographic parameters, procedural details and outcomes were reviewed retrospectively.
Results: 62 patients (46 female) with a median age of 1.2 years (range 0.43–76 years) and a median weight of 9 kg (range 4.7–108 kg) underwent attempted transcatheter ductal closure with the ADO II from March 2008 until December 2012. Retrograde arterial approach was used in 2 patients. In one patient with Loey Dietz syndrome and a large aneurysmal duct, 2 occluders were inserted from both transarterial and transvenous ends with good result. Ductal morphology varied but the majority were large and tubular or tortuous. Device sizes used were 3/4 (n = 14), 3/6 (n = 5), 4/4 (n = 11), 4/6 (n = 8), 5/4 (n = 7), 5/6 (n = 10), 6/4 (n = 3) and 6/6 (n = 2). The median fluoroscopy time was 7.4 minutes. ADO II was released in 60 patients (96.7%). Two patients had significant residual shunting following deployment of ADO II and underwent closure with

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Amplatzer ductal occluder I (ADO I). Another four patients required ADO II resizing following deployment of the initial device. Device embolisation of ADO II to the pulmonary artery occurred in 5 patients. All occluders were retrieved successfully at a second catheter procedure. Of these, one underwent successful surgical closure; three were closed with ADO I; and one was closed with a larger ADO II. Complete occlusion on echocardiography was noted in 87% of the remaining occluders (48/55) pre-discharge and 98% at first follow up (54/55). One patient had persistent mild flow acceleration (2.5m/s) in the left pulmonary artery at follow up.
Conclusions: The ADO II is highly effective for occlusion of arterial ducts with variable anatomy from either an arterial or venous approach with a low profile delivery system. Stable occluder position is highly dependent on accurate device diameter sizing, good quality imaging to visualise device configuration after deployment, and operator experience.

P-172
Long-term follow-up 10 years after catheter closure of atrial septal defect in 241 children
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Objectives: After the first percutaneous ASD closure (1975) and material improvement with the Lock Clamshell® in 1985 and Amplatzer® devices in 1995, several teams published results, limited to small numbers of patients and/or short-term follow-up (FU). This study analyzed effectiveness and long-term FU after ASD, PFO or fenestration cath closure in a large number of children in a single institution.
Methods: This retrospective study included 258 children after attempted cath closure from 1/2000 to 4/2011. Seventeen children have ultimately been operated. Analysis focused on 241 children who underwent effective percutaneous closure. Patients were divided in 3 groups: ASD (203 patients), PFO (10 patients) and fenestration post-Fontan repair (28 patients).
Results: Device closure was effective in 241 patients (93.4%), 17 ASD patients were operated (ASD to large for device closure in 15 and device embolization in 2). Mean age at closure was 8.2 ± 4.3 yrs (range 0.5–17.8 years) and weight 29 ± 18 kg (range 3.2–108 kg). Major complications observed in 5 patients (2%) were 3 device embolization (2 treated by surgery and 1 device recovered by cath), 1 pericardial effusion requiring drainage, 1 stroke after Fontan fenestration closure. Minor complications consisted mainly of transient arrhythmias (5.4%) with spontaneous resolution or under medication. The major strength of this study was a complete FU obtained in 98.8% of the patients (mean 5.5 ± 3.3 years, range: 0.1–12.2 years) showing no further complication. After ASD closure, the number of symptomatic patients (exercise intolerance, growth delay, recurrent respiratory infections) diminished from 47% to 13%. Echo showed a complete occlusion in 74.3% of the patients at hospital discharge, and 93.5% at late follow-up (ASD 94%, PFO 100%, Fenestration 86%).
Conclusions: This study is one of the few with extended FU and a large number of pediatric patients. Failure, success and complications rates were similar to those found in the literature and compared favorably with surgical closure. Cath closure was effective with disappearance of clinical symptoms and the absence of significant residual shunt on echo. Major procedural complications were rare and long-term FU revealed no complications.

P-173
Risk of cancer associated with cardiac catheterization procedures during childhood: a cohort study in France
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Objective: Children with congenital heart disease frequently undergo cardiac catheterization procedures for diagnostic or treatment purposes. Despite the clear clinical benefit to the patient, the complexity of these procedures may result in high cumulative radiation exposure. Given children's greater sensitivity to radiation and the longer life span during which radiation health effects can develop, an epidemiological cohort study is being launched in France to evaluate the risks of leukaemia and solid cancers in this specific population.
Methods: The study population will include all children who have undergone at least one cardiac catheterization procedure since 2000 and were under 10 years old and permanent residents of France at the time of the procedure. Electronically stored patient records from the departments of paediatric cardiology of the French national network for complex congenital heart diseases (M3C) are being searched to identify the children to be included. The minimum dataset will comprise: identification of the subject, characteristics of the procedure and technical details, such as fluoroscopy time and dose area product (DAP), which are needed to reconstruct the doses received by each child. The cohort will be followed up through linkage with the two French paediatric cancer registries, which have recorded all cases of childhood leukaemia and solid cancers in France since 1990 and 2000, respectively. Radiation exposure will be estimated retrospectively for each child included in the cohort. Organ doses, especially to the lung, the oesophagus, and the thyroid will then be calculated with PCXMC software.
Results: In all, 4500 children with catherizations between 2000 and 2011 have been already included in the cohort, and recruitment is ongoing at the national level. The study is expected to finally include a total of 8000 children.
Conclusion: This French cohort study is specifically designed to provide further knowledge about the potential cancer risks associated with paediatric cardiac catheterization procedures. It will also provide new information on typical dose levels associated with these procedures in France. Finally, it should help improve awareness of the importance of radiation protection in these procedures.

P-174
Overexpanding a large PTFE-Conduit with a Melody® Transcatheter Pulmonary Valve to an advantageous size
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Introduction: Percutaneous pulmonary valve implantation (PPVI) is an excellent alternative method in paediatric patients with right

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

ventricular outflow tract (RVOT) dysfunction to avoid open-heart surgery. We carried out PPVI in a nine-year-old girl presenting gradually worsening tricuspid valve insufficiency due to right-ventricle dilatation caused by free pulmonary regurgitation of a 16-mm polytetrafluorethylene (PTFE) valve-less conduit. Clinically she displayed normal exercise tolerance, no signs of heart failure or cardiac rhythm anomalies.
Methods: The intervention was carried out under general anaesthesia. She underwent a complete haemodynamic examination once transcutaneous femoral vessel access had been obtained. Angiocardiography revealed the aortic bulbus and coronary arteries in a safe distance from the RVOT. In addition, a coronary angiogramme was combined with balloon inflation in the conduit to rule out any coronary arterial compression. We successfully implanted a 22-mm bovine valve integrated in a covered stent (Melody® Transcatheter Pulmonary Valve) without pre-stenting the landing zone.
Conclusion: The optimal timing of intervening in the presence of severe pulmonary regurgitation is unclear. Generally speaking, severe pulmonary regurgitation after an RVOT intervention is well tolerated for years, but it is not a benign cardiac lesion. The clinical evidence strongly supports a timely intervention before the patient becomes symptomatic. The prevailing indication for pulmonary valve replacement in this patient was the gradual increase in tricuspid valve insufficiency. The decision favouring a catheter-based intervention rather than open-heart surgery was challenging. A PTFE conduit may not be the best candidate for PPVI because of its relative rigidity. Additionally, open-heart surgery would make tricuspid valve reconstruction possible. On the other hand, by alleviating the pulmonary regurgitation alone, the right ventricle's dilatation in turn leading to tricuspid valve insufficiency may resolve. We demonstrate an excellent outcome with PPVI in a PTFE conduit. Fortunately, the internal conduit diameter was enlarged without any complications from 16 mm to 18.5 mm at the narrowest position. The pulmonary valve's z-score raises from - 0.81 to +0.01 providing a new pulmonary valve's conduit-to-patientsize enabling postpone open-heart surgery for several years.
e enabling postpone open-heart surgery for several years.

P-175
A newborn with partial trisomy 10q and diminutive pulmonary arteries in DORV with critical right ventricular outflow tract obstruction and major aortopulmonary collateral arteries: How invasive to treat?

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Background: Distal 10q trisomy is an extremely rare chromosomal disorder usually caused by an unbalanced translocation, including a distinctive, dysmorphic appearance, growth retardation and psychomotoric disorders. Patients with trisomy 10q24->qter tend to be associated with cardiac, renal and ocular abnormalities. However, the severity varies greatly among affected individuals. Ethical aspects should therefore be carefully considered, but early rehabilitation is recommended in the setting of diminutive pulmonary arteries (PAs) to achieve favourable PAs growth and the goal of biventricular repair in the future.
Case and results: Mother 1G1P, no consanguinity, 40 SSW, birth weight 2765 g. The female newborn presented multiple dysmorphic craniofacial features including microstomia, a receding chin with a laryngeal aberrance, dysplastic, low-set ears, an epicanthic fold, and blepharophimosis along with microphthalmia. She also had musculoskeletal manifestations such as muscular hypotonia, talipes calcaneus and contracted joints affecting hands and limbs. Chromosomal analysis showed an unbalanced translocation of 10q [(t1;10), 10q25.2->10qter].
Echocardiography revealed a double outlet right ventricle (DORV) with critical right ventricular outflow tract obstruction (RVOTO) and extremely diminutive pulmonary arteries (PAs) with competitive major aortopulmonary collateral arteries (MAPCAs). To open the right ventricular outflow tract (RVOT) and subatretic pulmonary valve, we decided on initial palliation with balloon dilatation on day 7, followed by RVOT stent implantation eight weeks later. Repeated catheterisation with stent dilatation, valvuloplasty and embolism in the MAPCAs showed adequate PAs growth.
The child developed satisfactorily and underwent specific early ophthalmic and orthopaedic treatments. Parental bonding developed very well. Biventricular repair with a 12 mm Contegra valve was successful at the age of 14 months in the presence of well-developed PAs.
Conclusion: Instead of early high-risk unifocalisation, this early interventional approach with RVOT stent and repeated interventional cardiac catheterisation was a safe and effective treatment strategy that encouraged PAs growth. This approach provided time for early therapy of extracardiac dysmorphisms and for the family's ethical considerations.

P-176
Temporal witnesses about the evolution of interventional cardiac catheterisation for CHD: Interview with K. Amplatz (Minessota, USA)

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Background: The German Society of Paediatric Cardiology (DGPK) has started a project to document important developments of Paediatric Cardiology over the last 5 decades in Europe. Honorary members of DGPK are considered as leading pioneers of the evolving speciality of Paediatric Cardiology beginning in 1960. For the time being our German society (DGPK) has 7 honorary members (age > 70years). One of them is e.g. Kurt Amplatz (Minnesota, USA).
Aim: The aim of this project was to interview these temporary witnesses and to document their influence in establishing this new field of Paed. Cardiology.
Method: An interview with standardised questions was performed with each of the honorary members. The interviews were recorded on video. As an example of such an interview we will present video- sequences and photos from the interview with Kurt Amplatz (recorded in Germany, October 2012).
Interview with K. Amplatz
K. Amplatz was born 1925 in Austria. After his study of medicine in Innsbruck (Austria) in 1951, he immigrated into the USA in 1952. He thought to have better possibilities "in the new world" to work in an innovative surrounding. He considers himself as a tinkerer and experimenter since his childhood. From 1957 up to his retirement in 1999, he was appointed as a professor in Radiology, University of Minnesota (USA). During this time up to now he has invented numerous new devices to improve cardiac catheter technique e.g. specially shaped diagnostic cardiac catheters, heparin coated guide-wires, retrieval and occlusion devices. His most famous inventions were the Amplatzer devices for the occlusion of ASD, PDA, vessels and VSD. To manufacture

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

these devices he founded the AGA Company starting in a garage with an initial investment of 300 Dollar in 1995. Nowadays K. Amplatz is worldwide acknowledged as a one of the leading pioneers for interventional cardiology for CHD holding more than 30 patents.
Conclusion: This interview shows exemplary the importance to document the experience and opinion of leading contemporary witnesses about the evolution of interventional cardiology for CHD.

P-177
Transcatheter patent ductus arteriosus closure with echocardiographic guidance: Can Radiation exposure be reduced?
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Objective: The radiation dose from interventional cardiac catheterization is particularly relevant when treating children because of their greater radiosensitivity compared with adults. Transcatheter closure of patent ductus arteriosus (PDA) as well as other more complex pediatric interventions has raised concerns regarding radiation exposure. The purpose of this study is to show how to perform transcatheter closure of PDA in children while giving less ionized radiation and to prove that the amount of radiation as well as unnecessary additional contrast material can be reduced.
Methods: In this prospective study, we included 63 children those had transcatheter closure of PDA. Following appropriate device selection based on PDA morphology and diameter; transthorasic echocardiography images and control aortography findings were analyzed. Results were compared for presence of residue, the amount of contrast, radiation dose and follow-up data.
Results: The mean age, weight, and minimum ductal diameter were 5.1 ± 4.4 years, 19.3 ± 14.7 kg, and 3.0 ± 1.2 mm, respectively. Following devices were used during procedure: Gianturco coils (10/63), Amplatzer Duct Occluder (ADO, 31/63), Flipper coils (19/63), and Amplatzer vascular plug (3/63). Total durations of procedure and scopy, the amounts of radiation dose and contrast were as follows; 56.4 ± 19.4 mins, 12 ± 6.4 mins, 28.1 ± 14.7 cmgy/cm^2/kg and 4.2 ± 2.3 cc/kg. In control aortography shortly after the procedure, residual shunt was detected in different levels in 39.7% of patients and 9.5% demonstrated residual shunt in real-time echocardiography. In echocardiographic control one day later, occlusion percentile was 98.4%. In control aortography, exposure of radiation was 13.3% of total and the amount of infused contrast was 27.2% of total. If residual shunt was seen in control aortography but not in echocardiographic evaluation; those patients had no residual shunt in the follow-up.
Conclusion: With the appropriate device selection, patients may be exposed less radiation and contrast material if echocardiographic evaluation is performed after transcatheter closure of PDA instead of last control aortography injection.

