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Cardiology in the Young

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YIA: Young Investigators' Award; O: Oral Abstract Presentation; MP: Moderated Posters; P: Posters

YIA-1

Telmisartan improves RV function and hypertrophy by modulating processes of fibrosis and autophagy in PA banded rat

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Introduction: RV failure is a significant complication in patients with congenital heart disease with right-sided obstructive lesions, and PH. Effective medical treatment for decompensated RV remains to be elucidated. While ARBs are known to reduce mortality in patients with left side heart failure by inhibiting cardiac remodeling, their effects for RV failure are unknown. Pressure overload stress induces a robust autophagic response and hypertrophic changes in cardiomyocytes. However, previous studies showed conflicting results with regards to whether induction of autophagy was adaptive or maladaptive in overloaded ventricle.
Method: RV failure model rats were surgically generated by pulmonary artery banding using SD rats. Those rats were treated with oral telmisartan (T group: 5 mg/kg/day n = 24) or water as control (n = 12) for 4 weeks. RV-PV loops were examined using a micro catheter. For histological analysis of cardiac muscles remodeling, Masson's trichrome staining and electron microscopy were performed to obtain the view of morphological changes of cardiomyocytes. The level of LC3A/B, and p62 were measured by Western blotting.
Results: Median survival time for the T group was significantly longer than the control rats. There were significant increases in RV cardiac output ($p < 0.01$), RV EDP, end-systolic elastance, and end-diastolic elastance ($p < 0.05$) derived from RV PV loop in the T group. RV/LV + S was decreased significantly in the T group ($p < 0.01$). Quantitative analysis for autophagy showed significant reduction of the number of autophagosome in the T group. Both LC3A/B and p62 expressions were reduced in the T group compared with control. The rate of fibrosis in RV was significantly lower in the T group ($p < 0.01$) as well as mRNA expression of Pro-collagen 3, CTGF, and periostin.
Conclusions: In PA banded rats, telmisartan had effects to improve RV cardiac output and to decrease mortality without reduction of RV pressure by inhibiting cardiac fibrosis, autophagy and RV hypertrophy. Decreased expressions of LC3A/B and p62 indicated that the reduction of autophagy was induced by mitochondria dysfunction. These results suggest that telmisartan prohibit overload-dependent RV failure and fibrosis by blocking mitochondrial dysfunction and telmisartan may be an effective treatment option for RV failure.

YIA-2

The Impact of the Stage I Palliative Strategy on Echocardiographic Changes in Cardiac Size and Function in Children with Hypoplastic Left Heart Syndrome. A comparison between the Norwood and the Hybrid approach

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Background: Hybrid palliation may have theoretical advantages to preserve right ventricular (RV) function in hypoplastic left heart syndrome (HLHS) patients when compared to the Norwood procedure. The aim of this study was to compare echocardiographic indices of RV function in patients after the hybrid and Norwood procedure throughout all stages of HLHS palliation.
Methods: A retrospective study was performed in 76 HLHS patients (34 after hybrid and 42 after Norwood) with available protocol-driven echocardiography. Indices for RV size, shape and function (systolic, diastolic and global), tricuspid valve (TV) size and degree of regurgitation (TR) were obtained at 5 specific time points: 1. at baseline before stage I procedure, 2. before stage II, 3. after stage II, 4. before Fontan, 5. after Fontan.
Results: Median follow-up was 4.9 years (range 1.1-8.5 years) of all 76 participants, of whom 50 underwent second stage palliation and 23 Fontan completion. Baseline characteristics before stage I procedure of both groups were comparable. At pre-stage II evaluation, RV fractional area change (FAC) and E/E' ratio ($32 \pm 7\%$ vs. $27 \pm 6\%$; $p = 0.04$ and 12.14 ± 4.11 vs. 8.66 ± 3.58; $p = 0.02$, respectively) were significantly higher in

Norwood patients as compared to hybrid. After stage II, the FAC difference became insignificant (29 ± 7% vs. 25 ± 8%; p = 0.08), although E/E'-ratio remained significantly higher in Norwood patients (18.65 ± 8.30 vs. 11.07 ± 7.01; p = 0.04). Before and after Fontan completion, RV systolic function equally improved in both groups as expressed by a higher FAC and improved qualitative RV function. Moderate/severe TR was frequently present especially in Norwood patients after stage II (13 patients after Norwood vs. 4 after hybrid), which significantly improved at pre-Fontan assessment (moderate/severe TR in 3 Norwood patients and 1 hybrid) and after Fontan completion (moderate TR in only 1 patient per group).
Conclusion: Norwood and hybrid surgical strategies had equivalent echocardiographic indices of RV size, shape, systolic and diastolic function throughout the full course of palliation of HLHS, offering no favorable Fontan candidacy by either of the two techniques. Small differences in individual indices of RV function are likely to be explained by differences in physiology or surgical timing between both groups, rather than by intrinsic differences in myocardial and valve function.

YIA-3
The importance of right atrium tension in patients after atriopulmonary Fontan procedure
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Introduction: Elevated venous pressure is thought to play a pivotal role in the development of characteristic complications such as liver fibrosis/dysfunction, arrhythmias, protein-loosing enteropathy and thrombosis after Fontan surgery, particularly that for the atriopulmonary connection (APC). On the other hand, there are many patients who have dilated RA after APC-Fontan. We hypothesized that the right atrial tension (RAtension) rather than RA pressure (RAp) is important for the progression of these complications after APC-Fontan.
Methods: We studied 51 consecutive APC-Fontan patients who underwent cardiac catheterization (median postoperative period; 14 years). We calculated the RAtension from RAp and RAradius to be counted backward by RAvolume (RAv) assuming that RA as a sphere according to Laplace's law [tension (dyne/cm) = transmural pressure (dyne/cm2: Central Venous Pressure (mmHg) × 1333) × radius (cm:$3\sqrt{(3 \times \text{volume (cm2)}/4\pi)}$ / 2]. RAv was calculated by a biplane Simpson's method using RA-graphy. We investigated the correlation between the hemodynamic data by their cardiac catheterization (RAtension, RApressure, RAradius, Qs, Rp, EDP and EDV) and the complications of APC-Fontan
Results: Out of 51 patients after APC-Fontan, 27 patients had these complications (Liver fibrosis; 23, arrhythmias; 2, protein-loosing enteropathy; 1 and thrombosis; 1). Age and postoperative period were not correlated with these complications. Among the hemodynamic data, only the RAtension and RAradius were significantly correlated with these complications (p = 0.02 and p = 0.03). In this study, cut-off level of RAtension for presence of Fontan complications was 23,050 dyne/cm by receiver operating characteristic curve (sensitivity; 81.4% and false positive rate; 33.3%). Moreover, it was very interesting result that the RAtension was most strongly correlated with log] BNP (brain natriuretic peptide)](r = 0.438) among the hemodynamic data.
Conclusion: The present results indicate the importance of RAtension rather than high venous pressure for the development of post-Fontan complication and elevation of BNP. However, we should pay attention that even a little elevation of RApressure caused by exercise increase the RAtension on a large scale under the circumstance of RA dilation according to Laplace's law.

YIA-4
Transcatheter Stenting and Upsizing of Stenosed GoreTex Grafts Delays Further Surgery in Complex Congenital Heart Disease
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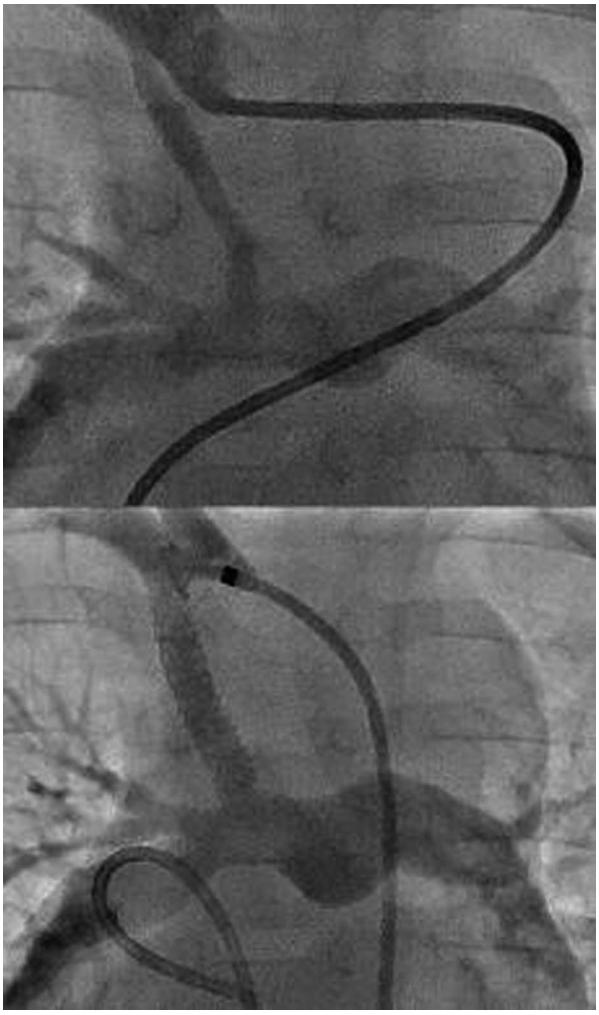
Introduction: Small GoreTex grafts are frequently used in the initial palliation of cyanotic congenital heart disease and increasingly for PA reconstruction or as an alternative to small homografts used as RV-PA conduits. Catheter interventions have been described to address graft failure. This study seeks to review our 10-year experience of stenting with a view to upsizing GoreTex grafts in an attempt to delay or obviate surgical re-intervention.



Methods: Retrospective case-note and angiography review of all patients undergoing stenting of GoreTex grafts between 2003 and 2013 at a single tertiary referral center.

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Results: Fifty-nine patients underwent 68 interventions at a median interval of 126 (3-3379) days after GoreTex graft insertion. At catheterization, median age was 131 days (17-2530), median weight was 5.2 Kg (3.2-28.6). A total of 76 stents were implanted into 68 GoreTex grafts. Coronary stents were used in 84% of the procedures. Grafts stented were RV-PA conduits (n = 40 (58%)), central/BT shunts (n = 23 (33%)) or reconstructed PAs/Fontan (n = 6(8%)). Two or more stents were placed at 18 procedures (26%).
In patients with Shunts or RV-PA conduits, oxygen saturations increased from median of 68% (50-82) to 82% (60-94), [$p < 0.001$]. From 2007, the choice of chosen stent size routinely exceeded nominal graft size. (See fig 1.) Where graft upsizing was achieved (n = 26), median nominal graft size increased from 3.5 (3-8) mm to 4.75 (4-12.3) mm [$p = 0.01$]. Median nominal graft cross sectional area increased from 38.5 (28.2-201.1) mm^2 to 70.9 (50.2-475.9) mm^2 [$p = 0.03$].
Median deferral of further surgical intervention was 191 days (SD 266, IQR 39-233). For patients <4 months of age awaiting cavopulmonary connection (n = 38), surgical graft revision was obviated in 58% (n = 27).
There was 1 procedural death and 2 deaths within 7 days of the procedure. There were 7 other major complications, mostly in patients post Norwood stage 1.
Conclusions: Stenting and over dilatation of GoreTex shunts is feasible and relatively safe. The size of GoreTex can be significantly increased, delaying subsequent surgical intervention or obviating the need for additional surgical shunt procedures in the majority of patients.