P-178
Transcatheter Closure of Perimembranous Ventricular Septal Defects (PmVSD) with Nit Occlud® Lê VSD Coil: Early and Mid-Term Results
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Background and Aim: Percutaneous techniques have been developed in order to reduce the impact of drawbacks of surgery. Since the first VSD closed by a transcatheter approach by Locket al. various devices and techniques have been used. Here in we present our early and mid-term results of patients who underwent transcatheter closure of VSD using Nit Occlud® Lê VSD Coil.
Patients and Methods: We retrospectively reviewed our echocardiographic, angiographic, electrocardiographic and 24-hour Holter ECG data of transcatheter closure of VSD using Nit Occlud® Lê VSD Coil.
Results: Our approach to place a Nit Occlud® Lê VSD Coil were successful in 13/14 patients (93%). The median age at closure was 7.6 years (range 1.7–17 years) and the median weight was 25.5 kg (range 10 to 58 kg). All of the PmVSDs were with ventricular septal aneurysm except one. The median device size used was 8/6 (range 8/6 to 12/6). The median procedure and fluoroscopy time were 93.8 minutes (range 40–180 min.) and 32 minutes (range 13.3–67.4 min.) respectively. The median aortik rim was 4.7 ± 1.8 mm (range 0–8 mm). When the procedures were completed the total occlusion rate was 11/13 (84%). One patient's residual VSD closed spontaneously in a month. But in the other one we observed severe hemolysis which was resistant to medical therapy. An additional coil was placed in to the residual VSD but it was not successful to reduce hemolysis. After seven days this patient underwent surgical VSD closure. None of the patients developed valvular regurgitation during follow up. During the procedure or in follow up (mean 7.7 months) none of the patients developed high degree AV-block. No deaths occurred.
Conclusion: Transcatheter closure of PmVSD with shorter or without aortic rims and ventricular septal aneurysms can be performed by Nit Occlud® Lê VSD coil. The most important advantage of this device is seem to be free of high degree AV block. For the evaluation of the clinical implications and long term complications, studies with longer follow up periods are needed.

P-179
Evaluation of the efficacy of ductus arteriosus stenting in neonates and infants to maintain pulmonary blood supply in cyanotic congenital heart disease: acute and midterm outcome after stent implantation, single center experience
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Introduction: We aimed to assess the efficacy and outcome of transcatheter ductus arteriosus stenting in newborns and infants with ductal-dependent or decreased pulmonary circulation. With today's generation of coronary stents which have better profile, flexibility and trackability, ductal stenting may be achieved safely and with considerably less difficulty than previously described.
Methods: Between April 2010 to January 2013, fourty patients underwent PDA stenting after full assessment by echocardiogram and angiogram, 4 of patients had PA & VSD, 19 patients had PA with intact ventricular septum, 6 patients had critical pulmonary stenosis, 4 patients had hypoplastic left heart syndrome and 7 patients had different types of congenital heart diseases with ductal dependency. 7 patients had radio frequency perforation of the pulmonary valve at the same time.
Results: Stenting was successfully performed in 97.5% of the patients. Median age of patients is 8,5 days and mean weight is 3.14 ± 0.74 kg. Procedure related death observed in one patient, just after stenting due to hypotension. The mean post procedural follow-up period was 190 days. The causes of death after stenting were pulmonary hemorrhage ($n = 1$), thrombus formation after

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Glenn procedure (n = 1), sepsis (n = 7), pneumonia (n = 2) and one patient sudden cardiac death. Mean arterial oxygen saturation before and after stent implantation was 78.5% and 87.4%. Seven patients had Glenn procedure performed successfully, five of them still on follow up. Four patients who have hypoplastic left heart syndrome, had hybrid procedure performed, one of which was successfully operated with hybrid stage II and the other three died after hybrid procedure due to sepsis. Central shunt operation is planned for one patient due to stent obstruction. Two patients are now ready for Glenn operation and soon will be operated. Three patients dropped out of follow up. Remaining patients are periodically followed up until Glenn procedure.
Conclusion: Stent implantation of ductus arteriosus can be a good alternative to surgery for initial palliation in severely cyanotic newborns and infants until the latter stage palliative surgery or total repair. Patients with ductal stenting may provide growth for pulmonary artery which achieves more additional time for surgical repair.

P-180
CP Stent Implantation in the Children with Coarctation of the Aorta: Short-Intermediate-Long Term Results From Turkey in a Single Centre

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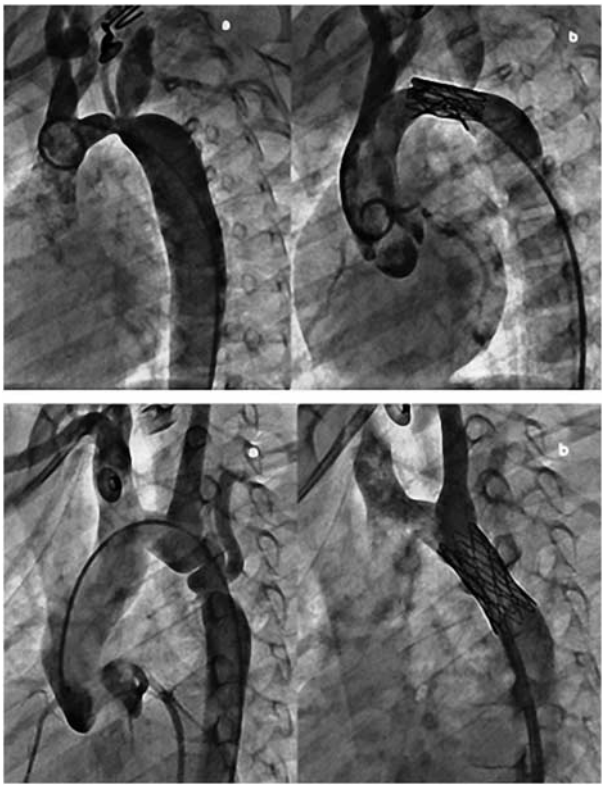
Objectives: Our aim was to evaluate patients with coarctation, who were treated by percutaneous stent implantation.
Method: Patients with coarctation (n = 36, 26 male) who had been treated with 39 stents (12 bare, 27 covered) were evaluated. The demographics, procedural and follow-up data were recorded from hospital registers and compared among the patient specifications (e.g. weight, coarctation nature).
Results: Mean follow-up time was 39 ± 16 months. There was a statistically significant difference between the patients with native coarctation (n = 17) and recurrent coarctation (n = 19) in terms of pre-procedural blood pressures, systolic gradients, coarctation diameters and the ratio of coarctation site diameter to descending aorta (CoA/DAo). While all patients received antihypertensive drugs before the procedure, the drug was discontinued in 27 patients during follow-up ($p < 0.001$). The procedure was successful in all patients. Stent migration was observed in four patients (11.1%) (all of them with recurrent coarctation) and peripheral arterial injury was seen in three patients (8.3%). On average 21(6–42) months after the procedure, six patients underwent cardiac catheterization. Four of these patients underwent balloon dilatation. At least two years following the procedure, multislice CT was performed in 21 patients (58.3%). The patients who were evaluated by MSCT revealed no pathology such as restenosis, intimal proliferation in the lumen of the stent, aneurysm formation, stent fracture and migration. Five patients (13.9%) were <20 kg, 16 patients (44.4%) were <30 kg (11–70). There was no statistically significant difference between the five patients weighed <20 kg and the other patients in terms of demographic-procedural characteristics, procedure success and complication rates and follow-up data (Figure).
Conclusion: Stent implantation for coarctation of the aorta is a method with satisfactory results in the reduction of both invasive and non-invasive gradients and in the efficient enlargement of the lesion area. CP stents may also be applied to the selected patients whose anthropometric measures are below the age and weights recommended by the manufacturer. Although multislice CT results alone do not seem to be a marker in the determination of patients who need a re-intervention, multidisciplinary evaluation using the available imaging methods seems to be the best follow-up model in the patients at risk.



P-181
Causes and management of systemic arterial desaturation in children with congenital heart defects and functionally single ventricle after the Fontan procedure. A single center experience

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Introduction: Important complications in children with CHD and single ventricle after the Fontan procedure (FP) include systemic arterial desaturation (Sa02).
Objective: To assess the Sa02<90% causes and management in post-Fontan pts, we analyzed cardiac catheterization (CC) and intervention results in a total 52 children, between January 2000 and December 2012.
Material/Methods: Sa02:56%–89% was seen in 19 (36.5%) pts aged 1.5–21 years (x-7.5 ± 5.76 years), body mass: x-21.9 ± 12 kg. Depending on CHD type, the pts were divided into two groups: Group-I: HLHS (n = 8), and Group-II: other complex CHD's (n = 11). The pts underwent staged transition to their completed Fontan circulation, mostly (18) with fenestration.
Results: Sa02 in Group-I was x–84.8% ± 3.7% while in Group-II: x-80.1% ± 9.1%. Age at the Fontan procedure in Group-I (x = 3.73 ± 2.64 yrs) vs. Group-II (x = 4.4 ± 1.7 yrs) and time between surgery and catheterization (Group-I: x = 2.4 ± 4.3 yrs vs. Group-II: x = 4.5 ± 4.9 yrs) differed insignificantly (p = 0.5,

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

p = 0.3, respectively). In Group-I, 6 pts showed significant LPA stenosis treated with balloon angioplasty (BPA) and stents implantation, 1 – restrictive ASD demanding reoperation, 1 – recanalization of LSVC to LA, 1 – large fenestration (both closed percutaneously). One teenager with severe RV dysfunction was qualified to heart transplantation performed 3 mo later. In Group-II: 3 pts showed intracardiac tunnel-RA shunts (closed in 1 percutaneously, in 2 surgically), 3 had PA branch stenosis (PBS) (treated with BPA and stents implantation), 3 – multiple intrapulmonary venous fistulas and 1 - SVC-to-PV fistula (closure possibility in 1 child). Fenestration occlusion was performed in 1 of 2 qualified to this procedure. In all pts treated interventionally, Sa02 increased from x = 82.5% ± 4.6% to 89.2 ± 5%.
Conclusions: Most reasons causing systemic arterial desaturation in post Fontan pts was possible to correct interventionally. Their different nature: more frequent PBS in Group I (easier to treat) vs multiple v-v fistulas in Group II (difficult to eliminate) despite similar time from FP to CC was a result of CHD's type and initial stage of operation.

P-182
Use of covered Cheatham-Platinum stents in congenital heart disease
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Aim: Evaluate possibilities and safety of covered Cheatham-Platinum (CCP) stents in congenital heart disease (CHD).
Methods and results: Single-center retrospective CHD-database study of all CCP stents, 2003–2012. Three study groups: aortic coarctation (CoA), right ventricular outflow tract pre-stenting for percutaneous revalvulation (RVOT), and miscellaneous. Data expressed as median (range). 114 CCP stents in 103 patients, 67% male, age 16.8 years (4.2–71.2).
CoA group: 54 CCP stents in 51 patients: 3/54 for aneurysm exclusion, in 51/54 covering used "prophylactically" because increased risk for vessel tear. CCP stent was dilated to desired dimension at implantation in 39/51 patients, further stent dilation to optimal dimension or following somatic growth in 12/51 patients after 4.5 months (1.6–28). Overall, CCP stenting increased coarctation diameter from 6 (0–15) to 15 (10–20)mm ($p < 0.001$) and decreased PTP gradient from 23 (0–86) to 2 (0–5)mmHg ($p < 0.001$).
RVOT group: 39 CCP stents in 37 patients: the graft lumen had shrunken from nominal 21 mm (10–26) down to 13 mm (5–22); with the CCP stent the graft was redilated prior to revalvulation up to 22 mm (16–26, $p < 0.001$ vs stenosis).
Miscellaneous group: 21 CCP stents in 15 patients: closure of Fontan-circuit fenestration (n = 5), expansion of cavopulmonary conduit (n = 2), restoration of superior caval vein (n = 2) or pulmonary artery (n = 2) patency, relief of supra-pulmonary stenosis (n = 2), intrapulmonary anastomosis (n = 1), exclusion of aberrant pulmonary arteries (n = 1). Hybrid procedures in 3/15 patients to make sutureless connections in distal lung vessels. CCP stent as rescue treatment in 2/15 patients to seal iatrogenic bleeding.
Specific techniques were required to optimize the results with CCP stents in selected patients: stent delivery through 10F sheath with a thin low pressure low profile 7F balloon, flaring to seal the expected tear, delayed dilation, double wire delivery, retrograde puncture to reopen a side-vessel subclavian artery.
Conclusion: CCP stents can safely be used in CHD patients, requiring specific techniques in selected targets. The covering allows adequate sealing of existing or expecteded tears, thereby increasing the safety margin with more complete dilation in selected patients, and extending the possibilities of transluminal interventions.

P-183
The Amplatzer vascular plugs to occlude various shunts in congenital heart disease
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The Amplatzer Vascular Plugs (AVPs) are self-expandable devices made from a nitinol wire mesh that have been used to occlude various collateral vessels and shunts in congenital heart disease. We report here one centre experience.
From 2004 to January 2013, 65 AVPs were implanted in 53 pts (24 females and 29 males). AVPs were employed to occlude arterial systemic shunts (n = 14), venous systemic shunts (n = 11) mainly in Fontan circulation, arterio-venous fistulas in Rendu-Osler (n = 9), patent vertical vein (n = 8), fenestration (n = 4), Blalock-Taussig shunts (n = 3), coronary artery fistulas (n = 2), VSD (n = 1) and PDA (n = 1).
Mean age of patients was 13.9 ± 10.5 years, mean weight 28.8 ± 11 kg. The AVPs implanted included: Plug I (n = 31, mean size 8.3 ± 4.0 mm), Plug IV (n = 30, mean size 5.6 ± 1.4 mm), and Plug II (n = 4, mean size 17 ± 3.8 mm). Implantation succeeded in all but 3 in whom the plug was retrieved. One patient with PDA underwent surgical ligation subsequently, a second had successful occlusion of a Blalock-Taussig shunt with another plug, and the remaining had complete VSD occlusion with another larger device. One device embolized in pulmonary artery that could be retrieved during the procedure. The fluoroscopic time was 19 ± 10.4 minutes and radiation dose 26 ± 28 $Gycm^2$. Full occlusion was confirmed by control angiography, CT scan or echocardiography. During follow-up, no evidence of hemodynamic or vascular compromise was noticed.
AVPs are effective and safe in the percutaneous closure of collateral vessels and shunts. The AVP IV is really appropriate to occlude difficult-to-reach tortuous vessels/shunts using small delivery catheter; it is a good alternative to the classic detachable coil. Plugs I and II are appropriate for occlusion of larger vascular shunts. The choice of AVP is mainly determined by the anatomy and morphology of the shunt.

P-184
Transvenous Edwards SAPIEN valve implantation in the tricuspid position
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Ebstein anomaly is a complex disease and surgical results may be not as good as expected. We report here 2 patients in whom percutaneous tricuspid valve implantation was realised after previous surgical repairs. Two boys underwent tricuspid valve plasty at the age of 4 and 12 years respectively. Because of persistent regurgitation, both of them had subsequently tricuspid valve replacement by a bioprosthesis (27 mm Mosaic Medtronic valve in pt 1 at the age of 7 years and 33 mm Carpentier-Edwards Perimount in pt 2 at the age of 12.5 years) in combination with a Glenn procedure. During follow-up, both of them had dysfunctional tricuspid bioprosthesis. Percutaneous implantation

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

of the tricuspid valve performed under general anaesthesia and from a femoral venous approach at the age of 16 years in pt 1 and the age of 15 years in pt 2. Rapid pacing by a pacemaker lead placed in the left ventricle from the femoral artery was used during valve deployment. In pt 1 during balloon inflation, the 23 mm Edwards Sapien slipped into the right ventricle where it became free. Using the extrastiff guidewire, this stented valve was anchored to the tricuspid annulus by different overlapping stents (Intrastent, EV3 and a self-expandable stent) and then a second 23 mm Edwards Sapien was implanted with success within the annulus. In pt 2, pre-stenting of the tricuspid annulus was realized first with a bare stent (Intrastent, EV3), and then a 26 mm Edwards valve was implanted with success.

Tricuspid valve-in-valve implantation using the Edwards Sapien valve is feasible. To secure valve positioning, it may be advisable to prestent the tricuspid annulus to have adequate landing zone for further Edwards Sapien valve implantation. To conclude, the Edwards Sapien valve seems appropriate for implantation in the tricuspid position in selected patients with prior tricuspid valve surgery but additional long-term results including more patients are mandatory.

P-185

Aortic Coarctation Stenting Procedure Assisted by Patient-Specific Models

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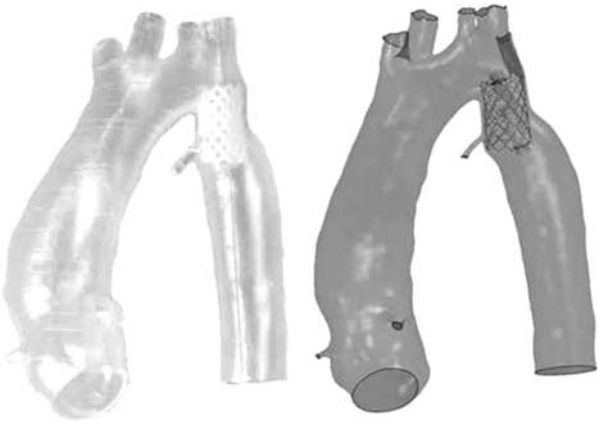
Introduction: Stent therapy is currently considered an effective alternative to surgery for treatment of aortic coarctation. However, procedural complications such as wall dissection and vessel obstruction are associated with multiple stent implantations. In this context, patient-specific models of stenting procedure might help the decision making process by realistically testing different treatments and comparing their outcomes.

In this study, we report the use of patient-specific models to optimise stent implantation in a case of aortic re-coarctation.

Methods: The patient is a 19-year-old male with native aortic coarctation and aberrant right subclavian artery (aRSCA). A bare stent was used in 2001 to treat the coarctation and subsequently redilated (14 mm) in 2007. Four years later, a follow-up catheter examination showed a 15 mmHg pressure gradient across the stent. In addition, the angiogram highlighted the presence of an early aneurysm in the proximal portion of the bare stent. A covered stent was evaluated as potential treatment, but the origin of the aRSCA in the distal portion of the bare stent precluded immediate insertion. The patient was sent for computed tomography (CT) examination. CT images were used to print a three-dimensional (3D), transparent, physical model (figure, left) of the patient's aorta and bare stent using rapid prototyping. In addition, computer analyses (figure, right) were performed to simulate different scenarios of CP stent implantation at diameters varying between 14 and 20 mm.

Results: The rapid prototyping model aided in fully understanding the patient's implantation site anatomy and the 3D relationship between bare stent, aneurysm and origin of the aRSCA. Outcomes of the computer simulations suggested a maximum expansion diameter of 18 mm in order to relieve the stenosis without obstructing the aRSCA. In agreement with the modelling results, the implantation of a 16 mm CP covered stent was successfully performed and pressure gradient reduced to 1 mmHg with no further obstruction of the origin of the aRSCA.

Conclusions: A case of re-stenting of aortic coarctation was successfully approached by integrating the results of advanced patient-specific modelling techniques into conventional decision making process. This work suggests the benefits of such a multidisciplinary approach to optimise intervention in patients with congenital heart disease.



Rapid prototyped (left) and computational model (right) of the pre-operative scenario for patient-specific simulations the CP covered stent implantation.

P-186

Complications of Catheterisation Procedures in Children With Pulmonary Hypertension: A 10 year Single institution Experience

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Introduction: Right heart catheterisation, is considered the gold standard for diagnosing and grading severity of pulmonary hypertension. We sought to describe the frequency of complications associated with catheter procedures in children with pulmonary hypertension at our institution and to identify possible predictors of serious adverse events.

Methods: A retrospective case note review of consecutive catheter procedures was performed in children with pulmonary hypertension between November 1990 and December 2010. Data were collected on diagnostic group, associated intracardiac shunts, WHO functional class and medications. Outcomes included; unplanned intensive care admission, arrhythmia requiring intervention, need for cardiopulmonary resuscitation (CPR), and death within 24 hours.

Results: 204 catheter procedures were performed in 149 children. Mean age at catheterisation was 8.4 years (range 0.1–19 years). Diagnosis was idiopathic in 57 (38%), associated with CHD in 56 (38%), lung disease in 8 (5%) and other causes in 28.

There were 51 (25%) interventional procedures (of which 41 had atrial septostomy, which was combined with Hickman line

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

insertion in 10). The remaining 153 (75%) were diagnostic catheters (of which 21 were combined with Hickman line insertion). 29 (14%) were considered urgent procedures the remaining 175 (86%) were elective.
At catheterisation patients had significantly elevated mean pulmonary artery pressure, mean 50 mmHg (SD 20); and pulmonary vascular resistance, mean 18 WU.m^2 (SD 14.2).
There were a total of 19 adverse events in 11 patients resulting in an overall risk of any adverse event of 5.4%. 5 patients experienced arrhythmia requiring intervention, 5 required CPR which returned spontaneous circulation in 2 patients. 8 patients had unplanned ITU admission.
3 patients died (overall mortality 1.47%); one following BAS, one with PVOD and loss of output on induction of anaesthesia and the third with pulmonary hypertensive crisis post procedure.
Of the 51 patients who had interventional catheters 7 (13.7%) experienced complications, compared with 4 (2.6%) complications in the 153 patients who had diagnostic catheters.
Conclusions: In experienced centers, right heart catheter procedures in patients with pulmonary hypertension are associated with low morbidity and mortality rates. Interventional catheters are likely to represent an incremental risk.

P-187
Use of Transesophageal Echocardiography in Aortic Valvuloplasty in children. Advantages?
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Background and objective: Aortic valvuloplasty has become well established palliative treatment of congenital aortic stenosis, as a high percentage of these children undergo posterior hemodynamic and/or surgical procedures.
We speculate that the use of transesophageal echocardiography (TEE) can help optimize this procedure, guiding technical aspects of the catheterization, and providing continuous hemodynamic assessment.
Methods: Retrospective analysis and comparison of aortic valvuloplasties guided by TEE versus those not guided by TEE at our institution over a 9 year period.
Results: Fifty-six primary valvuloplasties were included, of which 40 (70.2%) were performed under TEE guidance (35 with ACUNAV intravascular catheter, 3 with standard paediatric TEE probe, 1 with the microTEE probe and 1 with the 3D-TEE probe). The median age was 1.15 months (IQR 0,20–4,21). Associated congenital cardiac defects were present in 46.9% of the patients. 76.8% of the procedures were performed via femoral access, and right carotid surgical approach was used in the rest. Comparing the Fluoroscopy time in procedures performed with TEE guidance was 6,3 minutes less as compared to non TEE guided catheterisations ($p = 0.01$). The mean number of balloon dilatations per procedure was 2,05 in the TEE group, and 2,76 in the group without echo guidance ($p = 0.02$). We compared the measurement of the aortic ring by echocardiography and by angiography obtaining an intraclass correlation coefficient of 0.99. 78.6% were uncomplicated catheterisations with a lower complication rate in the TEE group: 12.8% versus 29.4%. There were no differences in the follow-up regarding aortic regurgitation, aortic valve gradient and subsequent interventions.
Conclusions: In our experience TEE guided aortic valvuloplasty provides a shorter intervention with less fluoroscopy time and less number of balloon dilatations. The use of TEE is also related to smaller complication rates. We found great correlation between echocardiography and angiography in the measurement of the aortic ring,
Otherwise we have not found differences in the immediate and mid-term follow-up results between TEE and non-TEE guided aortic valvuloplasty.

P-188
Interventional treatment of aortic arch obstruction after norwood procedure: a good alternative to surgery.
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Background: Despite significant advances in the surgical management of patients with hypoplastic left heart syndrome, aortic recoarctation after Norwood procedure continues to be a relevant problem. Catheter-based treatments, balloon aortoplasty and stent implantation, have been established as first line treatment.
Objective: To review the incidence of recoarctation after Norwood procedure and the results at short/mid term follow up after percutaneous treatment.
Methods: All patients undergoing interventional catheterization to treat aortic arch obstruction after Norwood procedure from January 2006 to December 2012 were studied. Clinical, echocardiographic and haemodynamic data were reviewed.
Results: During the study period 83 patients underwent Norwood procedure and 22 required arch interventions (26%). Patient median age was 4 months and median weight was 5 kg. All aortic recoarctations were treated in the cath lab: 12 with stent implantation (7 re-expandable) and 10 with balloon dilation. Femoral artery was the most used access (72.7%), followed by antegrade femoral vein (18.2%) and carotid artery (9.1%). Both types of procedures were considered acutely successful with an overall median gradient reduction of 25 to 5 mmHg ($p < 0.001$) and median coarctation index (coarctaton diameter/aortic diameter) reduction of 0.47 to 0.82 ($p < 0.001$). The most common site of recoarctation was distal to left subclavian (62%), but seven patients had proximal obstructions. 63% of arch obstructions developed after first stage Norwood procedure and 31.8% after second stage. 72.7% of patients were critically ill at the moment of catheterisation and 5 procedures were performed with the patient in ECMO. The median follow up period was 2.58 years (1 day–5.89 years) with a global survival rate of 59.1%. Recurrence of recoarctation appeared in seven patients (31.8%): 2 of them treated with balloon aortoplasty, 1 with stent implantation and 4 by surgery. No differences were observed between balloon aortoplasty and stent implantation in acute efficacy, need for reintervention ($p = 0.45$) or survival ($p = 0.47$).
Conclusions: In our experience balloon angioplasty and aortic stenting are effective in the treatment of recoarctation after Norwood procedure, acute and in the mid-term. Both interventional treatments have limited morbidity and mortality, and could be a good alternative to surgery.

P-189
Arrhythmia outcome in patients who are over 40 years of age after device closure of secundum atrial septal defect. A medium-term study
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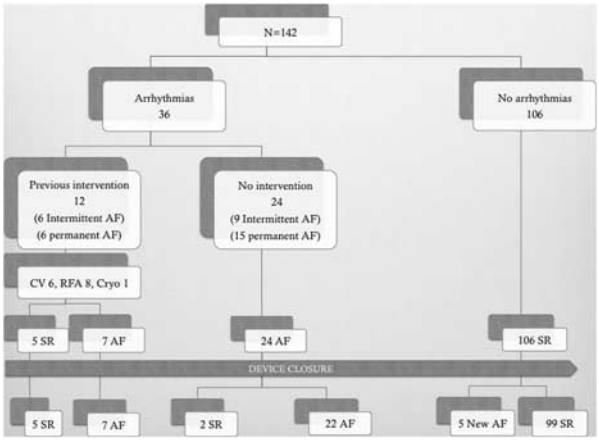
Introduction: It is recognized that surgical closure of secundum atrial septal defects (ASD) will improve symptoms but may have

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

little positive effect on arrhythmia control. Many patients now have device closure as an alternative to surgery and the purpose of this study was to investigate arrhythmia status in a consecutive group of older adults after device closure.
Method: Retrospective of 142 patients (>40 yrs old at time of device implantation) from a single centre (mean follow-up 2.8 yr, range 1–5 yrs). Patients were stratified to three groups based on arrhythmia status before the closure procedure: A (n = 24), preexisting atrial arrhythmia (intermittent/persistent) with no invasive electrophysiology (EP) treatment; B (n = 12), documented atrial arrhythmia (intermittent/permanent) but who had at least one therapeutic EP procedure and C (n = 106), patients without arrhythmias. All patients had a clinical history, ECG and 14 patients had a 24-hour ECG carried out (mean follow-up 3.2 yrs).
Results: The median age at time of ASD closure was 66.8 yrs (group A), 56.9 yrs (group B) and 51.8 yrs (group C). There was a significant difference between A and C. The mean size (mm) of device were 26.5, 22.1 and 24.4, and the mean PA pressure (mmHg) were 29, 24.8 and 20.1 in group A, B and C respectively. Arrhythmia persisted in 92% of Group A but 40% of Group B were in sinus rhythm and without anti-arrhythmic medication after a mean follow-up of two years. Five percent of those with no previous arrhythmia developed clinical atrial arrhythmia (mean age 51.8 yrs at onset of arrhythmias) requiring treatment and 3% remained in atrial fibrillation. The mean time to onset of arrhythmia was 2.5 months following device closure.
Conclusions: ASD device closure alone has little impact on arrhythmia outcome in those with documented atrial arrhythmias who are over 40. EP Intervention prior to ASD closure appear to play a role as about 40% of that group are on no antiarrhythmic medication on medium term follow up. The management of such patients remains a challenge but it appears that a proactive approach with regards to ablation procedures deserves prospective study. The fact that 5% of patients with no previous history develop arrhythmia also requires further investigation.