YIA-5
Survey of School Students indicated of Implantable Cardioverter-Defibrillators

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Objectives: With the spread of public access defibrillation and the advance of management for life-threatening arrhythmia, reports of survivors from cardiopulmonary arrest in school students are increasing. As some survivors among them are supposed to be indicated implantable cardioverter-defibrillator (ICD), those demographical data were surveyed.
Methods: Cases were searched and picked up with the keywords of ICD, AED, defibrillation, or ventricular fibrillation from all injury or illness reported to the mutual aid insurance system under school supervision in Japan from 2007 to 2012. Their school grade, sex, and original disease that have possibility of ventricular fibrillation, are summarized.
Results: Twenty-four cases were reported that ICD was implanted after resuscitation. One case occurred in 2007, 3 each in 2008 and in 2009, 8 in 2010, 6 in 2011, and 3 in 2012. The youngest case was in the 1st grade in middle school, 2 cases in the 2nd of middle school, 3 in the 3rd of middle school, 5 in the 1st of high school, 8 in the 2nd of high school, and 5 in the 3rd of high school. High school students occupied 75%. Twenty-one cases (87.5%) were male students. Basic diseases diagnosed before event occurrence were 7 hypertrophic cardiomyopathies (HCM), 1 operated congenital heart disease. Other 16 cases were not diagnosed before event and detail examinations for them revealed 1 case each of HCM, dilated cardiomyopathy, Brugada syndrome, long QT syndrome, ventricular tachycardia, vasospastic angina, and 10 cases were assumed to be idiopathic ventricular fibrillation (IVF). All events except one were occurred during or immediately after hard exercise. Exceptional case occurred during playing saxophone. Eighteen cases fell while running, 2 while basketball game, 2 while swimming, and 1 in kendo (Japanese swordsmanship). Schoolteacher or bystander equipped and operated automated electrical defibrillator (AED) in all cases except one case of operated congenital heart disease.
Conclusions: Number of patients who received ICD implantation is increasing in school, especially for patients with HCM and IVF. Pediatric cardiologists and caregivers are requested to have knowledge to take care of ICD implanted students.

YIA-6
3rd trimester fetal heart rate discriminates between non-carriers, carriers of single and double LQT1 mutations in Swedish Long QT Syndrome families

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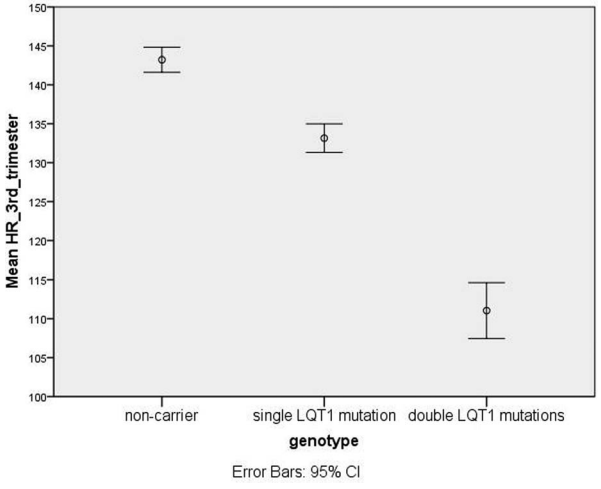
Introduction: Early diagnosis is of utmost importance in the congenital Long QT Syndrome (LQTS), especially for fetuses carrying double mutations, a condition associated with a high risk for life-threatening arrhythmias early in life. Here we investigate 3rd trimester fetal heart rate, routinely recorded within public maternal health care, as a possible early marker for LQTS genotype.
Methods: In large Swedish LQTS families where molecular genetics cascade screening for LQT1 (KCNQ1 gene) mutations had been performed in the clinical setting, fetal heart rates during the 3rd trimester (gestational week 29- birth) were noted for 124 fetuses with ascertained genotype, whereof 45 were non-carriers of the familial mutation(s), 67 were heterozygous mutation-carriers and 12 carried double mutations (3 homozygous and 9 compound heterozygous mutation-carriers). Among the heterozygous mutation-carriers, genotypes included p.Y111C (n = 63), p.A525 T (n = 3) and p.R518X (n = 1). Among the carriers of double mutations, genotypes included R518X/R518X = 3, A525T/R518X = 4, M159sp/R518X = 2, R190W/R518X = 1, R530W/R518X = 1 and S349W/R518X = 1. Sex was equally distributed within each genotype group (females 62%, 46% and 50%, per group).



The mean fetal heart rate per individual, calculated from all recordings from gestational week 29 and onwards, noted in the maternal health care records, were summarized and

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compared between genotype groups (no mutation, single mutation, double mutations) using t-tests and non-parametric tests, as appropriate.
Results: Mean 3rd trimester heart rate correlated with fetal genotype (no mutation 143 ± 5 bpm; one mutation 133 ± 8 bpm; double mutations 111 ± 6 bpm, $p < 0.001$). Mean heart rates were statistically lower per added mutation (no mutation vs. single mutation $p < 0.001$, single mutation vs. double mutation $p < 0.001$). Fetal sex did not significantly affect mean 3rd trimester heart rate ($p = 0.246$).
Conclusions: In this study including 124 fetuses with ascertained genotype from Swedish LQT1 families, 3rd trimester fetal heart rate discriminated between fetal genotypes (no mutation, single mutation and double mutations). This finding strengthens the role of fetal heart rate in the early diagnosis of familial LQTS, and importantly for the identification of fetuses with double mutations, at high risk of early life-threatening arrhythmias.

O1-1
Importance and implications of the occurrence of supraventricular arrhythmia in children with Catecholaminergic Polymorphic Ventricular Tachycardia

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Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) in children is rare malignant arrhythmia with a high risk of syncope and sudden cardiac death (SCD) due to the stress-induced polymorphic VT. In addition to ventricular arrhythmias different types of supraventricular tachyarrhythmia (SVT) is the common cause of inappropriate ICD shocks. The aim was to assess the prevalence and clinical significance of SVT types in children with CPVT.
Patients and Methods: 24 pts (15 males -63%) aged from 4.5 to 14.0 with exercise- induced CPVT and syncope were enrolled in the study. Family and clinical history, 12-leads ECG, 24-hour ECG monitoring, stress-testing, echocardiography, blood chemistry, end EP study (11 pts) were done. Follow-up time varied from 3 to 18 years.
Results: According to Kaplan-Meier survival analysis, 50% of CPVT children experienced first syncope before the age of 5 and more than 35% developed aborted cardiac death or SCD before the age of 13. Family history of SCD or syncope took place in 38% of pts. ECG and Holter revealed sinus bradycardia in 83%, PQ interval ⩽110 ms – in 68%, ventricular premature beats – in 55%, and AV-dissociation – in 55% of pts. SVT as a trigger of CPVT was confirmed in 77% of pts. Among them typical or atypical atrial flutter (AFl) was found in 32% of cases. 11 pts were implanted with ICD. Inappropriate shocks were mostly associated with SVT with high ventricular response. Adding to the beta-blocker therapy the sodium-channel blockers has allowed to control supraventricular triggers and improve the prognosis. In one patient with polytopic SVT and high AV conduction (210 bpm) AV node cryomodification reduced ventricular rate to 125 bpm.
Conclusions: Vulnerability to ventricular and also to supraventricular arrhythmia characterizes young patients with CPVT. SVT including atrial flutter is significant as trigger of malignant ventricular arrhythmia and as a cause of inappropriate ICD shocks. Combined antiarrhythmic therapy and cryomodification of AV-node could be suggested for SV triggers control.

O1-2
Risk factors predicting the future presence of long QT syndrome-related symptoms in pediatric patients diagnosed by school-based electrocardiographic screening programs in Japan

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Background and Objectives: Few data are available regarding risk factors predicting the future presence of long QT syndrome (LQTS)-related symptoms in pediatric patients who were diagnosed by school-based electrocardiographic screening programs.
Subjects: Subjects were 281 children and adolescents with LQTS (M/F = 144/137; age at diagnosis, 10.9 ± 3.2 years; observation period, 4.0 ± 4.0 years) who were screened by the program. LQTS-related symptoms were defined as syncope, sudden death, and aborted cardiac arrest. In the regression analyses, age, sex, QTc values by Fridericia's formula at diagnosis, past history of LQTS-related symptoms, the presence or absence of family history of LQTS or sudden cardiac death and observation periods were used as independent variables.
Results: Of 281 subjects, 23 (8%) had past history of symptoms. Triggers were exercise in 9 and swimming in 8 subjects. After diagnosis, 28 subjects (10%) developed symptoms; triggers were exercise in 11 and swimming in 1. The prevalence of exercise as a trigger was not different between before and after diagnosis. However, swimming as a trigger significantly decreased ($p = 0.007$) after diagnosis compared with before diagnosis. One child died. Logistic regression analysis showed that QTc value ($p = 0.001$), past history ($p = 0.0002$), the presence of family history of LQTS ($p = 0.0003$) and observation period ($p < 0.0001$) were predictive for the future presence of symptoms. Multivariate logistic regression analysis showed that observation period was a sole risk factor to predict the future presence of symptoms (Coefficient/SE, 2.84; p value of 0.005; Odds ratio, 1.14; 95%CI, 1.04-1.25), and that among patients who were treated (50 patients), drug incompliance was also a sole predictive factor for future presence of symptoms (Coefficient/SE, 2.67; p value of 0.008; Odds ratio, 7.88; 95%CI, 1.73-35.8).
Conclusions: Pediatric LQTS patients who were screened by programs should be monitored for long periods of time, in addition to individuals who were visited and diagnosed by the presence of LQTS-related symptoms. New strategies are needed to prevent exercise-triggered symptoms after diagnosis in these patients. Drug compliance should be monitored after initiation of therapy.

O1-3
Heating Effects of Magnetic Resonance Imaging on Epicardial Pacing Leads

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Introduction: Nowadays, most manufacturers offer standard transvenous cardiac pacemaker systems that are not contra-indicated for MR imaging (MRI conditional). In infants and children, epicardial pacing leads (epiL) are preferably used compared to the transvenous pacing leads (transL). Up to now, no epiL has been labeled as MR conditional. The aim of this study was to investigate the potential risk of epiL during MRI with a series of in vitro measurements. The situations of an intact pacing system and of abandoned leads were simulated.
Methods: Heating effects of MRI in a 1.5 T Scanner (Achieva, Philips) were measured at the tip of the pacing lead during a TurboSpinEcho sequence with a whole body SAR of 2 W/kg (upper limit for normal operating mode) during 2 minutes. The transL (CapSureFix MRI SureScan, 5086-45 cm, Medtronic) was compared with the epiL (CapSureEpi 4968-35 cm, Medtronic) in a gel filled tank (as described in ASTM F2182-11a) in a worst case linear configuration parallel to the tank wall.
Results: Three series of temperature measurements were undertaken: (1) Lead connected to a pulse generator (PM; EnRhythm MRI SureScan, Medtronic): There was a temperature rise of +2.5°C in the transL (= reference for all following results. This setup is MR conditional). The epiL showed a 4 × higher heating, (2) lead without PM: TransL 4 × higher heating, epiL 30 × higher heating and (3) epiL coiled to 20 cm length without PM to mimic the shortened (cut-off) lead: 9 × higher heating.
Conclusion: There is a significant heating effect and the current epicardial pacing systems cannot be exposed to an MRI without risk of thermal damage at the tips of the lead, even in case of abandoned leads. MRI of a patient with an abandoned epicardial lead cannot be performed without a high risk of thermal damage at the tips of the lead. Therefore, these patients must be excluded from MRI for lifetime. With increasing role of MRI in postoperative imaging of the heart, concerted efforts are warranted to develop MRI compatible electrodes for children.

O1-4
Determinants of left ventricular dysfunction in children with frequent premature ventricular complexes and/or asymptomatic ventricular tachycardia

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Introduction: For a long time idiopathic frequent Premature Ventricular Complexes (PVCs) and asymptomatic Ventricular Tachycardia's (VTs) were considered benign. However, over the past decade PVCs have emerged as a cause of Left Ventricular (LV) dysfunction in adults. This study aims to assess which determinants of PVCs and VTs are associated with development of LV dysfunction in children.
Methods: For this retrospective study, databases for pediatric patients with the diagnosis of idiopathic frequent PVCs and asymptomatic VTs were searched. Frequent PVCs were defined as 5% or more ventricular ectopic beats in 24 h. LV dysfunction was defined as a shortening fraction of 28% or less. Electrocardiograms, holter recordings and echocardiograms were reviewed.
Results: Seventy-two children were included. Seven showed LV dysfunction at diagnosis (4 (57%) males, age 11 ± 7 years, 3 (43%) had heart failure symptoms) and 65 showed normal LV function (37 (57%) males, age 8 ± 6 years, 21 (32%) with symptoms). Patients with LV dysfunction compared to normal LV function had a higher percentage of PVCs on holter (43 ± 18% vs 16 ± 11%, p = 0.002), higher prevalence of VT (6 (86%) vs 26 (40%), p = 0.048), and a higher number of couplets (7 (100%) vs 33 (51%), p = 0.015). Other determinants which were analysed showed no significant difference. In the group of patients with LV dysfunction, 3 responded to medication (class Ic, II and III) and 6 underwent ablation, of which 1 was unsuccessful. During follow up, LV function normalized in 6 patients. In 1 patient LV dysfunction persisted at follow up, even after a successful ablation procedure. In this patient severe LV dysfunction was possibly related to multiple ablation lesions during prior procedures. In the group of patients with a normal function, no one developed LV dysfunction during follow up.