P-190
Folded Melody Valve Technique for percutaneous valvulation of Complex Right Ventricular Outflow Tracts
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Introduction: Percutaneous Melody valve implantation has achieved standard of care for the management of patients with dysfunctional right ventricle (RV) to pulmonary artery (PA) conduits. The landing zone right ventricular outflow tract (RVOT) dimensions may vary significantly making it unfavorable for Melody valve implantation. We report a novel technique in patients with complex, repaired RVOT anatomies.
Methods: All patients were candidates for Melody insertion were evaluated with angiography. The landing zone was carefully studied. Patients with short landing zone or landing zone consisting of a retrosternal part were valved using a special technique. The insertion of the Melody valve in retrosternal continuity is at risk of stent fracture. In patients with short landing zone, there was a risk of Melody valve impingement with the RV muscle at the proximal end and the risk of accidental covering of PA distally. To circumvent these potential complications, we modified the Melody valve before implantation. The terminal open stent struts of the Melody valve were folded over itself from inside out to reduce its effective length.
Results: From 2008 to 2012, four patients (out of 4) received a shortened, folded Melody. The uncovering, crimping and loading of the Melody valve was technically simple in all. The folded and crimped valve moved easily in the delivery system and the deployment was done using standard technique. All valves were positioned where intended. Patients were discharged the day after the procedure. Evaluation showed excellent performance of the Melody valve with no paraprosthetic leak, no erosion, no perforation, no stent fracture, no residual stenosis, no valvular or para valvular regurgitation immediately after implantation or at follow up.
Conclusion: The "Folded valve technique" is a safe addition to the interventional armamentarium allowing the implanting physician to modify the valve in patients with complex short RVOTs. By implanting the Melody valve far from the sternum, this technique may reduce the overall incidence of stent fractures reported with this device. In the future, this technique may also be a good option for patients with vulnerable RVOT neighborhood that may preclude conventional technique.

P-191
Use of covered stents for closure of fenestration in extracardiac cavopulmonary connection: technical aspects and mid-term results
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Objective: Closure of fenestration in total cavopulmonary connection (TCPC) is usually performed with devices used to close intracardiac or vascular connections. This study presents our data regarding the use of covered stents in that context.
Methods: We retrospectively reviewed data of all the patients receiving a covered stent to close a fenestration of TCPC between 2005 and 2012.
Results: 50 patients met the inclusion criteria. Median age and weight were respectively 7.7 years and 20 kg. Median interval between Fontan completion and fenestration closure was 13 months. Procedures were done through the femoral vein in 42 and jugular vein in 8 patients. 57 stents were used. 7 patients received 2 stents (CP group) because of incomplete sealing of the fenestration. Covered stents were CP stents in 42 patients and Atrium Advanta

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

V12 in 8. For stents insertion we used BIB balloons in 24 patients or simple balloons (TYSHAK or Balt) in 18 patients. In 8 cases 2 different balloons were used: 5 in patients receiving 2 stents, 3 in patients receiving an Atrium stent needing post dilatation. 5 patients had simultaneous occlusion of venous collaterals. Median procedural and fluoroscopy times were respectively 49 ± 29 and 8 ± 7 minutes. Mean central venous pressure rose from 10 to 12 mmHg. Mean oxygen saturation increased from 88% to 96%. Full occlusion was confirmed in 47 patients. The remaining had significant residual shunts: two in patients with intracardiac Fontan and one in a patient where stents could not be fully opened. Following the procedure, 5 patients had local bleeding with one needing blood transfusion, and three delayed discharge at day two following the procedure. There was no thrombo-embolic event reported after a mean follow-up of 49 months.
Conclusion: Covered stent is a good option to close fenestration in extracardiac TCPC. It is safe, easily achievable with low fluoroscopy time, very low risk of thrombo-embolic events or failure. The good results are sustainable when excluding patients with none circular pathway.

P-192
Patent Ductus Arteriosus Stenting (Transcatheter Potts Shunt) for Palliation of Suprasystemic Pulmonary Arterial Hypertension: A Case Series
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Introduction: Side to side surgical Potts shunt connecting descending thoracic aorta to the left pulmonary artery have been tried in the past for patients with symptomatic severe and refractory idiopathic pulmonary arterial hypertension with right ventricular failure and syncope. The promising results from this surgical technique inspired us to try interventional methods in opening and stenting residual patent ductus arteriosus (PDA) for effective post tricuspid decompression of the pulmonary circuit in patients with chronic suprasystemic PAH who failed multiple medical therapies.
Methods: We look in our database for patients with suprasystemic PAH who underwent PDA stenting to decrease PA pressure.
Results: Three patients were found. All three had successful PDA stenting allowing for equalization of aortic and pulmonary systolic pressure. We chose stent diameter based on our experience of surgical Potts shunt. The criteria we used for selecting initial stent diameter were a compromise between age, "surgical" target diameter and narrowest PDA diameter. Moreover, we chose to gradually enlarge the diameter of the connection and thus flow by sequential balloon inflation with the aim of equalizing the PA and aortic systolic pressures, maintaining pulmonary blood flow and limiting the lowering the desaturation of the inferior limbs. A 6-mm stent was used in the smallest patient and 9-mm stent for the oldest patient. Post dilatation and over-expansion was necessary in one patient. The objectives were obtained with the initial stent in the other two. The saturation in lower limbs decreased as a result. All patients had improvement of their clinical status following the procedure.
Conclusions: The promising results from these cases compelled us to change the approach and protocol for management of patients with suprasystemic PAH. We now do a detailed cardiac catheterization in all patients with suprasystemic PAH to look for a PDA. Even tiny it can be opened by stent insertion to equalize aortic and pulmonary pressure and improve RV function.

P-193
Comparison of immediate results of percutaneous balloon valvuloplasty performed from two different arterial accesses for aortic valve stenosis in neonates
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Introduction: percutaneous balloon valvuloplasty (BVP) for aortic valve stenosis in neonates is the treatment of choice in many centres. Mainly two arterial accesses are utilized: femoral artery and carotid artery. Separate results are well known, but there is lack of direct comparison. We compare immediate results of percutaneous BVP in infants depending on arterial access used.
Methods: In this retrospective study, we collect data regarding 64 neonates with critical or severe congenital aortic valve stenosis from last 12 years. 29 consecutive procedures were performed through surgically exposed carotid artery (CA group). 35 patients undergo BVP through puncture of femoral artery (FA group). All procedures were performed in the first 28 days of life. We compare relief of aortic stenosis, degree of aortic regurgitation (AR), procedural time and vascular complications.
Results: All procedures in FA group and all but one in CA group were performed. In one patient from CA group we were not able to cross severely stenotic aortic artery. Median age in CA group was 5 days (range 1–28 days) and in FA group 8 days (1–27 days). The median systolic pressure was 67 mmHg (5–122 mmHg) in CA group and 58 mmHg (5–110 mmHg) in FA group. Post procedure median systolic pressure was 12 mmHg (0–44 mmHg) in CA group and 14 mmHg (2–50 mmHg) in FA group. Immediately after the procedure there was one patient with severe AR (FA group), moderate AR was present in 2 patients in CA group, and in 6 in FA group, mild AR in 12 patients in CA group, and 16 in FA group. Trivial or no AR was find in 15 patients from CA group and 12 patients from FA group. Procedural time was longer in CA group 93 ± 31 minutes compared to 76 ± 25 minutes in FA group. There was no immediate vascular complications in CA group, in 3 patients from FA group critically impaired circulation to the leg occurred during procedure.
Conclusions: BVP performed both from carotid artery and femoral access is effective and safe. There is higher incidence of severe and moderate AR in FA group. More studies are needed to access long time safety and efficacy.

P-194
Therapeutic strategy for the treatment of Swiss Cheese VSDs
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Introduction: Surgical repair of Swiss Cheese VSDs is still challenging. We describe interventional or combined surgical and interventional strategies for the treatment these lesions.
Patients and methods: During the last 4.5 years 10 patients with Swiss Cheese VSDs were presented. 6/10 had additional complex

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

malformations. They were treated either by a combination of surgery and intervention (9/10) or by intervention exclusively (1/10). Altogether 16 surgical procedures were performed to address the VSDs or the Banding of the pulmonary artery, two hybrid procedures inclusively. Nine patients underwent pulmonary artery banding, in 6/9 a partial occlusion of the VSDs were performed and in three patients additional mayor surgery was necessary. The age at first surgery ranged from 2 weeks to 15 months, mean 3.5 months. The weight ranged between 2.6 and 6 Kg mean 3.8. In 5 patients a second operation was performed for Debanding and closure of additional VSDs, unsuitable for interventional closure (2/4). Two patients underwent 3 surgical interventions.
All patients underwent interventional VSD closure. During 14 procedures 16 devices were implanted, 4 muscular VSD Plugs, 3 vascular Plug IV, 8 PDA Plug II and 1 vascular Plug from SJM company (former AGA). The age at the time of intervention ranged between 2 months and 42 months, mean 16.7 months, and the bodyweight was measured between 4 and 12.5 Kg, mean 7.6 Kg. Additional interventions were performed in 4 patients.
Results: 7/10 patients showed complete closure or no relevant residual shunt during the follow up time after the first interventional procedure of 15–53 months, mean 31 months. One child needs further interventional treatment, one another operation. One child had a hybrid procedure in the cathlab under ECMO support died two weeks after the procedure on a septicaemia.
Conclusion: For the treatment of Swiss Cheese VSDs a combination of surgical and interventional procedures shows encouraging results. Particularly patients with additional complex malformations can profit from the cooperation between cardiologists and surgeons.

P-195
Percutaneous closure of atrial septal defects in children under deep conscious sedation and spontaneous breathing - a feasible and safe concept

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Background: Interventional cardiac catherization (CC) is usually performed under general anaesthesia and endotracheal intubation. However, as CC in children under deep sedation and spontaneous breathing has been proved to be a feasible concept, percutaneous closure of atrial septum defect (ASD) without general anaesthesia is nowadays attempted in a growing number of children
Objectives: To objectively evaluate the success and complication rate of percutaneous ASD closure in children under deep conscious sedation and spontaneous breathing.
Methods: Retrospective analysis of all consecutive children undergoing percutaneous ASD closure at a tertiary care paediatric cardiology centre. Study patients received premedication with intravenous (i.v.) midazolam, followed by i.v. ketamine for sedation induction and propofol continuous infusion for the procedure as standard medication.
Results: Median age of study patients 6.1 years (minimum 0.5; maximum 18.8), and median body weight was 21.6 kg (5.3; 92). Median cumulative midazolam dose was 0.08 (0.02; 0.18) mg/ kg body weight, median ketamine dose during CC was 2.7 mg/kg (minimum 0.3; maximum 7), and median propofol infusion continuous infusion rate was 5 mg/kg/h (1.1; 10.7). After transoesophageal echocardiography and balloon sizing of the defect, percutaneous ASD closure was attempted in 174 patients and device implantation performed successfully in 163 patients (94%). There were no major cardiorespiratory complications associated with deep sedation, and only two patients (1%) required endotracheal intubation during CC due to bronchial obstruction. Seventeen patients (8%) had minor respiratory complications and required frequent oral suctioning or temporary mask ventilation. Median length of inpatient stay after CC was 2 days (1; 32).
Conclusions: Percutaneous ASD closure can be performed safely and with high success rates under deep conscious sedation and spontaneous ventilation in children.

P-196
Systemic-to-pulmonary venous collateral vessels after cavo-pulmonary connection

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Introduction: Venous collaterals connecting superior and inferior caval veins systems, as well as systemic to pulmonary venous collateral vessels (SPVCV) are commonly found in patients with partial or total cavo-pulmonary connection. SPVCV seem to be especially interesting from anatomical, physiological and therapeutic point of view. Their development is related to normally existing anastomoses of bronchial and mediastinal veins draining to the left atrium and pulmonary veins. The aim of the study was to describe anatomy and clinical importance of SPVCV in patients after cavo-pulmonary connection.
Methods: Cardiac catheterization results of 100 patients examined at our institution were reviewed. The group included 99 bi-directional Glenn patients and 1 patient after Kawashima procedure. Fontan procedure was completed in 12 of 99 Glenn patients. All existing examinations of patients were reviewed and the presence of SPVCV was assessed.
Results: In 17 of 100 patients SPVCV were visualized on angiography. SPVCV were found in 13 of 99 Glenn patients and in Kawashima patient. In one patient another SPVCV was discovered after interventional closure of previously existing collateral vein. Among Fontan patients, in 1 case the SPVCV existed after Glenn procedure and in 3 cases it appeared after Fontan circulation have been achieved. There were 19 supplying veins visualized. They originated from left brachiocephalic vein (12), azygos vein system (5), right superior phrenic vein (1) and right venous angle (1). Most of them (14) drained into a single vessel or chamber: to the left atrium (4), left superior pulmonary vein (4), right superior pulmonary vein (3), right inferior pulmonary vein (2), left inferior pulmonary vein (1). Five remaining veins emptied to both right pulmonary veins (2), both left pulmonary veins (1), left atrium and right inferior pulmonary vein (1), left atrium and left superior pulmonary vein (1). In most of cases they produced limited right-to-left shunt. Four patients were accepted for intervention, which was completed in 1 case.
Conclusions: SPVCV develop in almost one fifth of cavo-pulmonary anastomosis patients. They drain through bronchial and mediastinal veins to pulmonary veins and/or left atrium. Most of them are small vessels with limited clinical significance.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

P-197

Interventional closure of secundum type atrial septal defect in children with a bodyweight less than 10 kg: Indications, feasibility and outcome

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Background/Objectives: Interventional device closure of secundum type atrial septal defect (ASD) in children can be performed safely at the age of 2–4years. Aim of our study was to evaluate indications, feasibility, periprocedural complications, and outcome of interventional ASD closures in very small children with a bodyweight less than 10 kg.
Methods: Retrospective analysis (2005–2012) of ASD closures in children less than 10 kg bodyweight.
Results: In 25 children (13 male) 24 successful interventional ASD device closures were performed at a median age of 1.2 years (0.2–2.8) and a bodyweight of median 7.5 kg (4.6–9.9). RV volume overload was present on echocardiography in 24 patients, resulting in a Qp/Qs of mean 1.7 (±1). Clinically 9 (36%) pts presented with failure to thrive. 8 (32%) were past preterm infants with bronchopulmonary dysplasia (BPD). In 12 pts a genetic syndrome (Trisomy 21 in 9pts, 36%) was present. In more than 2/3 of the pts (n = 18, 72%) pulmonary hypertension (PAH) was suspected on echocardiography. Haemodynamic evaluation revealed baseline PVR of mean 3.8 WU/m^2 (±6.7) and mean PAP 25 mmHg (±10). ASD size was mean 8.1 mm (±2.7) with a ratio of bodyweight/defect-size of median 0.87. Mean femoral sheath size used was 7Fr (±1.5) and ASD closure devices of 10 mm (±3 mm), procedure time 84 min(±34), fluoroscopy time 13 min (±7). There were no periprocedural complications, postinterventionally prolonged femoral bleeding (n = 1) and femoral venous thrombosis (n = 1) occurred.
Median follow-up was 0.5 years (0.1–6) showing complete closure of ASD with no residual shunt in all 24 pts. All pts showed regression of PAH measured by echocardiography, one pt still being treated with sildenafil.
Conclusions: Interventional ASD closure in children weighing less than 10 kg can be performed without any additional risks and shows a favorable outcome with a slight device oversizing in defect sizes up to 8 mm. Defect sizes in these small children, which comply with the bodyweight of the patient in kg, mostly can be addressed successfully with a percutaneous device closure. Indications for early closure are co-morbidities as BPD, failure to thrive or suspected PAH on echocardiography. As these pts do benefit significantly from the procedure, there are no restrictions to withhold timely procedures from this specific group.

P-198

Inhaled Iloprost Treatment of Pulmonary Arterial Hypertension Before And After The Surgical Repair of Ventricular Septal Defect in Children

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Introduction: Pulmonary arterial hypertension (PAH) is an important factor for morbidity and mortality in children with ventricular septal defect. We evaluated the use of iloprost in children undergoing congenital heart surgery.
Methods: Fifteen children with VSD, 1 with VSD-ASD, 1 with PDA were included in this study. Children aged between 1–17 years (median, 4 years) with long-standing and severe PH were studied.
Various hemodynamic parameters were measured before and after iloprost inhalation, and vascular resistance was determined. Responders to the iloprost test were defined as those with a decrease in both pulmonary vascular resistance (PVR) and pulmonary-to-systemic vascular resistance ratio (Rp/Rs) of > % 10. Before surgery all patients were treated by aerosolised iloprost for two weeks. After hospitalization all patients who were switched to IV iloprost.
First routine detailed echocardiography was performed at first month and two year.
Results: Median age at surgery was 4 (1–17) years, 9 patients (%53) were male. At the catheterization laboratory, the baseline median mean PAP (MPAP), PVR, PVR/SVR was respectively 63 (45–72) mmHg, 8,3 (6.2–11,2) WU, 0.41 (0,35–0,7). After iloprost vasoreactivity testing median MPAP, PVR and PVR/SVR ratio significantly decreased, respectively 52 (41–71) mmHg, 6.4 (5.8–10.4) WU, 0,28 (0.18–0.48) ($p < 0.001$). Pulmonary artery systolic pressure fell below 75% of systemic arterial pressure in all patients after surgery. All the patients were administered inhaled iloprost after catheterization until the surgery was performed. Prior to surgery the median SPAP was 71 (59–89) mmHg. Postoperative immediately after SPAP was detected as median 47 (35–58) mmhg.
Three patients suffered PHC in postoperativetive 3,4, 30th days. Two patients died.
Fifteen patients recovered well and discharged. Median follow up time was 17 (6–42) months.
Discussion: In our study; aerosolized iloprost therapy significantly reduced the PAP in patients who suffering from PH before surgery of congenital heart defects. A significant reduction in PAP after cardiac surgery was observed in patients with positive response to inhaled iloprost especially PVR/SVR<0.5. The prognosis for patients undergoing closure of large VSD with increased PVR is dependent on the age, degree of PVR and PVR/SVR.

P-199

Congenital Supravalvular Aortic Stenosis: Twenty-five years' experience at the Deutsches Herzzentrum Berlin

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Objective: Congenital supravalvular aortic stenosis (SVAS) is an uncommon obstructive arteriopathy of varying severity with frequent association with stenoses of systemic and pulmonary arteries. Several techniques for symmetric reconstruction of the aortic root in SVAS have been developed, but it remains unclear what is the optimal surgical procedure. We reviewed our experience with surgical management of SVAS.
Methods and results: Forty patients underwent operations to treat congenital SVAS at our institution between 1987 and 2012. Twenty-six patients were male (65%) and nine under 1 year old (22%). Twenty-five patients had associated Williams-Beuren syndrome (62%) and 49% had pulmonary artery involvement. The mean preoperative gradient pressure was 79.85 ± 27 mmHg. Surgical procedures included patch enlargement of the noncoronary sinus only (McGoon's technique) (n = 4), inverted bifurcated patch plasty (Doty's technique) (n = 24), triple-sinus

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

reconstruction of the aortic root (Brom's technique) (n = 6) and sliding aortoplasty (Myers-Waldhausen's technique) (n = 6). Nine patients had associated procedures (22.5%). The mean post-operative pressure gradient was 16 ± 7.1 mmHg. There was one early death. Among those who survived the early postoperative period, 95% were alive at 5 years, 86% were alive at 10 years, and 72% were alive at 20 years. According to time-related analysis there was no difference in terms of survival and reoperation between different surgical techniques (p = NS). Patients under 1 year old have worse prognosis for survival, freedom from reoperations and re-interventions ($p < 0.05$).
Conclusions: In our cohort, the most important prognostic factor was the age at the first operation. Surgical treatment is palliative and requires careful follow-up examinations. Good surgical outcome can be achieved with the appropriate method of treatment in patients with both localized and diffuse SVAS, but because each case is different, the surgeon should individualize the approach based on the anatomic findings.

P-200
Surgery for pediatric active infective endocarditis: 24-year single center experience
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Objectives: Active infective endocarditis (AIE) occurs less commonly in children. We retrospectively analyzed the clinical and microbiological status and the results of surgical treatment over a period of 24 years.
Methods: Between April 1988 and December 2012, 8706 pediatric patients (<18 years) were operated upon. Of these, 31 (0.3%) patients (n = 21 men, median age 14 years, 7 mo - 17 years) underwent following surgery: aortic valve replacement (AVR) (n = 5), homograft aortic root replacement (ARR) (n = 6), Ross operation (n = 3), mitral valve (MV) repair (n = 7), MV replacement (n = 2) and combined tricuspid valve (TV) surgery (n = 8) in 20 (62%) cases of native and 13 (38%) of prosthetic AIE. Underlying congenital heart disease (CHD) was present in 62%. Follow-up (0–24 years) was completed in 91%.
Results: Preoperatively cerebral emboli were seen in 6 (19%), renal insufficiency in 4 (12%), aortic root abscess formation in 6 (19%) and sepsis in 4 (12%) patients. There were no operative deaths; 30-day mortality was 12.5% with 2 patients suffering from myocardial failure, 1 from septic multiorgan failure (MOF) and 1 from hemorrhagic shock after ECMO implantation. One-year survival was 87.5%. Actuarial freedom from reoperation and actuarial survival after MV repair at 1 and 10 years were 100%. Early endocarditic re-infection occurred in 1 patient after ARR. In the long term 1 patient underwent reoperation due to homograft degeneration. Staphylococci species (31%) were the most frequent microorganism.
Conclusions: AIE occurs among young children with complex CHD more frequently.
Repair of atrioventricular endocarditis yields excellent results in children and should be considered as the primary surgical option in these patients. Homograft ARR and Ross operation are associated with low operative mortality and provide satisfactory early and long-term survival and favorable freedom from recurrent endocarditis and repeat operation.

P-201
LeCompte Maneuver for Airway Compression Management in Late-presenting Absent Pulmonary Valve Syndrome
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Introduction: Patients with absent pulmonary valve syndrome (APV) often present early with airway compression, from diffuse aneurysmal dilatation of the pulmonary artery branches. Repair usually includes pulmonary artery reduction plasty to relieve proximal obstruction of the mainstem bronchi. The LeCompte maneuver has been proposed to address this issue, although there is limited data available. This study reviews our recent experience in managing APV syndrome in later presenting children, and surgical techniques used for managing airway compression.
Methods: This study is a retrospective chart review of all patients who underwent repair of tetralogy of Fallow and APV from 2000 to 2012 at our institution. Patients with clinical evidence of airway compression undergo systematic pre- and post-operative bronchoscopy. The primary endpoints were post-operative bronchoscopic and clinical evidence of persistent airway compression, and need for reinterventions or reoperations on the pulmonary arteries.
Results: 19 patients were included during the study period. The mean age at repair was 4.1 ± 3.0 years (range, 10 months – 11 years). 6 patients had associated anomalies: 3 with discontinuous left pulmonary artery from major aorto-pulmonary collaterals (MAPCA), 1 with a MAPCA to the LPA, 1 with a right aortic arch, totally anomalous pulmonary venous return and infradiaphragmatic MAPCA to the right lung, and 1 patient with 22q11 microdeletion. There were no perioperative deaths. 8 patients with respiratory symptoms had preoperative bronchoscopy, which showed airway compression in 7 patients and managed by pulmonary artery reduction plasty in 4 patients, and LeCompte maneuver in 3 patients. In the first group, 2 patients had no postoperative airway compression, 1 patient had improved compression, and 1 patient had unchanged compression. In patients managed with a LeCompte maneuver, 2 patients had no or trivial airway compression and 1 had improved compression (P = 1.0). There were 6 late reinterventions or reoperations on the RV-PA conduit (2/4 in the PA plasty group, 1/3 in the LeCompte group, P = 1.0).
Conclusions: In patients with APV and airway compression, either pulmonary artery reduction plasty or the LeCompte maneuver can relieve proximal airway compression, without a significantly different risk of pulmonary artery reintervention between techniques.

P-202
Acute systemic-pulmonary shunt occlusion in cyanotic congenital heart diseases: Are hereditary thrombophilic factors really matter?
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Objective: Acute thrombosis of systemic to pulmonary shunts (SPS) in cyanotic congenital heart disease is a crucial complication for patients have shunt dependent pulmonary blood flow.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Our aims were to determine the outcomes in patients early after undergoing SPS operations and to assess the presence of inherited thrombophilia factors in the children experienced occlusion of SPS.
Material and methods: A prospective randomized clinical trial was performed in a tertiary referral congenital cardiac center. Patients with hypoplastic left heart syndrome were excluded. The patients experienced shunt thrombosis were assessed for inherited and acquired thrombophilic factors. Vitamin B12 and folic acid levels and antithrombin III, protein C (PC), protein S (PS), anti-cardiolipin antibody (ACA), homocysteine, factor 5 Leiden and prothrombin G20210A mutations were studied.
Results: From October 2010 to September 2012 seventy-seven children underwent first shunt operation with a median age of 61 days (range: 4 days–5,6 years) and median weight of 8,8 kg (range: 2.1–14 kg). Thirty-three (%43) of them were neonate. The shunt sizes varied from 3 to 5 mm.The rate of acute shunt blockage was 10% (8/77), all within the first 24 h. There were 24 hospital deaths (31%), three of them was associated with shunt occlusion. There were 3 hereditary thrombophilia (1 antiphospholipid syndrome, 1 protein C deficiency, 1 active protein C resistance) in patients who had shunt thrombosis.
Conclusion: SPS occlusion can be potentially life-threatening in the case of patients have shunt dependent pulmonary blood flow. When shunt occlusion occured especially in patients with recurrent shunt thrombosis, inherited thrombophilia factors should be investigated.

P-203
The use of dornase alpha for postoperative pulmonary atelectasis after congenital heart surgery
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Objective: To investigate the efficacy of dornase alpha (DA), a mucolytic agent, in children who developed pulmonary atelectasis following congenital heart surgery.
Method: Design: Retrospective observational study.
Setting: Pediatric cardiac intensive care unit at a tertiary care education hospital.
Patients: Between July 2011 and July 2012, 41 patients who underwent cardiac operations due to congenital heart disease and developed postoperative pulmonary atelectasis after congenital heart surgery that was resistant to conventional treatments and chest physiotherapy.
Interventions: Twenty-six patients received DA treatment. As a control group, 15 patients who were treated with conventional medications were chosen. Study groups were matched for age and diagnosis.
Results: The median age of patients in the study and control groups was 25.5 (3–480) days and 50.0 (3–480) days, respectively. While 15 (57.6%) of 26 patients from the study group were male, 8 (53.3%) of 15 patients from the control group were male. The median weight was 4.2 (2.9–14.2) kg in the study group and 4.0 (3.5–13.6) kg in the control group. In the study group, pulmonary atelectasis was diagnosed at a median period of 5 (2–18) days after operations, whereas in the control group atelectasis was diagnosed at a median period of 6 (3–19) days after operations. In the study group, the median atelectasis score decreased from 3.4 (1–6) to 0.8 (0–3) ($p = 0.001$). The median pO2 level increased from 69 (17–142) mmHg to 89 (30–168) mmHg ($p = 0.04$). Besides, heart rate and respiratory rate per minute were significantly decreased ($p < 0.05$). There was no significant change in the control group.
Conclusions: The use of DA can be effective and safe for the management of pulmonary atelectasis that develops following congenital heart surgery.