Table – Comparison of determinants of PVCs in patients with different LV function

	SF < 28% (n = 7)	SF >= 28% (n = 65)	P value
Follow up (years ± SD)	3.8 (± 4.0)	3.9 (± 4.1)	0.830
QRS axis: Inferior (n %)	5 (71)	53 (84)	0.407
Block pattern: LBBB (n %)	3 (43)	42 (65)	0.218
QRS duration of PVC (ms ± SD)	172 (± 20)	162 (± 28)	0.383
QTc of PVC (ms ± SD)	484 (± 30)	487 (± 65)	0.915
Coupling-interval/RR-interval (ratio)	0.61 (± 0.13)	0.65 (± 0.13)	0.456
Couplets (n %)	7 (100)	33 (51)	**0.015**
Bigeminy (n %)	7 (100)	49 (75)	0.336
Trigeminy (n %)	5 (71)	44 (68)	0.841
Quadrigeminy (n %)	2 (29)	9 (14)	0.317
VT (n %)	6 (86)	26 (40)	**0.048**
PVC burden (% ± SD)	43 (± 18)	16 (± 11)	**0.002**
LVEDD Z-score (± SD)	1.9 (± 1.7)	1.3 (± 1.0)	0.195

QRS axis: Inferior = axis of PVC in frontal plane; Block pattern: LBBB = pattern of PVC left bundle branch block; PVC burden = Total percentage PVCs on holter; LVEDD z-score = adjusted value of LV end diastolic diameter by size.

Conclusion: In children with idiopathic PVCs and asymptomatic VTs, development of LV dysfunction, is associated with a higher burden of PVCs, the presence of VTs, and couplets. LV dysfunction appears to be reversible if the burden of PVC's is decreased by medication or ablation.

O1-5
Postnatal Outcome of Isolated AV-Block. A Dutch Retrospective Analysis

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Objective: Evaluation of the postnatal outcome in second and third degree congenital atrioventricular block (CAVB) in the Netherlands during the past decade.
Methods: A nationwide database of all Dutch paediatric cardiac centres was initiated to retrospectively analyse the postnatal outcome of neonates with a CAVB-II°/III° from 2003 to 2013. Exclusion criteria were LQTS, chromosome abnormalities or

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complex congenital heart disease in the absence of maternal autoantibodies.
Results: 48 liveborn children (29 female) with CAVB were included with a mean follow-up time of 5.8 years. One child was first diagnosed with CAVB at birth. Four had CAVB-II°, 43 CAVB-III° and 1 progressed from II° to III°. 42 children were positive for anti-SSA/Ro, 29/42 for anti-SSB/La and 1 for only lupus-anticoagulant. The mean gestational age at birth was 38.0 weeks, with a mean heart-rate of 77 bpm (range 53-110) in CAVB- II° and 59 (40-98) in CAVB-III°. 6 patients had a hemodynamic relevant secundum type ASD during follow-up.
Concomitant clinical details were hydrops (3 newborns), ventilation requirement (19 neonates) and pacemaker implantation in 33 children (69%). 11 children required a pacemaker within 2 days (mean heart rate 50), 9 in the neonatal period and 13 beyond. Initial pacemaker implants were predominantly VVI systems (78.8%) and had epicardial leads (93.9%).
Two children with CAVB-III° died, one postnatally, having hydrops, a VSD, hypertrophic cardiomyopathy and was positive for anti-SSA/Ro. The other died at 3.3 years due to cardiac strangulation by the pacemaker lead. Cardiac function was echographically impaired in 11 of 46 children (23.9%) during follow-up. Because of severe dilated cardiomyopathy, two children (one autoantibody negative) required heart transplantation. At last follow-up, 36 patients were in NYHA-I, 8 were in NYHA-II. Two were not classified.
Conclusion: The post-natal outcome of CAVB concerning functional class and the survival rate is satisfactory. 69% of the patients received a PM, for which a low heart-rate at birth was the main indication for early implantation. The prevalence of secundum type ASD was remarkably high in our study population. However, a national guideline for the follow-up of CAVB in the Netherlands is lacking, leaving room for improvement.

O1-6
Single catheter technique for radiofrequency ablation of drug-refractory tachycardias in infants

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Radiofrequency ablation is a standard technique used for definitive cure in most cases of tachycardias in old children. Number of catheters used for these procedure might be a handicap when patients are small in weight or age. Description of single catheter technique for ablation of tachycardia substrate in these small patients is presented and discussed. Results are compared to older pediatric population in a single pediatric center.
Patients, methods and results: From 1995 to 2013, 1280 cardiac ablations were performed in children under 18 years of age in a pediatric electrophysiology service. Of those, 111 procedures were performed to 101 patients weighting less than 15 kg due to drug-refractory tachycardias. All of them were done under sedation or general anesthesia. Radiofrequency energy was used in all cases. Single catheter technique was used in 85 procedures (75.2%). In the other 26 cases, only two catheters were used. Primary success rate in a single procedure was 98% of cases. Nine patients needed a second procedure for reccurency. In only one patient, affected of severe Ebstein disease, a third procedure was needed. The mean procedural time was 44.5 +/- 24.2 minutes, with a mean radiation time of 10.8 +/- 8.7 minutes. Complications were the following: one complete AV bock that completely recovered 24 hour after the procedure, 1 pericardial effusion that resolved with pericardial aspiration within the same procedure, 1 ventricular fibrillation that needed electric cardioversion with no further complications, 1 moderate mitral regurgitation freee of medication after 7 years of follow-up. These results are similar to those seen in older children in our series exept for the rate of single catheter technique that is less used in older patients (11%).
Conclusion: Ablation in small pediatric population is feasable and safe when performed in large pediatric units. Single catheter technique is an excellent option for those small patients requiring ablation.

O2-1
A 10-year study of planed delivery of foetuses with prenatally diagnosed congenital heart disease in a single institution

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Objectives: To describe the aims and rationale for planned delivery in a tertiary referral centre for foetuses with prenatal diagnosis of congenital heart disease.
Methods and results: 2130 consecutive foetuses with congenital heart disease diagnosed from January 2002 to December 2011 were included: 1258 (59%) in-born neonates whose delivery was planned in our institution, 799 (38%) terminations of pregnancy, and 73 (3%) foetal deaths. For in-born, planed delivery was classified as 'certainly warranted' for 899 (71%) for the following reasons: need for Rashkind atrioseptotomy in 344 cases, risk of aortic coarctation in 272 cases, ductal patency needed for pulmonary flow in 107 cases, ductal patency needed for systemic flow in 93 cases, need for an immediate intervention in 83 cases. For the remaining 359 in-born, planed delivery was classified as 'potentially warranted' for the following reasons: potential need for ductal patency for pulmonary flow in 156 cases, for systemic flow in 35 cases (3%), incomplete congenital heart disease diagnosis in 94 cases, need to monitor neonatal tolerance of the defect in 51 cases. In these 359 in-born at risk, rationale for planed delivery was reviewed after birth. A posteriori, in-born delivery was not necessary for 249 in-born (20%) in whom no intervention was needed during the first week, and confirmed to be necessary for 110 in-born (9%) -32 in whom diagnosis was different with a direct influence on management and with 78 who needed an intervention during the first week.
The median follow up was 3 years and 318 days. The initial announced therapeutic program was achieved at last news for 95,5% of in-born neonates (after excluding death).
Conclusions: Our study demonstrates that only a fifth of foetal congenital heart diseases delivered in a tertiary reference centre appears to be unnecessary. Conversely, a third of in-born with only potential post-natal risk of cardiac complication were appropriately delivered in our institution, as they needed immediate care in an expert centre.

O2-2
Prenatal detection of transposition of the great arteries affects mortality and morbidity

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Introduction: Transposition of the great arteries (TGA) is the most common cyanotic congenital heart defect requiring immediate treatment after birth. TGA has an excellent long-term outcome, after arterial switch operation. The pre-operative mortality is, unfortunately, 4 to 6%. A prenatal diagnosis reduces neonatal mortality. TGA is, however, commonly missed during prenatal screening. The study aim was to evaluate the prenatal detection rate of TGA and the effects of prenatal detection on pre- and post-surgical mortality and morbidity.
Methods: In a retrospective cohort study all infants with TGA with intact intraventricular septum or non-significant ventricular septal defect born in a large referral region between 1-1-2002 and 1-1-2012 were included. The cases were divided in two groups: with or without prenatal diagnosis. Pre-surgical morbidity was assessed in terms of; (1) significant cardiovascular compromise, (defined as resuscitation with inotropics and/or oxygen saturation lower than 61%), (2) metabolic acidosis, (pH $<$ 7.1 and/or lactate $>$ 5.0 mmol/l), (3) multi-organ failure, (abnormal renal and/or hepatic function), (4) closure of the arterial duct before initiation of prostaglandin therapy. Pre-and post-surgical mortality was assessed, follow-up was one year.
Results: Of all cases (n = 103) 25% were prenatally diagnosed, with 44% prenatal detection in the last five years. All deaths occurred in cases with a postnatal diagnosis (9.1%, with 4.9% pre-surgical mortality). The incidence of closure of the arterial duct before initiation of prostaglandin treatment and renal failure was significantly more common in the postnatal compared to the prenatal group (0% versus 19.7%, p = 0.002 and 0.06% versus 21.7%, p = 0.021). Other variables indicating morbidity showed a non-significant trend in favour of prenatal diagnosis (mean ASAT 204.5 versus 60.9, ALAT 90.2 versus 38.7).
Conclusions: This study presents one of the largest cohorts of neonates with TGA assessing morbidity to this extent. First-year mortality is significantly decreased by prenatal diagnosis from 9.1% to 0%, showing that despite modern resuscitation and intensive care for neonates with TGA, a prenatal diagnosis is the best prevention for mortality. Moreover, some morbidity indicators were significantly higher in the postnatal group. These results justify all efforts to improve prenatal screening programs.

O2-3
Characteristics and outcomes of fetuses with laterality defects. Comparison of the newer outcomes data with the more remote ones

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Characteristics and outcomes of fetuses with laterality defects. Comparison of the newer outcomes data with more remote ones

Background: Congenital heart defects (CHD) associated to laterality defects or heterotaxy syndromes have potentially a poor prognosis. Aim of this study was to analyse whether the outcomes of these defects diagnosed in fetal life modified in more recent years with respect to the previous era.
Material and methodology: Out of around 7000 fetuses at risk for CHD examined between 1995 and Dec. 2013 by echocardiography, 1320 had CHD and 81(6.1%) presented an abnormal visceroatrial situs: 42 in the period till Dec. 2002 and 39 afterthen. In total - 19 had left isomerism (Lisom), 21 right isomerism (Risom), 12 - situs inversus with dextrocadia (SVI-dx), 6 - SVI with levocardia (SVI-levo) and 24 situs solitus with dextrocardia (SVS-dx). Anatomical features of the fetal heart, association with chromosomal or extracardiac anomalies (CA, ECA), course in utero and after birth were analysed retrospectively from the data base and clinical documentation. The mortality results in two periods were compared.
Results: Thirteen cases (6,8%) showed normal intracardiac anatomy: 5/12 SVI-dx, 2/6 SVI-levo, 5/24 SVS-dx and 1 Lisom with an isolated anomaly of the systemic venous return. Out of the remaining 68 cases with CHD, the most frequent was complex atrioventricular defect (25 cases) - in Lisom, Risom, SVI-dx and SVSdx, with atrioventricular block (AVB) in 3, complex DORV and UVH, with AVB in one. Other fetuses had corrected TGA, VSD, pulmonary atresia + VSD, tricuspid atresia, truncus, coarctation, partial pulmonary venous drainage and DOLV. Six had associated chromosomal or extracardiac anomalies (in SVI-dx, SVI-levo, SVS-dx and Lisom).
Outcome: 20/68 cases (29.4%) opted for the termination of pregnancy, 4 died in utero, 8 after birth spontaneously and 10 after operation or pacemaker implant. Total mortality in all cases with CHD continuing pregnancy was 22 (45.8%) Comparing the 1st and the 2nd period it was 50% vs. 28.6% in SVS-dx, 25% vs. 45.4% in Risom and 71.4% vs. 33.3% in Lisom.
Conclusions: Our fetal cases with laterality defects and CHD presented a relevant mortality, in both periods, mainly in cases with isomerisms and SVS-dx. The outcomes seem slightly better in more recent period, despite the limits due to the small numbers.