P-204
Analysis of RV components after reoperation of the right ventricular outflow tract in patients with Tetralogy of Fallot
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Introduction: In patients with repaired Tetralogy of Fallot (TOF), reoperation of the right ventricular outflow tract (RVOT) is often necessary. This study aims to analyse the functional recovery of the right ventricle (RV) and the RVOT after a reoperation using volumetrical data from cine MRI.
Methods: 22 patients (25.0 ± 9.2 yrs) underwent an MRI examination before and after (9.6 ± 5.6 months) pulmonary valve replacement or reconstruction (PVR). For segmental analysis of the RV, a custom made analysis software was developed. The method provides an automatic contour detection algorithm for the determination of the RV blood volume and enables the user to segregate the blood volume of the RVOT based on anatomic landmarks and to generate volume-time-curves over a cardiac cycle. The analysis of volume-time-curves of the RV allowed calculation of Peak-Filling-Rate (PFR) and Time-to-Peak-Filling-Rate (TPFR), which are surrogate parameters for diastolic dysfunction.
Results: A significant reduction in blood volumes of the RV as well as the RVOT was present after a PVR (RV-ESV -19.0 ± 27.2 ml/m^2, $p = 0.004$, RVOT-ESV -5.5 ± 6.1 ml/m^2, $p < 0.001$, RV-EDV -36.5 ± 27.9 ml/m^2, $p < 0.001$, RVOT-EDV -7.3 ± 7.9 ml/m^2, $p < 0.001$). Pulmonary insufficiency was also significantly reduced ($-24.3 \pm 15.8\%$, $p < 0.001$). The PFR decreased (-149.2 ± 261.8 ml/s, $p = 0.001$) and the TPFR increased (0.06 ± 0.12s, $p = 0.041$), indicating improvement of RV diastolic function. Additionally, late gadolinium imaging showed that non-viable tissue was present in $5.7 \pm 3.8\%$ of the RV mass and in $11.9 \pm 7.2\%$ of the RVOT mass before operation and did not change on follow-up.
Conclusion: After reoperation of the RVOT in TOF patients the systolic and diastolic function of the right ventricular components improved within less than a year. The end-diastolic and end-systolic blood volumes decreased significantly in the RVOT and the total RV. The dedicated analysis software enables the user to assess the components of the RV in a time-efficient and precise manner.

P-205
Interleukin-6 as a marker of inflammatory related post-operative myocardial dysfunction
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Background: Cardiac operations with cardiopulmonary bypass in children are related to the systemic release of inflammatory mediators. Among them, interleukin(IL)-6, plays a central role as this cytokine possesses pro-inflammatory properties, causing in the early course of the systemic inflammatory reaction myocardial damage and as it controls acute phase response.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

This study was designed to test the hypothesis that IL-6 after cardiac operation is a marker of organ dysfunction and correlated with postoperative outcome.
Methods: In 58 patients (median age: 4 9/12 years), serum levels of IL-6 were measured immediately after end of cardiopulmonary bypass, 4- and 24 hours po and were correlated with po outcome. Cardiac outcome was quantified by using a score system at the end and 4 hours po that takes into account duration of surgery, po hemodynamics and need for inotropic- and pacing support.
Results: IL-6 levels were maximal at the end of CPB (202,4 ± 47,3 pg/ml, mean ± SEM), decreasing in the po period to reach 142 ± 42pg/ml and 51,2 ± 17,3 pg/ml 4- and 24 hours po, respectively.
IL-6 levels at the end of CPB correlated with duration of CPB ($p < 0.001$) and myocardial ischemia time ($p < 0.05$). It also correlated negatively with ultrafiltrated volume at the end of the operation ($p < 0.005$), with po blood pressure and with oxygenation index ($p < 0.05$, respectively).
Patients with a cardiac score > 90th Percentile had IL-6 concentrations after CPB > median value for the whole group. A cut off value could, however, not be defined.
Conclusions: IL-6 levels measured at the end of CPB correlated with postoperative morbidity, especially with impairment of cardiovascular function.
Although a clear cut off value could not be defined in this series, IL-6 levels measured immediately after cardiac surgery might allow identifying individuals at risk to develop postoperative organ dysfunction. These individuals would profit from early anti-inflammatory strategies in the post-operative course.

P-206
Short lasting anti-inflammatory reaction in neonates undergoing cardiac operations correlates with postoperative outcome
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Background: Cardiac surgery with cardiopulmonary bypass (CPB) in neonates is related to a systemic inflammatory reaction that jeopardises postoperative outcome. In this respect, the importance of the production of anti-inflammatory mediators such as interleukin (IL)-10 and cortisol, that might counterbalance systemic inflammation might be protective and improve postoperative outcome.
Methods: Serum levels of IL-10 and cortisol were measured during and postoperatively up to 10 days after cardiac surgery in 13 neonates (mean age: 7 days) operated on for transposition of the grat arteries ($n = 7$), total anomalous pulmonary venous return ($n = 3$), truncus arteriosus communis ($n = 1$) and interrupted aortic arch ($n = 2$).
Cardiopulmonary bypass protocol included use of moderate hypothermia, cardiocirculatory arrest if needed and preoperative administration of dexamethasone.
Results: In all patients, IL-10 levels increased from a preoperative value of 6,45 ± 1,14 to reach maximal values of 385 ± 65,3 pg/ml at the end of CPB. IL-10 levels decreased in the first postoperative days to reach preoperative values 48 hours postoperatively. Maximal IL-10 levels correlated with minimal core temperature during surgery (Spearmann: 0,63, $p < 0.05$). They also correlated with postoperative lactate – and creatinine levels and with the postoperative cardiac scoring system ($p < 0.05$, respectively).
Cortisol levels increased from preoperative values of 1188 ± 80 pg/ml up to 1281 ± 275 pg/ml at the end of cardiopulmonary bypass (not significant). Cortisol levels decreased thereafter to reach a minimal value 72 hrs after surgery (765 ± 191 pg/ml, $p < 0.05$ versus preoperative) and normalised thereafter. Cortisol levels measured after CPB correlated with CPB duration, with po lactate levels, creatinine concentrations and systolic blood pressure ($p < 0.05$, respectively). Po cortisol- and IL-10 levels were correlated to each others.
Conclusions: Our data show that neonates answer the inflammatory stress due to cardiac surgery with a significant production of IL-10 that is, however, short lasting. In contrast, the production of cortisol is significantly decreased postoperatively, suggesting insufficient stress response after cardiac surgery in neonates.
The correlations shown between IL-10 and cortisol, and markers of postoperative organ dysfunction, respectively, indicate that both anti-inflammatory mediators are up-regulated due to operative stress and might therefore be targets for pharmacologic modulation.

P-207
Paediatric high fidelity simulation is useful to improve resuscitation skills in paediatric cardiology
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Introduction: Life-threatening Paediatric Cardiac Arrhythmias are a low frequency event in paediatric resuscitation.
This can cause a high stress level on teamwork, which influences the resuscitation efficiency.
Patients have the best chance to survive when the team rapidly identifies the problem and performs life saving interventions. To achieve a good clinical outcome, the clinical skills as well as the non-clinical skills such as communication, leadership and teamwork have to be optimal.
Our hypothesis was that offering good education and team training, including high fidelity simulation on a regular basis would improve these skills.
Methods: A combined theoretical and practical course was put in place for a multidisciplinary group at our University Hospital. These groups were put together with nurses and Specialist registrars, who are working together in clinical practice.
The course dealt with the treatment of cardiac arrhythmias like ventricular fibrillation, ventricular tachycardia, supraventricular tachycardia and asystole.
After the theoretical course the group went to the simulation laboratory including a paediatric simulation mannequin, in whom they needed to handle cardiac arrhythmias scenario. During the debriefing clinical as well as the non-clinical skills were debriefed in detail. The main focus was put on the algorithm, including rapid diagnosis of arrhythmias, first choice medical treatment if needed, minimal interruption of cardiac massage, communication and leadership.
Results: After the first course several further simulation sessions were held. An improvement in dealing with critical paediatric resuscitation situations was observed, as quantified by a score system assessing time to obtain correct diagnosis, time to give the correct instructions with respect to stabilisation and treatment, quality of leadership and quality of debriefing.
A questionnaire was distributed after the session in which the candidates mentioned that regular training sessions were useful to maintain their resuscitation skills, give them more confidence in acute situations in general and in treating arrhythmias in particular and good teamwork.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Conclusion: High Fidelity Simulation is a necessary tool to prepare a multidisciplinary team for rare but serious situations in paediatric cardiologic resuscitation. This is expected to result in an improvement of patient safety and decreased stress of the team facing these acute situations.

P-208

Non-confluent pulmonary arteries- therapeutic approach and results

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Introduction: Non-confluent pulmonary arteries (PA) may occur in a variety of congenital heart diseases preventing uniform distribution of blood flow to both lungs. This is a risk factor for development of vascular injury secondary to overflow to one of the lungs and surgical correction is of particular importance to prevent those lesions.
Objective: Evaluate the therapeutic approach and results of treatment of patients with non-confluent PA.
Methods: Retrospective analysis of patients with non confluent PA from January 1995 to December 2012.
Results: 27 patients with non-confluent PA were identified. 16 patients (59%) had concomitant severe hypoplasia of one or both PA. In one third, one or two PA had origin in major aorto-pulmonary collaterals (MAPCAS). In over one third (37%) one of the PA had origin in a ductus arteriosus; 18.5% presented extra hilar PA agenesis with intrapulmonary arteries fed by multiple MAPCAS. 17 patients had pulmonary atresia with VSD's, 5 had tetralogy of Fallot (ToF) and 5 had other forms of congenital heart disease.
Re-establishment of continuity between the two PA was possible in 15 patients (56%). In 8 this was performed with a Goretex® tube between the two PAs, in 5 patients a patch was used (Goretex® in 3, pericardium in 2) and in the remaining 2, direct anastomosis was performed between the two pulmonary arteries. All patients who achieved total correction (n = 12) underwent previous palliative surgery, except for one patient with ToF. Of these 12 patients, 2 had PA continuity re-established before and 7 at the time of total correction. In 3 patients re-establishment of continuity between the two PA was not possible as there was extra-hilar pulmonary agenesis.
Three perioperative deaths occurred in 37 surgeries. Mean follow up after restoring confluence of PA was 8.7 years. Balloon dilation of PA or stent implantation was performed in five cases.
Non-confluent PA occurs in the setting of a variety of congenital heart defects and in the majority of cases surgical correction can be achieved. Presence of non confluent PA is an additional risk factor for mortality.

P-209

Fast-tracking in Paediatric Cardiac Surgery: A Retrospective Audit

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Introduction: Despite the evolving complexity of congenital heart disease cases requiring surgical intervention, improvements in antenatal screening, prompt neonatal diagnosis and advances in surgical techniques have seen survival rates rising. Traditionally post-operatively these patients spent prolonged periods intubated and ventilated in intensive care. More recently there has been a move towards an evolving practice known as 'fast-tracking' where patients are extubated and moved to HDU within a few hours of surgery.
Potential benefits of this approach include reduced length of ITU and total hospital stay, reduced incidence of complications associated with prolonged ventilation, reduced impact of hospitalisation on the child and family, improved resource allocation and reduced expenditure.
Methods: A retrospective audit of 157 cardiac surgical patients over a sixth month period at a single large tertiary referral centre in the North West of England was conducted by a single researcher. Cases requiring cardiopulmonary bypass and those not requiring bypass were included. A wide range of procedures were performed including TOF repair, PDA ligation, ASD and VSD closures, arterial switch procedures, PA banding and pacemaker insertions. Ages of cases ranged from 2 days to 19 years of age.
Results and Conclusions: Results revealed 33% of patients were extubated within eight-hours post-op despite no current protocol being in place for fast-tracking. The incidence of post op complications was 32.7% amongst the fast tracked group, compared to 54.3% of conventionally managed patients. No significant differences in rates of re-intubation were observed between the two groups. Length of ITU and total hospital stay were reduced within the fast-tracked cases.
Fast-tracking has the potential to improve service planning and encourage more cost-effective use of resources in addition to the benefits for the individual child and family. With Paediatric cardiac surgery provision in the UK currently under review any move than could potentially enhance the sustainability of such a fundamental service should be embraced. Whilst this audit is a small scale and single site it highlights significantly positive outcomes.
Further larger scale, multicentre audits are required to add further weight to these findings and provide comparison of mortality rates between fast-tracked and non fast-tracked cases.

P-210

Coronary Artery Anatomy in neonates with Transposition of the Great Arteries undergoing the Arterial Switch Procedure – Comparison of preoperative trans-thoracic echocardiography and intra-operative findings

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Objective: This study compares the pattern of coronary artery anatomy in Transposition of the Great Arteries (TGA) using 2D trans-thoracic echocardiography (TTE) with intra-operative findings during arterial switch procedure.
Methods: Single centre retrospective review of 213 children with TGA and underwent the arterial switch operation from January 2001 to December 2012.
Results: 213 patients underwent neonatal arterial switch procedure from January 2001 to December 2012. The male: female ratio was found to be 3:1. 96% patients had documented coronary artery (CA) anatomy delineated by TTE preoperatively (9 cases did not have documented echo findings describing CA anatomy). 165 cases (81%) showed correlation between

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

pre-operative TTE and intra-operative CA anatomy patterns. However, in 39 cases (19%) there was no correlation. Table 1 shows the overall comparison between the 2 groups. Where there was an abnormal CA pattern this was only detected in 21 cases (37%) prior to the arterial switch procedure. The most common abnormal CA variant found was that with left anterior descending artery (LAD) arising from anterior facing sinus (sinus 1) while right coronary artery (RCA) arising from posterior facing sinus (sinus 2) dividing further into main and circumflex branches. 14 cases had a single CA ostium giving rise to both the left and right coronaries. The remainder of abnormal patterns was composite of all coronary artery variations previously described, making a total of 57 cases with abnormal CA anatomy.

Table 1: *Comparison between preoperative echocardiography and intra-operative CA findings.*

Surgical intraoperative CA findings	ECHO Normal CA anatomy*	ECHO Abnormal CA anatomy^	Total
Normal	144	3	147
Abnormal	36	21	57
Total	180	24	204

** LAD & Circumflex arising from sinus 1 and RCA arising from sinus 2*
^ Any variation from normal CA anatomy defined

Conclusion: In conclusion, there was good correlation between pre-operative echocardiography and intra-operative findings when the coronary artery anatomy was found to be normal. With abnormal coronary anatomy a mismatch was found between the two modalities. However, this had no bearing on eventual outcome following the arterial switch procedure.

P-211
Patient handover at PICU: Introduction of a modified Formula 1 model

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Introduction: 2010, during, a visit to the CICU, Great Ormond Street Hospital (GOSH), England we observed a method for patient handover after congenital heart surgery. A large study (Ped. Anesth. 2007, 17:470) aiming at increasing quality and safety developed a method that was clear, systematic and based on teamwork. The idea came from Formula 1 racing, pit-stop technique and the method had been modified to suit the healthcare sector. The procedure was: written prehandover submitted by the surgical team to the CICU, clear roles and responsibilities for team members and an oral handover given by the pediatric anesthetist and pediatric cardiac surgeon to the CICU staff. Postoperative care was discussed and the responsibility handed over to the intensive care team.
We saw a potential in introducing the model at our PICU, Skane University Hospital Lund, Sweden, where approximately 300–350 children/year undergo cardiac surgery.
Method: A multi professional working group consisting of a pediatric anesthetist, an anesthesia nurse, a pediatric cardiac surgeon, two critical care registered nurses and a nurse assistant was created. The team reviewed the existing literature and designed an observational study for the handover at our institution. A modification of the GOSH Formula 1 method was developed to fit our needs. A prehandover report sheet, checklists and report templates were devised. All involved professionals were informed prior to project launching. After a short trial period the final project was introduced and later evaluated.
Results: A modified Formula 1 method was introduced and adapted to our PICU. The benefits of the project included clearer handover structure, coherent information to the PICU team, improved learning experience for all involved professionals, increased patient safety, time and economical saving.
Conclusions: We have with some modifications, successfully introduced a Formula 1 handover method for the cardiac patient in our PICU. We aim at applying the same strategy to all patient treated in our unit.

P-212
Kommerell diverticulum should be removed when operating symptomatic children with right aortic arch and aberrant left subclavian artery (vascular ring)

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Background: Right aortic arch with aberrant left subclavian artery is the most frequent cause of vascular ring. Usual treatment in symptomatic children is ligamentum arteriosus division, leaving the Kommerell diverticulum in place with potential risk of residual compression, aneurysmal dilation and dissection or even rupture. Translocation of the aberrant left subclavian artery to the left carotid artery together with removal of the Kommerell diverticulum and division of the ligamentum through a left thoracotomy is currently advocated to avoid those complications.
Methods and results: Between 9/2009 and 8/2011, 13 patients underwent above-mentioned procedure. Clinical findings, surgical procedure and complications, histopathological findings and follow-up data were retrospectively analyzed. Mean age at time of surgery was 7.2 years (median 4.3, range 0.9–18.9), mean weight 25 kg (median 18, range 8.4–59). All had respiratory symptoms, associated with dysphagia in 5. CT scan and/or MRI had demonstrated the arch anomaly and the dilated Kommerell diverticulum in all. A left postero-lateral thoracotomy was done in all. All had bilateral cerebral oxymetry monitoring. Post-operative complications included transient chylothorax in 4 and transient phrenic palsy in 1 patient. Mean follow-up reached 6,6 months (median 1,1, range 0.1–29). Mild residual respiratory symptoms were noted in 6 patients. Echo-Doppler analysis available in 11 patients showed a patent left subclavian to carotid artery anastomosis. Histopathological analysis of the resected diverticulum, available in 6 patients, showed cystic medial necrosis and inflammatory tissue in 3, borderline cystic medial necrosis in 1, hyperplastic myo-intimal lesions in 1 and non specific histological findings in 1.
Discussion: Translocation of the aberrant left subclavian artery together with Kommerell diverticulum resection and ligamentum division is a safe and efficient procedure for symptom relief. The observation of profound wall abnormalities such as medial necrosis in at least 50% of the analyzed diverticuli encourages us to maintain this strategy, in order to reduce the risk of aneurysm formation and dissection.

P-213
11 years follow-up of Left atrioventricular valve (AVV) stenosis after complete repair of Atrioventricular septal defect (AVSD): echocardiographic predictors of outcome

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https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Introduction: Despite of the excellent outcome of AVSD repair achieved in the last 50 years, the left AVV remains the "achilles" heel for surgeons. Several surgical techniques has been used, focusing in minimizing residual AVV regurgitation. However, there is a paucity of data regarding of left AVV stenosis. We sought to identify the incidence, late outcome and risk factors related with this morbidity.
Material & Methods: 221 patients underwent to all types of AVSD repair. Routine intraoperative transesophageal echocardiogram (TEE) were performed in all above 3.5 kg. Among them 18 presented residual Left AVV stenosis (mean gradient $\geq$ 8 mmhg) (group I). Age at the surgery (4 months to 22.5 years/median of 11 month). Group II comprised 26 patients with trivial/mild residual LAVV regurgitation. Measurements of AVV annulus, distance between the two left papillary muscle were obtained from the pre and post-operative TEE. Clinical and surgical data were obtained by chart review.
Results: There were 6 re-operations, including 2 valve replacement and 3 deaths in the early post-op. No late post-op death. Two patients lost follow-up. Among 13 remaining, there was one late re-operation. The LAVV gradient remained stable and/or lower over the time. Mean follow-up was 6.6 years (0.8 to 10.7 y).
Preoperative total common valve annulus and LAVV annulus, measured by intraoperative TEE, were not different between groups. However, the preoperative distance between the papillary muscles were significantly smaller in Group I ($p = .01$). Postoperatively, LAVV annulus were reduced comparing with preoperative values, in Group I ($p < .001$), but not in Group II. Partial AVSD, Down's Syndrome, Tetralogy of Fallot and Heterotaxy syndrome were not risk fact, as the surgical technique neither.
Conclusions: Residual LAVV stenosis occurred in 8.1%. The distance between the two left papillary muscles (which support the trileaflet LAVV and also predict the mural leaflet length) was significantly smaller in those patients with residual LAVV stenosis. The LAVV annulus decreased after repair in patients who's LAVV became stenotic. The need for further re-operation was low and overall patients remained stable from the clinical point of view.

P-214
The preservation of the pulmonary valve during early repair of Tetralogy of Fallot: anatomical substrates on surgical correction
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Background: Despite the excellent survival for tetralogy of Fallot (TOF) repair, long-term pulmonary regurgitation and subsequent morbidities remain a challenges for these patients. We investigated the feasibility and the impact of the preservation of the PV during early repair of TOF, focusing our attention on the anatomical substrates of patients with TOF.
Methods: All patients with TOF who were scheduled for PV's preservation between January 2008 and January 2013 were enrolled. Surgical correction involved the combination of early transatrial repair (TAP) and intraoperative balloon dilation of the PV annulus (IBPV). Outcome of this study was the evaluation of PV competence at follow-up outcome. To support this new developed technique a series of 101 specimens were analyzed focusing of the PV and the right ventricular infundibulum.
Results: Twenty-eight of 32 patients (875%) patients underwent a successful preservation of the PV. In the remaining 4 patients (12,5%) the procedure was converted to a classic TAP. Median age at surgery was 100,5 days (range 36–521 days). No procedure-related complications were reported. Median follow-up time for the IBPV patients was 610 days (207–1763 days). Twenty-four patients showed none-mild PV regurgitation (PVR) (85%), 4 had a moderate PVR (15%). Median RV function was 55% (range 45%–65%). Based on our autoptical series, the PV was predominantly bicuspid (65%), followed by tricuspid (23%) and more rarely unicuspid (12%). In 54 patients (53%) PV cusps were normal while in the remaining patients was dysplastic. PV dysplasia was found in almost 50% of all cases in unicuspid/bicuspid valves.
Conclusions: The integrity of the PV annulus and PV function can be preserved in selected patients during early repair of TOF by intraoperative PV balloon dilation. The PV preservation seems to preserve right ventricle function in the mid-term. The majority of specimens presented a normally conformed PV which represent a positive basis for the utilization of this novel technique.

P-215
Oral sildenafil early after cavopulmonary operations in children
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Introduction: Elevated cavopulmonary pressure early after surgical creation of cavopulmonary connections is an important hemodynamic problem with a lot of potential complications and prolongation of the intensive care unit stay. Positive role of the selective pulmonary vasodilators especially of inhaled nitric oxide (iNO) is well established and thoroughly investigated. This study aims to investigate the hemodynamic effect of oral sildenafil in children early after cavopulmonary operations.
Methods: Medical files of patients with cavopulmonary operations for three years period (2009–2012) were retrospectively analyzed. A total of 73 operations were done – partial cavopulmonary connection (PCPC) - 34 and total cavopulmonary connection (TCPC) - 39. 10 patients were treated with oral sildenafil - 6 with TCPC and 4 with PCPC. In 4 patients sildenafil therapy was preceded by administration of iNO. Therapy with oral sildenafil was considered for patients with elevated preoperative pulmonary arterial pressure $\geq$18 mmHg or elevated cavopulmonary pressure $\geq$15 mmHg early after operation. Cavopulmonary pressure was serially measured before sildenafil treatment and in the first 24 hours after initiating it. Data were presented as medians with range or means $\pm$ standard deviation. A parametric paired samples T-test integrated in the statistical software SPSS v.19 was used. A value of $p \leq 0.05$ was considered significant.
Results: Median age of the patients was 36 months (range 6–48 months). Median weight was 11,25 kg (range 6–15 kg). Median dose of oral sildenafil was 20 mg/24 h (range 12–30 mg), in three or four divided doses. Sildenafil therapy was started between 6^{th}–118^{th} postoperative hours (median 32,5 hour). Initial cavopulmonary pressure was 17,0 $\pm$ 3,09 mmHg and changed to : 15,2 $\pm$ 2,48 mmHg at the first hour ($p = 0.016$); 14,7 $\pm$ 3,36 mmHg at the 6^{th} hour ($p = 0.009$); 13,7 $\pm$ 1,94 mmHg at 12^{th} hour ($p = 0.007$) and 13,1 $\pm$ 2,94 mmHg at 24^{th} hour ($p = 0.01$).

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

Conclusion: oral sildenafil early after cavopulmonary operations seems to be effective in our small group of patient. We observed a significant reduction of the cavopulmonary pressure for 24 hours after initiation of treatment with effect onset at the very first hour.

P-216

Discharging home an eight years old child on intra-corporeal left ventricular assist device as a bridge to decision

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Introduction: Long-term mechanical circulatory support with ventricular assist device (VAD) has become a routine treatment option for heart failure in adults. The patients are discharged home on device to improve their quality of life. Pediatric VADs were mainly used as an inpatient treatment so far because of the relatively complexity and size of the equipments. Newer devices now offer the perspective of an ambulatory treatment also in children.
Case report: We report the case of an eight years old girl (body surface area 0.97 m^2) suffering from inotropic dependent rapid onset biventricular heart failure due to chemotherapy-induced acute toxic cardiomyopathy after treatment of a bone sarcoma. The child was provided with an intra-corporeal left-VAD (HVAD Heartware) as a bridge to candidacy. Apart from prolonged medical supportive right ventricular recovery post-operative course was uneventful. Ventilation time was 7 days, ICU stay 28 days. The girl was discharged home after three and a half months hospitalization on oral medications with diuretics, ACE-inhibitor and beta-blocker and a triple anticoagulation (phenprocoumon, acetylsalicylic-acid, dipyridamole). Prior to discharge an extensive training of the child, parents, family doctor, the whole school class including teaching staff and emergency services was provided. In a multipart concept institutional guidelines were created for emergency situations and provided for everyone. Training was offered at our institution including lectures (function of the systems, different alarms) followed by hands-on training with a simulation tool. A workshop for the emergency service was organized. Finally training was also done at the home village of the girl and in her school.
Follow up after nearly two months at home with daily school attendance and regular ambulatory visits was uneventful.
Discussion: Third generation implantable VADs like the Heart-ware HVAD are applicable as intra-corporeal devices in children with a body surface area $\geq 0.6\,m^2$. Discharge from hospital to habitual surrounding with improvement in the quality of life of the whole family is possible, if extensive training of the child, parents and the neighbourhoods including emergency services and school staff is ensured.

P-217

VSD closure using detachment of anterior leaflet of the tricuspid valve

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Objectives: To analyse the impact of detachment of anterior leaflet of the tricuspid valve in closing various types of ventricular septal defects.
Methods: 161 patients operated upon (2005–11) for closure of VSD (151 perimembranous, 4 malalignment, 4 inlet and 2 infundibular) using annular detachment of the anterior tricuspid valve leaflet were analysed. Median age and weight were 5 (Range 1–202) months and 5.3 (Range 2.6–53) kg respectively. VSD closure was performed using running polypropylene stitches. The detached leaflet was reattached while suturing the upper margin of the patch. Follow-up data was available from 148 patients with a median duration of 20 (Range 4–66) months.
Results: Early and late survival was 100%. Incidence of mild and moderate/severe tricuspid valve regurgitation was 13 and 1% at follow-up, 37 and 0% postoperatively as against 27 and 3% preoperatively. Incidence of mild aortic regurgitation was 1% at follow-up, 9% postoperatively as against 8% preoperatively; none had more than mild regurgitation. Trivial postoperative rest shunts seen in 23 (14%) were visible in 6 (4%) patients at follow-up. 3 patients (2 with inlet extension and tricuspid adhesions, 1 with DORV) suffered complete heart block.
Conclusion: Periannular detachment of the anterior leaflet of the tricuspid valve allows VSD closure staying clear of the sub-valvular suspension apparatus. Clear delineation of the inlet margin of the VSD facilitates its closure. Not only that elective detachment of the leaflet does not negatively impact tricuspid valve function but may even contribute to normal valve function.