O2-4
Isolated AV-Block in the Fetus. A Dutch Retrospective Analysis

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Introduction: As there is no consensus concerning the follow-up and treatment of congenital atrioventricular block (CAVB) the prenatal course of CAVB in the Netherlands of the last decade was compiled.
Methods: A nationwide database of all Dutch paediatric cardiac centres was initiated to retrospectively analyse the outcome of fetuses with a CAVB II°/III° from 2003 to 2013. Patients with complex congenital heart disease, LQTS or chromosomal abnormalities were excluded.
Results: 56 fetuses were indentified. 28 fetuses were initially diagnosed with CAVB-II° at a gestational age (GA) of 23.3 (17.6-41.2) weeks and mean heart-rate of 71 bpm (51-100). 27 were initially diagnosed with CAVB-III° at a GA of 22.6 weeks (range, 17.2-36.4) with a mean heart-rate of 57 bpm (30-71). 1 fetus progressed from CAVB-I° to CAVB-III°. 16 fetuses progressed from CAVB-II° to CAVB-III° prenatally. 5 (8,8%) women opted for termination. 7 (12.3%) fetuses (CAVB-III°) died in utero. Mean heart-rate in intra-uterine deaths at diagnoses of CAVB was 55 bpm (95% CI: 43.5- 67.1) at a mean GA of 20.4 weeks (95% CI: 19.0-22.0). Live-births had a heart-rate of 66 bpm (95% CI: 61.9-69.4) and GA of 24.0 weeks (95% CI: 22.4-25.4). 3 children with AVB-II improved prior to birth.

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One - negative for maternal autoantibodies - resolved spontaneously. 2 others receiving dexamethason converted to CAVB-I° and 0 respectively.
23 (41.1%) fetuses received therapy, 7 β-mimetics, 5 steroids and 11 both. 10 fetuses had hydrops, which resolved in 3 of 5 receiving B-mimetics, 3 of 5 receiving both, whereas 2 other fetuses receiving both medications died. In total 4 treated fetuses died. At the last fetal exam 2 had hydrops, 6 had pericardial effusion. In 9,1% the cardiac function was impaired at the last fetal exam.
Conclusion: The prenatal course of CAVB shows a moderate mortality rate, which is related to earlier gestational age at diagnosis and lower heart-rates. The number of initially diagnosed CAVB II° seems high and it has to be scrutinised whether the majority was CAVB III° instead.
Our aim is now to re-evaluate prenatal diagnostic work-up and follow-up and to develop a Dutch consensus on therapy.

O2-5
Intrauterine treatment of critical pulmonary stenosis/atresia with intact septum – preliminary experience of intrauterine and postnatal course

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Critical pulmonary stenosis/atresia with intact septum (CPS/IVS, PA/IVS) remains a therapeutic challenge with significant morbidity and mortality.
Between 10/2000 and 12/2013 a total of 10 attempts of in-utero pulmonary valvuloplasties have been performed in 8 fetuses (3 PA/IVS, 5 CPS/IVS). All had hypoplastic RVs with suprasystemic RV pressures. Median TV z-score was -2,48 (-1,26–-2,94). Median gestational age at intervention was 27 + 4 weeks (range: 24 + 6–32 + 2 weeks). Technical success was achieved in 6/10 procedures or 6/8 fetuses (75%). All procedures were carried out under general anesthesia of the mother. No maternal side effects were observed. Median procedure time was 1:51 hours (range 1:04-4:00 hours). There was no fetal death, no fetal bradycardia or significant pericardial effusion. Balloons with diameters of 4,3–4,8 mm were inserted through 17G (2 fetuses) and 18 needles. In all successful cases the pulmonary valve was perforated with the sharp needle before inserting the catheter. After successful intervention there was always a better (longer and biphasic) RV filling. A high gradient across the pulmonary valve remained in 5/6 fetuses with increased flow through the RV, in 1 case there was severe pulmonary regurgitation with almost no residual gradient. Continuous, but slower than normal growth of TV and RV was observed in all successfully treated fetuses. After birth, 5/6 newborns received a modified BT Shunt, in 1 newborn a balloon valvuloplasty alone was sufficient and still is at the age of 3 years. In 3 children the BT Shunt was successfully removed at 8 months of age, 1 patient received a Glenn Shunt (1,5 ventricle repair) and 1 BT shunt is still in place 6 months after surgery. At a median follow-up time of 3,8 years (0,6–13,2), 4/6 children are biventricular with acceptable RV size and function.
Conclusions: in-utero pulmonary valvuloplasties are technically more challenging, but seem to be better tolerated than LV interventions. Successful intervention resulted in improved flow through the right heart, which might has lead to better RV growth and function at birth. Postnatal transient RV support with a systemic to pulmonary artery shunt was necessary in most cases.

O2-6
Can postnatal aortic arch morphology predict neonatal coarctation of the aorta following prenatal diagnosis?

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Introduction: Antenatal diagnosis of coarctation is associated with false positive diagnoses. Postnatal assessment involves an extended neonatal hospital admission whilst awaiting ductal closure to confirm or refute a coarctation. In fetuses with an antenatal suspicion of coarctation, can the first postnatal echocardiogram predict which patients will develop neonatal coarctation?
Methods: The last 37 cases of prenatally suspected coarctation with normal connections diagnosed at our institution were reviewed. Neonatal echocardiograms performed within 24 hours of birth were analysed. Measurements of the aortic arch at end-systole were taken. The distal arch index (ratio of distance between left common carotid artery to left subclavian artery/distal arch diameter prior to the left subclavian artery) was calculated. Measurements were made blinded to the neonatal outcome. Published z scores (Detroit) based on weight were used. Mann Whitney and Fisher's exact test were used for analyses.

Table 1. Measurements of cardiac structures in neonates who develop coarctation following an antenatal diagnosis of coarctation. Values are expressed as median (range). * indicates $p < 0.05$.

Echocardiographic feature	Coarctation	Normal	*P* value
Presence of ventricular septal defect	9/14	7/23	*0.086*
Bicuspid aortic valve	5/14	2/23	*0.080*
Common origin of innominate and left common carotid artery	6/14	13/23	*0.508*
Aortic valve z score	-2.2 (-5.9 to +1.2)	-1.9 (-4.3 to +1.1)	*0.598*
Transverse arch diameter (mm)	3.3 (2.9–3.0)*	4.7 (3.6–6.2)	*< 0.0001*
Transverse arch z score	-5.7 (-6.7 to -4.1)*	-3.4 (-5.3 to -1.6)*	*< 0.0001*
Arch diameter at left common carotid artery/isthmus	1.2 (0.7 – 2.1)	1.1 (0.5–1.4)	*0.251*
Distal arch index	2.2 (0.8 – 3.4)*	0.9 (0 – 4.4)	*0.0005*
Isthmus diameter (mm)	2.2 (1.7 – 4.4)*	3.6 (1.9–6.8)	*0.001*
Isthmus z score	-5.6 (-7.4 to -1.3)*	-2.3 (-6.2 to +1.8)	*0.001*
Duct diameter (mm)	5.0 (3.1 – 6.3)	4.7 (2.0 – 7.3)	*0.633*
Duct/isthmus	1.9 (1.1 – 3.2)*	1.2 (0.4–2.5)	*0.006*

Results: 14/37 neonates developed neonatal coarctation requiring surgical repair. The arterial duct was patent in all patients at the time of echocardiogram. Mean weight was similar in in the coarctation and non-coarctation groups ($p = 0.975$). There was no significant association of bicuspid aortic valve or ventricular septal defect with development of coarctation ($p = 0.080$; $p = 0.086$, respectively).
The median transverse arch and isthmus z scores were below the normal range in both the coarctation and non-coarctation groups. However, transverse arch diameter, transverse arch z score, isthmus diameter and isthmal z scores were significantly smaller in neonates who developed coarctation. The distal arch index and duct:isthmus ratio were significantly larger in those who developed coarctation. Table 1 denotes p values.
A distal arch index of greater than 1.5 is associated with coarctation ($p = 0.001$), relative risk: 6 (95% CI: 2–17). Sensitivity: 73%, specificity: 87%. A transverse arch z score of less than -4 is associated with development of coarctation ($p = 0.0001$). Sensitivity for coarctation: 100%, specificity: 83%.
Conclusions: As expected, in these groups of neonates, the arch measurements are below the normal range. However, they are significantly smaller in those who develop neonatal coarctation. A distal arch index >1.5 and transverse arch z score <-4 should further raise the suspicion of development of coarctation.

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O3-1

Motor skills in children are no longer influenced by the existence of congenital heart defect

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Introduction: This study aims to compare sport motor skills of children with and without congenital heart defect (CHD). In addition, both the influence of sex and the severity of heart defect on motor skills were examined. Physical activity has cardiovascular, muscular, functional and psychological benefits on children with CHD. Without adequate exercise during childhood the patients can suffer from poor physical and mental health.
Methods: Sport motor skills of 205 children aged between 4 and 13 years were examined. 102 patients were divided into three groups depending on the severity of their heart defect (simple, moderate or great complexity). The remaining 103 healthy children served as control group. All children were tested in the areas of strength, speed, reaction time, mobility, coordination under time pressure, and balance. The test battery consisted of various parts of the German motor ability test, the Eurofit, "Motorik-Modul", "Kindergarten Mobil" and further tests using a pressure plate or the talent-diagnose-system.
Results: Sport motor performance of children with CHD cannot generally be classified as better or worse than the performance of healthy counterparts. Children with CHD dominated some reaction tests and sprint, while healthy children demonstrated their superiority in shoulder flexibility, coordination under time pressure, simple reaction and in the height of jump (all $p < .05$). Children with CHD revealed a consistent level of performance in nearly all tests regardless of the severity of their heart defect.
However, a gender-based analysis depending on health demonstrated worse performance results of girls with CHD compared to that of healthy girls, who dominated shoulder flexibility ($p = .004$), simple reaction ($p = .003$), jumping height ($p < .001$) and jumping distance ($p = .010$). Boys with CHD had the same performance level than their healthy controls. In fact, they were even better at sprint ($p < .001$).
Conclusion: Nowadays, boys with CHD seem to have caught up on healthy boys and show normal motor skills. Girls with CHD, however, lack competence in certain motor skills. Thus, recommendation of an active lifestyle seems to show positive effects. Individualized intervention, however, should be focused especially on girls. The severity of heart defect appears to have no major additional influence.

O3-2

Average-Intima-Media-Thickness and Intima-Media-Roughness as new parameters for cardiovascular health in children

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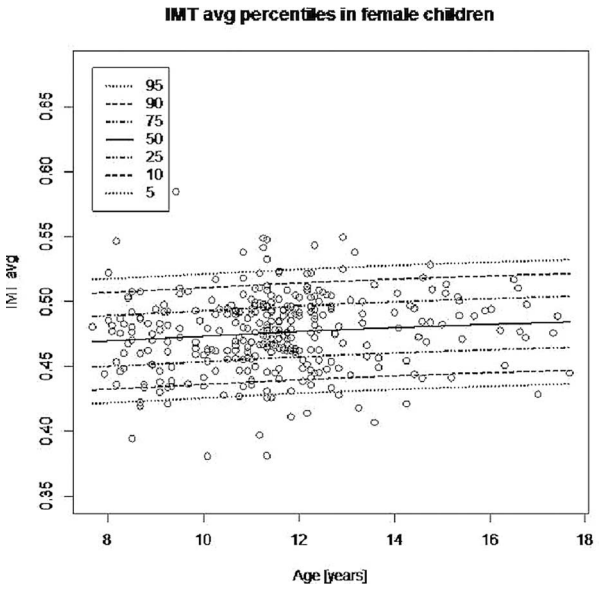
Introduction: Carotid artery intima media thickness (IMT) measurements and their interpretation in the paediatric age group represent a special challenge. On the other hand, this surrogate marker for atherosclerosis deserves special attention in counselling patients at risk for vascular complications as early as possible. Previously calculated reference values differed between studies. So, in the light of different reference data from different ultrasound systems and algorithms we are missing robust and easily applicable preventive parameters.
Methods: We calculated "average" IMT from sonographic studies of the carotid artery in 709 school children from 8–18 y. The algorithm included both end diastolic and end systolic IMT during a minimum of 3 heart cycles eliminating errors of manual tracing or indefinite measurement points. Moreover, IMT-"roughness" as a function of difference from the mean and as a surrogate for IMT irregularity was calculated as well. From these parameters, we calculated age- and sex-specific percentiles using the LMS-method.
Results: After eliminating studies from obese and hypertensive children, a total of 631 subjects were included (s. Table).