P-218

Total correction of Truncus arteriosus: Conduit versus conduitless reconstruction of the right ventricular outflow tract

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Objective: Retrospective analysis of repair of truncus arteriosus with focus on the right ventricular outflow tract repair strategy.
Methods: 14 patients undergoing truncus repair (2005–11) were analyzed. Median age was 24 (0–64 months). Based on surgeon's preference, 6 underwent RVOT reconstruction with a Contegra while 8 underwent conduitless RVOT reconstruction with a 3 patch technique and a monocusp (PTFE 4 and Contegra monocusp 4). Moderate to severe truncal valve regurgitation necessitated reconstruction in 6 patients. 2 patients had repair of interrupted arch and 2 reimplantation of arteria lusoria. Median follow-up was 40 (3–80) months.
Results: There was no mortality. 1/14 had severe truncal valve regurgitation at follow-up. 1 patient receiving a 12 mm Contegra needed revision with downsizing to 8 mm Contegra (bicuspidized) due to technical error. 2/6 patients in conduit group needed intervention for distal RVOT/branch PA obstruction at a median fup of 41 months against 1/8 in the conduitless group at 37 months. 2/6 conduits had moderate pulmonary regurgitation, while all conduitless repairs had moderate-severe pulmonary regurgitation. Median intubation duration was 6 and 4 days respectively.
Conclusions: Contemporary results of truncus arteriosus continue to improve despite the complexity of repairs involved. Conduitless RVOT reconstruction with a potentially growing posterior wall is an evolving option, with the objective of delaying the first reoperation. However, it needs long term follow-up to draw any conclusions.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

YIA-1

Fetal aortic stenosis : which fetuses are candidates for in utero dilatation : a retrospective review over a decade

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Introduction: Fetal aortic stenosis is a complex disease. The aim of antenatal diagnosis is to offer adequate prenatal and neonatal care.
Objective: We sought to investigate the outcome of fetuses with aortic stenosis in order to determine criteria of bad outcome and define candidate for in utero balloon dilatation.
Material and methods: We retrospectively reviewed data of fetuses with aortic stenosis over a decade (2002 to 2012). Fetuses with other cardiac defects were excluded. Fetuses were classified in three groups according the prenatal outcome and the neonatal strategy (group 1 termination of pregnancy (TOP), group 2 biventricular strategy and group 3 univentricular strategy).
Results: 73 fetuses were eligible. Data were available for 71 patients. 30 fetuses had TOP (41%, group 1). 33 (46%) were in group 2 and 8 (13%) in group 3. Patients in group 2 were diagnosed later than those in group 1 and 3 ($p < 0.05$). In group 2, 5 deaths (15,5%) were reported. Left to right shunt across the PFO, retrograde flow in the aortic isthmus, akinetic left ventricle and mitral valve inflow abnormalities were associated with extremely poor outcome (termination, univentricular physiology or death in case of biventricular strategy). When LV was of normal size or dilated, only four patients evolved to hypoplastic left heart syndrome (HLHS). However when considering hemodynamic criteria, no evolution from good form to HLHS was noted. In utero balloon dilatation was performed in 7 fetuses (10%) as an alternative to TOP. After intervention, only 2 had a biventricular physiology at birth, 2 a univentricular physiology and pregnancy was terminated in the remaining 3 cases due to evolution to HLHS.
Conclusions: Fetal aortic stenosis is a severe disease diagnosed at various ages of gestation Evolution to HLHS can be predicted by full echographic assessment including shunt across the PFO, LV size and function, flow across aortic isthmus and valve and MV inflow. In utero intervention should not be indicated in fetus with good prognosis. The number of eligible fetuses is very low in our experience (around 3 per year). The place of in utero dilatation remains to be precised.

YIA-2

Pulmonary valve regurgitation and the brain natriuretic peptide level in patients long after repair for Tetralogy of Fallot

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Objective: Since the first intracardiac surgical Tetralogy of Fallot (TOF) repair was reported, the outcome of patients after corrective surgery has improved over the years. However we are now faced with an increasing number of patients who present with pulmonary valve regurgitation (PR) or right ventricular (RV) dysfunction, and these patients always have a risk for sudden death. The severity of PR and the RV end-diastolic volume were reported to show a positive correlation with the brain natriuretic peptide (BNP) level in patients with surgically repaired TOF, but this has been controversial. The purpose of this study was to evaluate the BNP levels, and to address the issue of optimal surgical timing in patients who underwent TOF repair.
Methods: We retrospectively reviewed 33 patients (20 males, 13 females) older than 10 years of age who underwent TOF repair at our institution.
Results: The median age at the initial repair was 13 months (range, 4–38 months). The median time that had passed since the original corrective surgery was 12 years (range, 10–20 years). Ten reinterventions were required in our patient group, including pulmonary valve replacement (PVR), pulmonary artery plasty, tricuspid valve repair, right ventricular outflow tract reconstruction (RVOTR) and ventricular septum defect (VSD) closure. In the patients who underwent PVR, the BNP levels before the procedure were significantly higher than those in the other patients (median 54.0 vs. 21.8 pg/ml, $P < 0.05$). In the patients who did not have PVR, the BNP levels of the patients who had moderate or severe PR were significantly higher than those of patients who had trivial or mild PR in echocardiography (median, 26.0 vs. 17.0 pg/ml, $P < 0.05$). After the PVR procedure, the PR was improved all the patients and the BNP levels were significantly decreased (median, 54.0 vs. 32.7 pg/ml, $P < 0.05$).
Conclusions: The severity of PR may have a relationship with the BNP level long after repair for TOF. The BNP level may contribute to defining the indication and timing of TVR for TOF late after the initial operation. PVR after TOF repair significantly improves the RV function, and may reduce the risk for sudden death.

YIA-3

One hundred cases of arterial duct stenting in a single centre: a mid term follow up

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Introduction and aim: Arterial duct (AD) stenting is nowadays considered as a valuable alternative to surgical shunt in patients with congenital heart disease and duct-dependent pulmonary circulation (CHD-DDPC). The aim of this study was to review clinical, procedural and follow-up data of a large single-centre series of patients submitted to AD stenting as palliation of CHD-DDPC.
Methods: Between April 2003 and April 2012, AD stenting was attempted in 100 neonates with CHD-DDPC (n = 14 pulmonary atresia intact ventricular septum, n = 13 critical pulmonary stenosis, n = 73 complex congenital heart disease). Mean age and weight of these patients were 25 ± 50 days (range 3–255 days) and 3.4 ± 1.1 Kg (range 1.2–8 kg), respectively. AD morphology was conical in 30 pts, tubular in 38 pts and tortuous in 32 pts.
Results: Overall feasibility of the procedure was 93%. The procedure was not completed in 7 patient due to extreme ductal tortuosity. Mean procedural and fluoroscopy time were 128.8 ± 59 min and 26.5 ± 27 min, respectively. Ductal stenting was always performed with high-flexibility chromium-cobalt coronary stents. AD size increased from 1.6 + 1.3 to 3.3 + 1.3 mm ($p < 0.0001$) and percutaneous O2 saturation from 74.6% ± 1.1 to 90.4 ± 7.2% ($p < 0.0001$). None intra-operative

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

death was recorded. Overall in-hospital mortality was 3.2%, significantly associated to low-weight (<2.5 kg) at procedure. Complication rate was 4.3% (2 pts partial stent dislodgement after deployment, 1 pt mild right pulmonary artery stenosis, 1 pt femoral artery thrombosis), Over a mid-term follow up (31 ± 15 months), 12 pts (12.9%) needed early surgical shunt due to persistent cyanosis, 17 pts (18%) underwent late duct re-stenting before surgical repair, 18 pts (19.4%) were considered cured despite complete AD closure. Control angiography was performed in 37 pts destined to surgical repair, showing significant growth of the pulmonary arteries (Nakata index increased from 183 ± 151 to 293 ± 103 mm^2/m^2 $p < 0.0001$).
Conclusions: AD stenting is a reliable palliation of CHD-DDPC, supporting spontaneous clinical improvement or in view of later, lower-risk corrective surgery. Procedural feasibility depends on ductal morphology while procedural risks mainly depend on patients' demographic and clinical characteristics. Mid-term fate of the stented AD is spontaneous closure, although allowing significant pulmonary artery growth in view of corrective surgery.

YIA-4
Clinical practice method of evaluating pulmonary perfusion in children with pectus excavatum after surgical repair

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Objectives: We aimed to investigate the effects of surgical correction of left mediastinal displacement in children with pectus excavatum (PE) on pulmonary perfusion, using easily performed and minimally invasive methods, which minimize radiation exposure.
Methods: Consecutive patients with PE who underwent the Nuss procedure at Gunma Children's Medical Center from July 2003 to December 2009 were evaluated. Two of 42 patients with insufficient data were excluded. The remaining 40 patients were prospectively studied pre- and post-operatively (just prior to pectus bar removal). A chest CT scan, radiography, echocardiography, and pulmonary perfusion scintigraphy were performed before surgery and at medium-term follow-up (3.0 ± 0.2 years). The chest CT and radiograph were used to calculate the funnel index (FI) and left displacement index (LDI). LDI was defined as the ratio between the distance from the left border of the mediastinum to that of the thorax and the transverse thoracic dimension on a posteroanterior chest radiograph. Pulmonary perfusion scintigraphy was visually interpreted and the left-to-right count ratio for lung scintigraphy (Ls/Rs) was measured. Pre-ejection period (PEP), acceleration time (AcT), and ejection time (ET) of the right pulmonary artery (RPA) and left pulmonary artery (LPA) were measured by pulse Doppler echocardiography.
Results: Preoperatively, left pulmonary perfusion was significantly impaired compared with right pulmonary perfusion. Postoperatively, sternal depression expressed by FI and mediastinal displacement expressed by LDI were improved ($P < 0.001$). Postoperatively, Ls/Rs was significantly increased (0.85 ± 0.16 vs. 0.96 ± 0.17, $P < 0.001$) and AcT/ET was significantly altered (LPA: 0.29 ± 0.05 vs. 0.33 ± 0.05, $P < 0.001$; RPA: 0.42 ± 0.06 vs. 0.37 ± 0.07, $P = 0.003$). Echocardiography didn't show pulmonary hypertension in any patients. To investigate the useful method, we examined the relationships of FI and LDI with Ls/Rs and AcT/ETLPA. LDI was correlated with Ls/Rs ($R = 0.371$, $P = 0.001$) and AcT/ETLPA ($R = 0.546$, $P < 0.001$). FI wasn't correlated with Ls/Rs and poorly correlated with AcT/ETLPA ($R = -0.324$, $P = 0.003$). There was a significant correlation between ATILPA and Ls/Rs ($R = 0.338$, $P = 0.003$).
Conclusions: Imbalance of pulmonary perfusion in children with PE is improved after the Nuss procedure. There is a clear relationship between the degree of leftward displacement of the mediastinum and decreased left pulmonary perfusion. Children with PE can be followed up with minimal invasiveness.

YIA-5
Prevalence and Morphological Properties of Anatomical Isthmuses involved in Monomorphic Ventricular Tachycardia in Repaired Tetralogy of Fallot

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Introduction: Ventricular tachycardia (VT) is an important cause of late morbidity and mortality in repaired Tetralogy of Fallot (rTOF). The majority of the VTs in rTOF are monomorphic VTs based on re-entry over well defined anatomical isthmuses that can be successfully targeted with radiofrequent catheter ablation (RFCA) in most patients. The prevalence and properties of these isthmuses in the rTOF population are unknown. Extending this knowledge might facilitate RFCA of VT in rTOF.
Methods: In 28 post-mortem specimens with rTOF (age 80 ± 65 months) the presence and size of anatomical isthmuses were assessed. Isthmuses were defined as (1) the isthmus between tricuspid annulus (TA) and RV-scar or RVOT-patch, (2) the isthmus between RV-scar and pulmonary annulus (PA), (3) the isthmus between PA and VSD-patch and (4) the isthmus between VSD-patch and TA. Furthermore, presence of coronary arteries abnormalities, thickness between isthmus 3 and the aortic root and the degree of infundibular resection were assessed.
Results: Total correction with a transannular patch was performed in 75% (n = 21) of hearts; the remaining had RV myectomy with direct closure of the RV. Isthmus 1 (width 23 ± 9 mm) and isthmus 3 (width 13 ± 6, length 13 ± 5 mm) were present in all rTOF specimens. Isthmus 2 (width 7 ± 5 mm) and isthmus 4 (width 6 ± 4 mm) were present in respectively 21% (n = 6) and 11% (n = 3) of hearts. No major coronary artery abnormalities were found; in 11% of cases a large conal branch, anterior to isthmus 1, was present. The area between isthmus 3 and the aortic root had an average thickness of 7 ± 2 mm, independent of age. Infundibular resection was performed in 93% (n = 26) of specimens, which was mild to moderate in 39% and severe to extreme in 54% of cases.
Conclusion: Isthmus 1 and 3 were present in all rTOF specimens. The distance between isthmus 3 and the aortic root, as well as the normal coronary artery pattern of most specimens would allow RFCA. Infundibular muscle resection was severe to extreme in 54% of the rTOF specimens, which might result in significant scarring of isthmus 3.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press

YIA-6

Optimal timing of initial treatment in severe Kawasaki disease: A sub-analysis of the RAISE study

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Introduction: Kobayashi and colleagues showed that addition of prednisolone to the standard regimen of intravenous immunoglobulin improves coronary artery outcomes in patients with severe Kawasaki disease in Japan (RAISE study). The purpose of this study was to evaluate the relationship between the timing of initial treatment and clinical outcome in the same patients.

Methods: In the RAISE study, a total of 242 children with severe Kawasaki disease were randomised at 74 centers in Japan between 2008 and 2010, to intravenous immunoglobulin plus oral aspirin (group G, n = 121) or intravenous immunoglobulin plus intravenous prednisolone followed by oral prednisolone (Group P, n = 121). Z score of the proximal right coronary artery (RCA), the left main coronary artery (LMCA), and the proximal left anterior descending artery (LAD), and the maximum Z score of coronary arteries were estimated at weeks 1, 2, and 4. In this study, patients who received treatment at day 4 or earlier (group 1) were compared with those who received treatment at days 5 to 8 of fever (group 2) both in group G and group P. We did analyses with JMP9 (SAS Institute).

Results: Both in group G and group P, there was no significant difference in the needs of additional rescue therapy, relapse, and the total doses of immunoglobulin between group 1 and group 2. Both in group G and group P, absolute diameters of RCA, LMCA, and LAD were significantly lower in group 1 than in group 2 ($P < 0.05$; ANOVA). In group P, maximum Z score of LMCA was significantly lower in group 1 than in group 2 ($P < 0.05$; ANOVA). In group G, there was no significant difference in Z scores, maximum Z scores, and delta Z scores of coronary arteries during the study period between group 1 and group 2.

Conclusions: Optimal timing of initial treatment still remains controversial. In severe cases of Kawasaki disease, early treatment of KD resulted in lower absolute diameter of coronary artery both in group P and group G, and in lower maximum Z score of LMCA in group P.

https://doi.org/10.1017/S104795111300036X Published online by Cambridge University Press