Age (years)		1 (8.0-10.9)	2 (11.0-13.9)	3 (14.0-17.9)
n	w	111	134	59
	m	124	177	26
IMT	w	0.48 ± 0.04	0.49 ± 0.03	0.50 ± 0.04
(mm)	m	0.49 ± 0.03	0.49 ± 0.03	0.49 ± 0.04
IMR	w	0.037 ± 0.012	0.035 ± 0.010	0.036 ± 0.010
(mm)	m	0.035 ± 0.011	0.033 ± 0.010	0.038 ± 0.011

We calculated age- and sex-specific percentiles for average-IMT and IMT-roughness. Also, tables of mean of avg-IMT and IMT-roughness were calculated reporting the L-, M- and S-value and allowing for calculation of z-scores from absolute parameters.
Conclusion: Given the differences of actual IMT-normative values, we propose a more sophisticated calculation of IMT including diameters at end systole and end diastole. As these diameters are detected with an automated contour edge detection system and calculated from several measurements at different time points, they may represent more comparable surrogate markers for the "real" intima media thickness of the carotid artery. IMT-"roughness" may add valuable informations about the structure of the inner layer of the endothelium. Also, by using z-scores of both average-IMT and IMT-roughness, measurement results from different ultrasound systems and from different IMT measurement algorithms should be comparable throughout different studies.



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O3-3

Congenital Heart Disease is Associated with the Development of Diabetes Mellitus: A Nationwide Study

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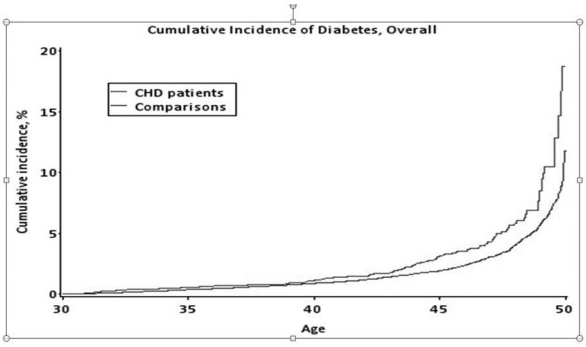
Introduction: An improved survival rate for children with congenital heart disease (CHD) has yielded a large population of adults living with CHD. It is important to understand this population's potential for acquired morbidity. The purpose of this study was to assess CHD survivors' risk of developing Diabetes Mellitus (DM) when compared to a non-CHD national cohort, and to determine risk factors that predispose CHD survivors to develop DM.
Methods: This retrospective cohort study aimed to include all Danish CHD survivors born from 1963-1984 alive at 30 years of age. CHD subjects born pre-1977 were previously identified by review of nationwide medical records. CHD subjects born between 1977-1984 were identified from the Danish National Registry of Patients (DNRP). For each CHD subject, 10 non-CHD subjects matched by sex and birth year were identified from the general population using the Danish Civil Registration System. Based on DNRP data, we computed cumulative risks and hazard ratios (HR) of time to diagnosis of DM from 1977 to 2013, and assessed for difference in the incidence of DM within the CHD cohort by specific demographic and clinical factors.
Results: We identified 5,295 CHD survivors. By 45 years of age, the cumulative risk of DM was 3% among CHD subjects (Figure). The HR of DM among CHD subjects compared to the non-CHD control population was 1.62 (95% CI: 1.33-1.97). CHD survivors born preterm had an elevated risk of DM (HR 12.58; 95% CI: 1.14-138.80), although those born term also had an increased HR (2.08; 95% CI: 1.21-3.57). Subjects with severe CHD had a HR of 2.17 (95% CI: 1.51-3.12), higher than among those with mild or moderate CHD (HR 1.45; 95% CI: 1.15-1.84). Those with cardiac surgery during the first year of life had a HR of 1.85 (95% CI: 1.06-3.22).
Conclusion: CHD survivors are at increased risk of developing DM. Adult CHD survivors born premature, with severe CHD, or requiring cardiac surgery during the first year of life were at higher risk. More research is needed to assess lifestyle elements in the CHD survivor population to determine interventions to mitigate the development of DM.



O3-4

Impact of lifestyles of children and their parents on cardiovascular risk factors in elementary school children

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Background and Objectives: Recently, the prevalence of obese children might be declining in Japan; however, longitudinal studies showed that the prevalence of obesity is still increasing during elementary school periods. Therefore, the present study aimed to evaluate the effect of lifestyles of children and their parents on the levels of cardiovascular (CV) risk factors in elementary school children.
Subjects: The study has been conducting since 2012 and announced through the local boards of education in seven areas in Japan. The study was included 976 healthy volunteers (478 boys, 498 girls) aged from 6 to 12 years with a medical examination and a questionnaire. The medical examination included the measurement of height, weight, waist circumference, and blood pressures, and blood sampling for CV risk factors. The questionnaire collected data on the lifestyles of the subjects and their parents. Screen time included time spent watching TV and playing games. The subjects were asked to walk with pedometer for 7 days. Obesity in the present study was defined using the age- and sex-specific International Obesity Task Force standard.
Results: Multivariate regression analyses showed that number of steps by pedometer measurement, screen time, sleeping time, and parental BMI were significantly and independently associated with the levels of one or more CV risk factors in elementary school children. Among these, screen time had a profound adverse effect on CV risk factor levels. Number of steps was positively associated with sleeping time and negatively associated with screen time. Screen time in children was strongly associated with parental screen time. The risk of obesity in boys was associated with paternal obesity, but not with maternal obesity. On the other hand, the risk of obesity in girls was associated with both paternal and maternal obesity.
Conclusions: Increase in number of steps and sleeping time and decrease in screen time may be the first-line approach for elementary school children to maintain favorable CV risk factor levels. An association between paternal or maternal obesity and obesity differs between genders in Japan; thus, approaches focusing on parents should take the gender of children into consideration.

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O3-5

Fitness is not associated with carotid intima media thickness in children

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Introduction: Carotid intima media thickness (cIMT) and cardiorespiratory fitness (CRF) are independently used as predictors of cardiovascular health. We combined these measures in a pediatric population to examine if fitness is associated with vascular status. Furthermore, we calculated cIMT reference values for children aged 7-17 years.
Methods: 1017 children, aged 7-17, from Bavaria, Germany, were prospectively studied. cIMT was assessed by B-mode ultrasonography (Hitachi Aloka prosound α6) in 736 children (330 boys/ 406 girls) and calculated as average of two measurements of the left and right common carotid artery according to the Mannheim cIMT consensus [1]. Measurements were performed by two examiners (coefficient of variation: 8.51%). CRF was determined by estimation of VO2max with the PACER test [2]. Further measures consisted of blood pressure, body weight and height, hip and waist circumference. Age- and height-normalized cIMT reference values were calculated for 690 non-obese children (310 boys/ 380 girls) applying the LMS method [3]. The study was funded by "Sternstunden e.V. and Landratsamt Berchtesgadener Land".
Results: cIMT increased with height, weight, hip circumference, systolic blood pressure, body mass index and age ($p < .001$). There was no significant correlation between cIMT and VO2max. Multiple regression analysis associated a small waist circumference with low cIMT (standardized $\beta = -.666$), the heavier and taller children were, the thicker cIMT. Boys within the youngest and oldest age categories (8-9.99 and 16-17.99 years) had significantly higher cIMT values than girls ($p < .05$). No sex differences occurred in children from 10-15.99 years.
Conclusions: In this study, CRF is not significantly associated with cIMT. Nevertheless, it would be wrong to conclude that fitness doesn't affect cardiovascular health. As structural changes in IMT develop over a long time, the same might be for positive effects due to good CRF. Parameters of arterial stiffness (pulse wave velocity, augmentation or stiffness index β) may respond faster and will therefore be analyzed in further studies.
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O3-6

Temporal Trends of Increasing Adiposity are Associated with Diminished Exercise Capacity in Children with Repaired Congenital Heart Disease

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Introduction: Children with repaired congenital heart disease are often less physically active than their healthy peers and, hence, at risk for obesity. We sought to determine the trends over time in adiposity and its association with exercise capacity.
Methods: n = 725 patients were included in this study and 4,153 height and weight measurements were available for analysis (average 6 ± 5 measurements per patient); 193 (27%) patients were followed for longer than 10 years. Underlying cardiac diagnoses included ASD 18%, VSD 21%, AVSD 16%, TGA 21%, TOF/DORV 18% and Fontan 10%. All analyses were performed using regression models adjusted for age, gender, repair status and repeated measures through an autoregressive covariance structure. Body mass index (BMI) and height z-scores could be calculated only for children >2 years old.
Results: Weight z-score at diagnosis was -1.1 (5th/95th percentile: -3.6; +1.1) and increased over time (+0.103 (0.007) SD/year, $p < 0.001$). At the age of 2 years old, BMI z-score was -0.2 (-2.6; +1.9) and increased over time (+0.042 (0.007) SD/year, $p < 0.001$) while height z-score was -0.3 (-2.7; +1.4) and did not change over time (-0.007 (0.006) SD/year, $p = 0.18$). A total of 45% of patients had at least one BMI measurement above percentile cut-points for overweight (28%) or obesity (17%). The proportion of overweight and obese children increased over time (OR: 1.09 (1.01-1.17) per 3 years, $p = 0.02$). A total of 153 exercise tests in 101 (14%) patients were reviewed. Overweight or obese patients had lower percent predicted maximum VO2 (-15.5 (2.4)%, $p < 0.001$), higher peak systolic blood pressure (+11 (4)mmHg, $p = 0.002$) and higher systolic blood pressure response (+7 (3)mmHg, $p = 0.01$) than those patients with BMI below the 85th percentile (normal weight). Overweight/obesity was not associated with percent predicted maximum heart rate (EST: +2 (2)%, $p = 0.35$).
Conclusions: Children with repaired congenital heart disease have an important risk of overweight and obesity, with a trend towards increasing adiposity with age, and an association with lower exercise capacity and higher blood pressure. Programs and counseling aimed at achieving healthy lifestyles and a reduction in adiposity are necessary.

O4-1

Model of hypoplastic left heart in the fetal lamb created using a percutaneous transhepatic technique - preliminary experience

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Introduction: Reduced blood flow into the fetal left ventricle (LV) may underlie development of Hypoplastic Left Heart Syndrome (HLHS), for which palliative surgery is the only treatment option. Currently there is no HLHS model in a large fetal mammalian animal for testing and clinical translation of new therapy. We have recently published a percutaneous trans-hepatic technique to catheterise the fetal heart, which could be utilised to produce an animal model of HLHS. Using a percutaneous trans-hepatic technique to deliver an occluder into the fetal heart, we aimed to create a fetal lamb model of HLHS by occluding the foramen ovale (FO).
Methods: Three fetal lambs (110-117days, term = 147days) under maternal general anaesthesia were used. Under ultrasound guidance, the fetal hepatic vein was percutaneously punctured through the maternal abdomen using a 14GA IV-cannula. A coronary catheter and guidewire were inserted and advanced into

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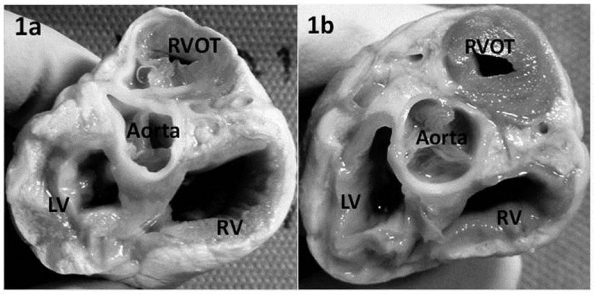
the fetal IVC and right atrium. An Amplatzer Duct Occluder II (ADO II 4 × 4 mm, St Jude Medical) was delivered to occlude the fetal FO. In 2 fetuses, a self-expandable stent (8 × 12 mm Superflex-DS 4Fr deliverable stent, Opti-Med Inc. Germany) was positioned across the FO first, and the ADO II was anchored within the stent. Serial ultrasounds were performed to monitor cardiac development. Euthanasia and post-mortem examination was performed 3 weeks after. A twin fetus which did not undergo the procedure was used as control for comparison. Morphometric measurements were made on digital images (Image J1.48b, NIH) of transverse sections of the heart at the level of atrioventricular valves.



Results: All 3 fetal lambs survived well to 3 weeks without any fetal compromise. At post-mortem examination, the LV lumen was markedly reduced (Fig 1a) with lower LV/right ventricular (RV) chamber volume ratio, LV/RV lumen area ratio and increased septal thickness at level of atrioventricular valves (Table), compared with the control fetal heart (Fig 1b).
Conclusions: Occlusion of the fetal FO using percutaneous catheterisation leads to phenotype simulating HLHS. Our results demonstrate the potential to develop the world's first large animal model of HLHS, critical for understanding the cardiac and pulmonary consequences and devising new therapies.

Fetal hearts	Ratio of heart weight/body weight (g/kg)	Ratio of LV/RV chamber volume	Ratio of LV/RV lumen area	Septal thickness (cm)
Occluded FO 1	3.7	0.63	0.79,	0.44
Occluded FO 2	4.5	0.30	0.88	0.45
Occluded FO 3	4.7	0.23	1.02	0.53
Control (n = 1)	4.1	1.0	1.54	0.23

O4-2
Pulmonary artery growth in congenital heart disease with duct-dependent pulmonary circulation: role of arterial duct stenting in hypoplastic pulmonary artery tree

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Background: Arterial duct (AD) stenting is increasingly deemed a reliable alternative to surgical shunt in congenital heart disease with duct-dependent pulmonary circulation (CHD-DPC). Allowing stent to conform to pulmonary artery (PA) anatomy might promote significant and uniform growth of the pulmonary vascular tree. Aim of this study was to assess the PA growth resulting from AD stenting in CHD-DPC with very hypoplastic PAs at the time of initial palliation.
Methods: Thirty-six of the 111 neonates submitted to AD stenting as palliation of CHD-DPC at our Institution between 04/2003 and 12/2013, underwent control catheterization in view of surgical repair. Global PA growth was evaluated as Nakata Index increase, while left and right PA growth was assessed as individual z-score changes. Finally, left-to-right PA diameter ratio was considered indicative of balanced PA growth. All data were assessed both in the whole population and in two subgroups identified on the basis of the PA size at the time of duct stabilization (Group I, n = 12, Nakata Index <100 mm2/m2) and (Group II, n = 24, Nakata Index >100 mm2/m2).
Results: AD stenting was performed using coronary stents dilated to 3.6 + 1.3 mm (median 3.4). Control angiography was performed 6.2 ± 3.4 months (median 5) after stenting, showing significant and balanced PA growth (Nakata Index increase from 148.6 ± 76.2 to 273.0 ± 92.4 mm2/m2, +113 ± 99%, $p < 0.0001$; LPA z-score from -0.57 ± 1.5 to 1.23 ± 1.47; RPA z-score from -0.61 ± 1.3 to 1.19 ± 1.3, $p < 0.001$ for both comparisons), without significant changes of the left-to-right PA diameter ratio (from 0.99 ± 0.18 to 0.92 ± 0.31, p = NS). The Group I showed a greater increase of both global PA growth (Nakata Index increase 194 ± 115 vs 75 ± 61%, $p < 0.001$) and individual PA z-scores (LPA z-score 106 ± 51% vs 78 ± 51%, p = NS; RPA z-score 99 ± 44 vs 95 ± 57%, $p < 0.05$) compared to the Group II. At control angiography, final PA size did not significantly differ between groups (241.6 ± 106.5 vs 285.2 ± 84.5 mm2/m2, p = NS).
Conclusions: Percutaneous AD stenting is highly effective in promoting significant and balanced PA growth in CHD-DPC. This approach seems to be even more effective in neonates with severe PA hypoplasia at the time of duct stabilization, thereby being highly advisable in this subset of patients compared to surgical palliation.

O4-3
PRDM16 is a possible therapeutic target for heart failure

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Introduction: Individuals with del1p36 syndrome commonly have cardiomyopathy and/or structural heart defects. We have recently described the role of the transcription factor PRDM16 (PR domain containing 16) in cardiomyopathy associated with monosomy 1p36, and confirmed its relevance in non-syndromic left ventricular noncompaction cardiomyopathy (LVNC) and dilated cardiomyopathy (DCM). PRDM16 has not previously been associated with cardiac disease.
Results: We identified a minimal deletion for cardiomyopathy associated with del1p36 that included only PRDM16. Resequencing of PRDM16 in non-syndromic patients with LVNC detected de novo mutations. In addition, in a series of cardiac biopsies from individuals with DCM, we found previously unreported non-synonymous variants in the coding region of PRDM16. Modeling of PRDM16 haploinsufficiency and a human truncation mutant in zebrafish resulted in both contractile dysfunction and partial uncoupling of cardiomyocytes, and also revealed evidence of impaired cardiomyocyte proliferative capacity. The cardiac phenotype in the zebrafish mutants can be completely rescued by the application of 5 structurally related compounds. Wildtype zebrafish demonstrate a significant increase in cardiomyocyte numbers after treatment with the

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compounds suggesting a pro-proliferative effect of the compounds. With the zebrafish model system we are currently identifying the role of PRDM16 in the heart. We will also present our approach on the underlying PRDM16 signaling pathway using in vitro culture systems.
Conclusion: The zebrafish model will serve to identify new interaction partners for PRDM16 in the heart. The rescue of the cardiac phenotype in the zebrafish might lead to novel therapeutic targets for heart failure.

O4-4
Pressure volume relations obtained by 3D-real-time echocardiography and mini-pressure wire - multimodal validation-studies with MRI and conductance-catheter in piglets

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Introduction: Pressure-volume relations (PVR) provide vital information regarding cardiac pathophysiology and function. In children, acquiring PVR by conductance technology (Cond) is – due to catheter size - restricted to older age. The aim of the study was to compare the results of PVR - calculated from 3D-echo (3DE) volume data and simultaneously obtained pressure data with conductance technology.

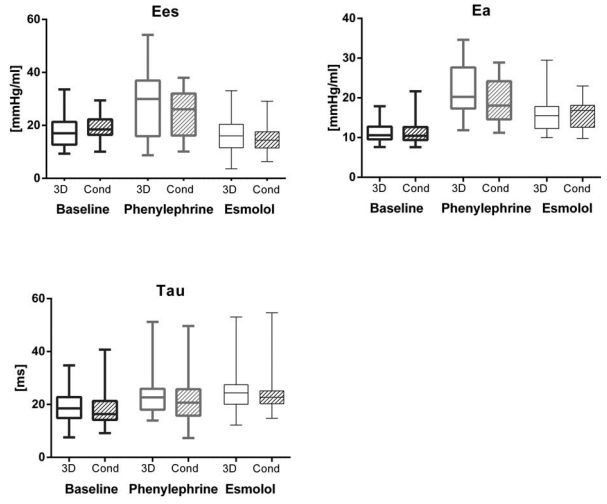

Figure: 3D-Echo (3D)-derived pressure-volume relations compared to parameters obtained by conductance technology (Cond) under various conditions (mean and 95% CI), n = 16.

Methods: In 16 piglets (weight: 3.6-8.0, mean 5.9±1.3 kg), dp/dt max, dp/dt min, systolic myocardial elastance Ees, arterial elastance Ea, diastolic relaxation constant Tau and end-diastolic PVR were determined under various conditions: baseline, epinephrine and esmolol infusion, using 3DE with simultaneous recording of ventricular pressure by a mini pressure wire (Radi, St. Judes) and compared to Cond. To validate the accuracy of 3D volume data, MRI was performed in 8 piglets.
Results: Computation of PVR was comparable to results obtained by Cond (Fig). Agreement between MRI and 3DE was good (Table). Inter- and intraobserver- coefficients of variation were below 5% for all parameters.

Bland Altman Analysis MRI vrs 3D

Parameter	Bias	95% Limits of Agreement	Parameter	Bias	95% Limits of Agreement
EDV [ml]	-0.03	-1.2; 1.1	SV [ml]	-0.01	-2.4; 2.2
ESV [ml]	-0.12	-1.1; 0.9	EF%	2.71	-5.6; 11

Conclusions: Calculation of PVR from 3DE volume curves is a feasible and reliable method to assess different conditions of cardiac function in small hearts. Due to its minimal invasive character this methodology may be implemented into daily practice and contribute to clinical decision making.

O4-5
Defining the pathogenic autoantibodies in congenital heart block

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Objectives: Maternal autoantibodies directed against the amino acids (aa) 200–239 of Ro52 (Ro52-p200) is associated with risk of foetal AV-block. The objective of this study was to identify the epitope specificity of human anti-Ro52-p200 antibodies.
Methods: Sera from 19 mothers of foetuses with CHB and 8 babies with CHB were analysed for binding to synthetic peptides representing variants of human and rodent Ro52-p200 and overlapping peptides by ELISA. Competition experiments were performed to confirm specificity and affinity of the binding. Secondary structure of the peptides was analysed by circular dichroism spectroscopy (CD).
Results: Analysis of autoantibody-binding to human p200 and a highly overlapping peptide including aa 197-232 (p197), demonstrated significantly higher reactivity towards p200 than to p197 ($p < 0.0001$). The relevance of C-terminal aa of p200 was confirmed using truncated human p200 peptides, demonstrating that deletion of aa in the C-terminal of p200 completely abolished antibody binding. Furthermore, taking advantage of the fact that human sera do not bind rat p200 (r-p200), peptides based on the rat p200-sequence, with selected crucial residues mutated into the human counterparts were generated. C-terminal mutations including a glutamic acid substitution for an aspartic acid in position 233 reestablished binding of sera, while there was no gain of reactivity by substitutions in the N-terminal or mid part of the peptide. Analysis of the peptides by CD confirmed correct folding of the peptides. Finally, we generated peptides with alanine substitutions of each residue from position 233 to 239 (pA233-pA239). Substitution of the aspartic acid 233 abolished antibody binding, while the other mutations did not affect binding. Preincubation of the sera with the rat-to-human 233 mutated peptide blocked antibody reactivity to p200 to the same degree as the p200 peptide itself.
Conclusions: Our study suggests that the aspartic acid at aa residue position 233 of the Ro52-p200 peptide is crucial in forming the main epitope of the Ro52-p200 peptide bound by CHB-related human Ro52 antibodies. This specificity might be used as a tool to identify high risk pregnancies for CHB, and for identification

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of the cross-reactive target in the fetal heart bound by the maternal autoantibodies.

O4-6
Impact of anti-HLA antibodies on survival and the development of coronary artery vasculopathy in paediatric heart transplant recipients

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Introduction: It is now well established that pre-transplant detection of donor-specific and non-donor-specific anti-HLA antibodies (DSAs/NDSAs) in heart transplant recipients is associated with a worse outcome. In adult patients, DSAs and NDSAs have been found to be associated with increased episodes of acute rejection, and more severe coronary artery vasculopathy (CAV), measured by intra-vascular ultrasound (IVUS). In paediatric recipients, DSAs have been associated with increased graft failure rates, but the link between DSAs and CAV in children has not previously been examined.
Methods: Since mid-2007, all heart transplant recipients at Great Ormond Street Hospital have been routinely screened for anti-HLA antibodies using the Luminex testing platform. Routine screening for CAV using IVUS started in late 2004. Data on antibody status and IVUS measurements, along with further clinical and demographic information, were extracted from the relevant clinical databases and collated using Excel. Using the R statistical package, the maximum intimal thickness (as measured by IVUS) at various time-points post-transplant (2-4 months, 1 year, 3-4 years) was compared in those with and without pre-transplant DSAs and NDSAs (using a threshold of 1000 mean fluorescence intensity) using Welch's two-sample t-test. Differences in survival between groups was compared using Kaplan-Meier survival curves and the log rank test.
Results: 97 paediatric heart transplant recipients had pre-transplant antibody testing using the Luminex platform. Of these, 32 patients had at least one IVUS study post-transplant. The average maximal intimal thickness at the 1 year IVUS study in recipients with DSAs was 1.17 mm versus 0.76 mm in those without DSAs ($p = 0.01$). Pre-transplant DSAs did not significantly affect survival, but the presence of NDSAs was associated with increased post-transplant mortality ($p = 0.03$).
Discussion: CAV is a leading cause of death and graft failure in paediatric heart transplant recipients. These results indicate that even very low levels of pre-transplant DSA may lead to more severe CAV compared to recipients without DSAs, and that pre-transplant NDSAs are associated with increased post-transplant mortality. Our sample size is currently small, and more advanced analyses examining the role of DSAs and NDSAs in post-transplant outcome will be possible as our cohort grows over time.

O5-1
Prognostic Variables and Their Powers Change Over Time in Fontan Survivors

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Introduction: Unique impaired hemodynamics and exercise capacity may predict morbidity and mortality in Fontan survivors (F). However, the prognostic power (PP) may change over time in the long-term postoperative course in those patients.
Objectives: To clarify whether the PPs of major hemodynamic variables, brain natriuretic peptide (BNP), and exercise capacity change during the long-term follow-up.
Methods and Results: In 362 F, we have undergone catheterizations at postoperative years of 1 ($n = 362$, group 1), 5 ($n = 280$, group 2), 10 ($n = 193$, group 3), and 15 ($n = 112$, group 4), and compared the hemodynamics (central venous pressure: CVP, cardiac index: CI, ventricular ejection fraction, and arterial oxygen saturation: Sat), BNP, and peak oxygen uptake (PVO2) with clinical events that required unscheduled hospitalization (USH), including all-cause death, and calculated the all hazard ratios (HR) as an index of PP. We encountered 128, 78, 61, and 30 USHs in the groups of 1, 2, 3, and 4, respectively. The HRs for all variables in all groups were as follows: CVP (1.13**, 1.18**, 1.22**, 1.08), CI (0.81, 0.59**, 0.49**, 1.02), EF (0.99, 0.97*, 0.97*, 1.00), SaO2 (0.94**, 0.90**, 0.92**, 0.93), PVO2 (0.97, 0.95**, 0.94**, 0.94**), BNP (1.02**, 1.07**, 1.08**, 1.10**) (*: $p < 0.05$, **: $p < 0.01$). Those results indicated that CVP and Sat had strong PP in the early postoperative phase, however, the PPs disappeared in the late phase. BNP had significant PP during the entire postoperative phase. In contrast, PVO2 had a stronger PP in the late postoperative phase.
Conclusions: Prognostic variables and the PPs significantly change over time in F. Our results indicate that, after intensive management strategies for impaired hemodynamics in the early postoperative period, additional interventions to improve physical fitness may be required for the better long-term outcome.

O5-2
Cellular immunodeficiency in patients with Failing-Fontan

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Introduction: 3-15% of all Fontan-patients develop a failing of their Fontan circulation with manifestation of a protein-loosing enteropathy (PLE). Clinically a PLE includes hypoalbuminemia, hypogammaglobulinemia and dysregulation of the salt water homeostasis. According to low IgG levels, an altered risk of bacterial infections is expected but can not be observed. Instead they show an increased risk or vulnerability of acute and chronic viral and fungal infections.
Methods: Venous blood was obtained from Failing-Fontan ($n = 10$) and Non-Failing patients ($n = 15$) and a control group of patients with biventricular heart (after thymectomy during cardiac surgery) ($n = 20$) at different time points. The control group minimized the predescribed effect of temporary T-cell depletion after thymectomy. Absolute and relative count of T-cells (CD3+, CD4+, CD8+), natural killer cells (CD16, CD56+, CD3-) and B-cells (CD19+) were determined using flow cytometry. Duodenal and colorectal biopsies of Failing-Fontan patients with PLE ($n = 2$) who received a coloscopy and gastroscopy were analysed by immunohistochemistry to differentiate between B- and T-cell infiltration.

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Results: Patients with Failing-Fontan had significant ($p < 0.01$) lower absolute and relative T-cell subsets of CD3+, CD4+ and CD8+ cells than Non-Failing patients and the control group. Absolute count of CD45 lymphocytes was significantly ($p < 0.01$) reduced in Failing-patients. Relative count of natural killer cells was significantly ($p < 0.05$) higher in Failing-patients. Failing patients showed a significant ($p < 0.05$) higher B-cell count.
The immunohistochemistry analysis of the duodenal and colorectal biopsies showed an increased CD3+ T-cell infiltration in the descending colon and duodenum.
Conclusions: For the first time we describe a cellular immunodeficiency in patients with Failing-Fontan with an extensive T-cell depletion. This can explain the clinical vulnerability against recurrent viral and fungal infections. Based on our results frequent substitution of immunoglobulins does not seem reasonable since the B-cell lineage is increased and frequent bacterial infections are not present in these patients. Long term effects on immunomodulation due to chronic immunoglobulin substitution can not be estimated. The alteration of T-cells in the descending colon and duodenum might represent a migration of T-cells from peripheral blood into the gastrointestinal wall. Underlying pathophysiological mechanisms of T-cell depletion in Failing patients need to be further analyzed.

O5-3
Paediatric Heart Failure from Heart Muscle Disease in the UK and Ireland: 5+ Year Follow Up

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Introduction: Our original study, the first national prospective study of new-onset heart failure from heart muscle disease in children, showed an incidence of 0.87/100,000 with an overall 1-year survival of 82%, and an event (death or transplantation)-free survival of 66%. (1) This study aimed to evaluate 5+ year outcomes of this important cohort of patients.
Methods: All centres in the UK and Ireland with event-free survivors from the original study participated (n = 14). Consent for inclusion was sought from patients & families. Anonymised data based on the last hospital attendance and echocardiograms (where available) were forwarded for analysis. The investigator (RA) was blinded to outcome at the time of echo review.
Results: Of 69 event-free survivors at 1 year, data was obtained on 64, with 3 lost to follow-up & 2 moved abroad. Results of investigations or further clinical developments resulted in a changed diagnosis in 16%. There were 3 deaths at 2.2, 3.3 & 9.0 years after presentation and 1 transplant, at 5.2 years. Overall 5-year survival was 79% and event-free survival 63%; or 78% and 61% respectively at last follow-up. For the 60 event-free survivors, median(range) follow-up duration was 9.04(5.0-10.33) years for those still under review (n = 46), or time to discharge 5.13(0.67-10.0) years (n = 14). Fifty-eight were in NYHA Class 1, and two in Class 2. Twenty-seven patients (45%) required one or more readmissions to hospital. On echocardiogram, 41/60 had normal studies at last follow-up. Fourteen cases had mild functional impairment, and one moderate. Mitral regurgitation was mild in 8 and moderate in 1. No intracardiac thrombi were seen. Twenty-three patients remained on medication including ACE inhibitors(n = 22), β-blockers(n = 13 of which 9 on carvedilol), diuretics(n = 2), digoxin(n = 3), aspirin(n = 3), losartan(n = 1), & flecainide(n = 1).
Conclusions: Children who survive the first year following a diagnosis of heart failure from heart muscle disease have a good longer-term outcome and quality of life although there remains a small attrition rate. Good heart failure management remains important in those with persisting echocardiographic abnormalities or symptoms.

Reference:
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O5-4
Reversible Pulmonary Artery Banding for Left Ventricular-DCM with preserved RV function: effect on BNP biomarker and MRI-imaging

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Background: dilated cardiomyopathy (DCM) in childhood has a considerable morbidity, mortality and high incidence of heart transplantation (HTX). Recently we published our initial experience with reversible pulmonary artery banding (rPAB) as an additional strategy in children with LV-DCM and preserved-RV-function instead HTX. Purpose of the current paper is to demonstrate the course of BNP-serum levels with focus on the pre-PAB medical treatment effect as well as MRI-data during further follow-up.
Methods: retrospective single center observational study. Since April 2006 24 children with LV-DCM (age <3 y) were treated by rPAB, thereof 18 without associated open heart surgery. Anti-congestive/anti-remodeling treatment with highly specific long acting β1-blocker Bisoprolol, with angiotensin-converting-enzyme inhibitor Lisinopril (goal dosages 0.1-0.2 mg/kg/d) and spironolactone (2 mg/kg/d) represent cornerstones of pharmacotherapy; optionally digitalis was used for heart rate modulation. Except for present lung edema furosemide therapy was abandoned. Milrinone (1 μg/kg/min) and levosimendan (0.1-0.2 μg/kg/min) were administered routinely one day before rPAB; milrinone was post-operatively stopped when oral anticongestive therapy was re-established for-long term treatment. Laboratory monitoring on the basis of BNP-serum levels was routinely performed pre- and post-operatively and continued in long-term follow-up. A Siemens 3-thesla cardiac machine performed MRI imaging during pre- and post-operative follow-up.
Results: there was no hospital mortality. Except for three cases all patients show clinical improvement. The pressure gradient across the PAB increased significantly correlating with the RV-function. The MRI-LV-EF increased from a median of 15% pre-PAB to 43% at discharge home and to 47% 3-6 months postoperatively accompanied by functional class improvement ($p > 0.001$); the median LVEDD/Z-score decreased ($p > 0.001$) respectively. Plasma BNP-levels dropped significantly after establishing the initial anti-congestive therapy with ß-blocker and ACE-inhibitor; immediately after surgical rPAB BNP-levels rose to their peak-level to decline again continously in all rPAB-responders, even partly to BNP-levels of health population, if transcatheter de-banding was successful.
Conclusions: rPAB improves LV-DCM if reactive hypertrophy of right ventricle can be observed. The amelioration of ventriculo-ventricular interaction can be best followed by MRI-imaging. The clinical course of patients with rPAB in LV-DCM correlates with BNP-levels.

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O5-5

First experience of Tolvaptan therapy in patients with Failing-Fontan and diuretic resistance

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Introduction: Up to 15% of all Fontan patients develop a so-called failing of the Fontan circulation. Symptoms are protein-losing enteropathy (PLE), which results in hypalbuminemia, hypogammaglobulinemia, lymphopenia, profound diarrhea, dystrophy and severe imbalance of the salt water homeostasis with serum-hyponatremia (negative predictor of morbidity and mortality in heart failure) pleural and pericardial effusions, ascites and edema. These patients need high doses of three- to fourfold diuretic therapy and electrolyte substitution. We report about our first experience of Tolvaptan treatment in patients with Failing-Fontan. Tolvaptan is a vasopressin receptor antagonist and has been demonstrated to be effective in patients with heart failure refractory to diuretic therapy.
Methods: We treated four patients with Failing Fontan, three with PLE and one patient who suffered from congestive heart failure of his single ventricle. All patients showed signs of water overload with hyponatremia (<135 mmol/l), effusions and edema. Before and during Tolvaptan therapy we monitored laboratory parameters (electrolytes, neurohumoral parameters, liver enzymes, urinary sodium and osmolality) and clinical symptoms (weight, effusions, edema, abdominal girth, etc.). In one patient with PLE we performed serial experimental 23Na-MRI (before and during therapy), to illustrate and quantify interstitial sodium and water in muscle and skin.
Results: During Tolvaptan therapy all patients showed significant reduction of weight, pleural effusions, edema and ascites. Serum-sodium stabilized under Tolvaptan in the normal range (135-145 mmol/l). Tolvaptan reduced hepatical congestion and significantly decreased liver enzymes. Conventional three to fourfold diuretic therapy and electrolyte substitution could be considerably reduced under Tolvaptan therapy and therefore side effects of chronic conventional diuretic therapy. These effects were confirmed in long-term follow up.
23Na-MRI showed a significant sodium overload in muscle and skin before Tolvaptan treatment which could be reduced and normalized under therapy.
Conclusions: Tolvaptan improves clinical state of health and symptoms of water overload in patients with failing Fontan. It reduces effusions, ascites, peripheral edema and normalized serum sodium and therefore eliminated the negative predictor of hyponatremia. Its value as rescue diuretic in patients with Failing Fontan, with water overload and diuretic resistance need to be discussed and further evaluated.

O5-6

Usefulness of three-dimensional echocardiography in the evaluation of intraventricular dyssynchrony in children with refractory heart failure secondary to dilated cardiomyopathy and its correlation with QRS morphology and duration

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Introduction and objective: Intraventricular dyssynchrony is common in patients with dilated cardiomyopathy (DCM). Adults with DCM and QRS duration >120 msec may benefit from cardiac resynchronization therapy (CRT), but children with heart failure (HF) caused by MCD often show a narrow QRS. The aim of the study was to evaluate intraventricular dyssynchrony with 3D echocardiography in children with DCM and analyze its correlation with QRS morphology and duration.
Methods: Transversal, observational, and analytical study. Echocardiographic studies were performed using a Philips IE 33 echocardiography system.
Results: 10 patients were studied. Six women. Age: 81.6 ± 61 months. Four children were in functional class III, five in class II and in one in class IV. LVEF was 8-49%, QRS duration 93 ± 31.5 msec. Three children had a QRS duration >120 msec. The ECG showed LBBB in 2 patients with a QRS duration of 122 and 158 msec, respectively. RBBB in addition with anterosuperior fascicle block was observed in one patient (QRS 128 ms). The asynchrony index (AI) assessed with real time three-dimensional echocardiography was 6.78 ± 3.66. The time to reach the minimum volume of the last segment was 110-670 msec. Although the three children with QRS > 120 msec showed abnormal AI (7.49, 8.24 and 3.98 respectively), these did not correspond with the largest AI. The largest AI matched with narrow QRS (AI 11.66/QRS 100 msec, AI 10.4/QRS 86 msec and AI 10.3/QRS 80 msec). The asynchrony index did not correlate with the morphology or the QRS duration but correlated with functional class.
Conclusions: The real-time three-dimensional echocardiography is a useful tool in the evaluation of intraventricular dyssynchrony in children with refractory heart failure secondary to dilated cardiomyopathy. The AI showed no correlation with the QRS duration in the surface 12-lead electrocardiogram in this small group of patients.

O6-1

The relative role of echocardiography and magnetic resonance imaging in identifying critical lesions in patients with single-ventricle physiology, prior to bidirectional cavo-pulmonary connection (BCPC)

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Introduction: This study compared the ability of simultaneous, pre-operative echocardiography and magnetic resonance imaging (MRI) to predict the need for intervention on lesions at the time of bidirectional cavopulmonary connection (BCPC) or during the 6-month postoperative period.
Methods: All patients were included who had undergone BCPC for single ventricle palliation between 2007–2012, and who had pre-operative assessment with echocardiography and MRI. The outcome measure was the need for surgical or catheter intervention on additional lesions at the time of BCPC or within 6 months. The indices assessed were: MRI and echocardiography dimensions of right pulmonary artery (RPA) and left pulmonary artery (LPA) (indexed for body surface area using z-score), coarctation of the aorta (COA), adequacy of interatrial communication, and degree of atrioventricular valve regurgitation (AVVR). A 4-point scoring system was used to grade the adequacy of interatrial communication and AVVR.

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Binary logistic regression analysis was used to identify imaging covariates associated with the need for intervention.
Results: A total of 72 patients satisfied the inclusion criteria. Their median age at BCPC was 160days, (IQR 121-284). Echocardiographic measurements were inferior to MRI for predicting the need for additional intervention on RPA, LPA or aortic arch. The MRI z-score for RPA dimensions predicted intervention, OR 1.77 (95% C.I. 1.12-2.79, p = 0.014). LPA intervention was associated with both MRI LPA z-score, OR 1.45(1.04-2.00, p = 0.027) and MRI report conclusion (OR 1.57 (1.06-2.33, p = 0.025). The MRI report conclusion predicted aortic arch intervention OR 11.5 (3.5–37.7, p = 0.00006). The need for additional AV valve repair was associated with MRI regurgitation score, OR 22.4 (1.7-295.1, p = 0.018), but not echocardiographic assessment. Echocardiography assessment was superior to MRI for predicting the need for intervention on the interatrial septum, OR 27.7 (6.3-121.6, p = 0.00001).
Conclusion: This study demonstrates that for branch pulmonary artery, aortic arch, and AVVR, MRI parameters more reliably predict the need for intervention. However, the adequacy of interatrial communication was more accurately identified by echocardiography. The complete assessment of patients with single ventricle physiology approaching BCPC, requires the cumulative strengths of multimodality imaging. The relative strengths of MRI and echocardiography should be acknowledged when recommending intervention.

O6-2
NT-proBNP in Acute Kawasaki Disease is Predictive of Coronary Artery Involvement

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Background: Natriuretic peptides are natural endogenous diuretics released by the myocardium in normal state, and to a high extent with increased myocardial distension and inflammation. We have lately documented the importance of N-terminal B-type natriuretic peptide (NT-ProBNP) aiding the diagnosis of Kawasaki disease (KD).
Objectives: We sought to investigate the potential value of NT-proBNP pertaining to the prediction of coronary artery (CA) involvement and of resistance to IVIG therapy. We hypothesized that increased serum NT-proBNP correlates with increased resistance to IVIG and CA dilatation.
Methods: Prospective study involving newly diagnosed KD patients treated with 2g/Kg IVIG within 5-10 days of onset of fever. All subjects had echocardiography at onset, then weekly for 3 weeks, then at month 2 and month 3. CA were measured at each visit and CA Z-score was calculated (Dallaire & Dahdah JASE 2010). All subjects had NT-proBNP serum level measured at onset Electrochemiluminescence IA (Roche-Dx), and Z-core calculated (McNeal-Davidson et al, Pediatrics International 2012). The aim was to determine if elevated NT-proBNP (Z-score > 2.0) was predictive of CA dilatation (Z-score > 2.5) and/or resistance to IVIG (fever 36 H after IVIG).
Results: There were 109 patients enrolled, at 6.58 ± 2.82 days of fever, age 3.79 ± 2.92 years. High NT-proBNP was predictive of CA dilatation at onset in 22.2% vs 2.6% for normal NT-proBNP (OR 4.8 [95%IC 1.05-22.4]; p = 0.031). This was also predictive of cumulative CA dilatation for the first month (p = 0.04-0.025), but not during convalescence at 2-3 months (OR = 1.28 [95%IC 0.23-7.3]; p = NS). This observation reflects therapeutic response to IVIG. In fact, elevated NT-proBNP did not predict IVIG resistance, 15.3% vs 13.5% (p = 1).
Conclusion: Elevated NT-proBNP predicts acute CA dilatation in treated KD, but not IVIG resistance. Normal NT-proBNP is associated with a diminished risk of persistent CA dilatation in IVIG responsive patients.

O6-3
Early detection of isolated coarctation of the aorta – still a challenging task

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Objectives: Newborns with coarctation of the aorta (CoA) are at a particular risk of being discharged from newborn nurseries without a diagnosis. These neonates often have a normal routine physical examination before closure of the arterial duct.
We aimed to identify symptoms and signs that led to the diagnosis and to determine to what degree prenatal screening and neonatal pulse oximetry screening contribute to the diagnosis.
Methods: Data were reviewed retrospectively for infants with isolated CoA, born between January 2003 and December 2012 in our referral area for cardiac surgery, who either died before surgery or underwent repair of CoA before 2 months of age. Cases were identified from our surgical files and from the causes of death registry (National Board of Health and Welfare). Data on clinical presentation, diagnosis, management and outcome were collected from hospital charts.
Results: Eighty-eight infants fulfilled inclusion criteria. Five neonates died before coming to surgery. The remaining 83 infants were diagnosed at a median age of 5,6 days (0-51). Only 2 were detected prenatally and 4 of 19, who were born in units using pulse oximetry screening, had a positive screen. Forty-two presented with a systolic murmur with (18) or without (24) weak femoral pulses. Eleven had weak femoral pulses but no murmur. Forty-six (55%) were discharged without a diagnosis, and 7 of them underwent echocardiography (normal in all) before discharge. Twenty-two of the discharged neonates (48%) were in circulatory shock on re-admission. Two babies died after surgery, but CoA was probably not the primary cause of death.
Conclusions: We showed that CoA is still rarely diagnosed prenatally and is also usually not detected by pulse oximetry screening. In addition neonatal routine physical examination also fails to diagnose a substantial proportion of children with CoA.
Early diagnosis depends on careful palpation of femoral pulses and wide indications for echocardiography. However, even echocardiography sometimes fails to diagnose CoA, or the obstruction has not yet developed.

O6-4
High yield and therapeutic implications justifies genetic investigations in childhood hypertrophic cardiomyopathy

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Introduction: The genetic background of hypertrophic cardiomyopathy (HCM) diagnosed in childhood was earlier considered

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be characterised by a lower proportion of sarcomeric mutations and a higher proportion of metabolic or storage disorders than HCM diagnosed in adulthood, although lately data from tertiary referral centres has suggested a similar spectrum of sarcomere gene mutations also in childhood HCM. However, geographically-based population data are lacking, particularly in the paediatric age range.
Methods: Patients belonging to the West Götaland region with presentation of non-syndromic HCM either in childhood or in adulthood were offered genetic investigations; only one family declined. In total 47 patients with a childhood presentation, and 55 patients with adult presentation had samples sent for assay at accredited laboratories (in Oxford, Copenhagen and Helsinki). A minimum screen of 13 genes were performed (MYBPC3, MYH7, TNNT2, TNNI3, MYL2, MYL3, TPM1, ACTC1, CSRP3, PLN, FHL1, PRKAG2, and GLA; in patients with marked ECG changes LAMP2 was also analyzed) and in those negative on this screen a 100-gene survey is being undertaken.
Results: Causative mutations were identified in a significantly higher proportion of childhood HCM, 38/47 (81%), than in adult HCM, 33/55 (60%; $p = 0.016$). Mutations in MYBPC3 were most common both in childhood (40%) and in adulthood (36%), followed by MYH7 30% in childhood, versus 16% in adults. Further causative mutations found in childhood were ACTC (5%), MYPN, MYL2 and LAMP2 all 2.6%. An additional LAMP2 mutation was found among 9 tertiary referral patients from other regions, so a mutation causing Danon disease was unexpectedly found in a total of 3.6% of paediatric patients with non-syndromic HCM. Among adults mutations were found in TNNI in 5.5%, and MYL2 in 1.8%. 10.6% of childhood HCM and 22.8% of adult HCM had mutations of unknown significance that were not known polymorphisms, and 8.5% of childhood HCM and 17.5% of adult HCM had no identified mutations or known polymorphisms.
Conclusions: Genetic investigation of childhood presentation of non-syndromic HCM has a higher yield than in adult HCM with sarcomere genes dominating, and sometimes has major therapeutic consequences such as the early recognition of Danon disease leading to early consideration of cardiac transplantation.

O6-5
Neurodevelopmental Outcomes after Staged Palliation for Hypoplastic Left Heart Syndrome – Impact of Cerebral Tissue Oxygen Saturation

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Background: Patients undergoing the Norwood procedure are at risk for impaired neurodevelopmental outcome. Perioperative cerebral hypoxia might be causative. We evaluated the impact of cerebral tissue oxygenation on neurodevelopmental outcome.
Methods: Neurocognitive testing was performed in 22 patients with hypoplastic left heart syndrome (HLHS) at a median age of 4.0 (3.1-5.0) years. Verbal IQ, performance IQ and full scale IQ were evaluated with the Hannover-Wechsler-Intelligence scale (HAWIVA-III). The German "Kognitiver Entwicklungstest für das Kindergartenalter" (KET-KID), which is composed of a global scale for cognitive development, a verbal and a nonverbal scale, was applied to asses cognitive functions. Cerebral oxygen saturation (cSO2) was recorded for 24 hours before and 48 hours after the Norwood procedure. Mean preoperative cSO2 values and the mean cSO2 of the first 4 postoperative hours were calculated. The duration of cSO2 below 40% was determined.
Results: Median verbal IQ was 100 (78-127), performance IQ was 93 (84-112) and median full scale IQ was 96 (81-111). Full scale IQ was below average in 4 cases; additional 5 cases had results in the low normal range. Median percentile ranks of the KET-KID were 38 (0-88) for the global scale, 48 (0-96) for the verbal and 39 (0-77) for the nonverbal scale. Results on the global scale were below average in 5 patients; another 5 had results in the low normal range. Overall, HAWIVA-III or KET-KID results were below average in 6 patients. In 13 patients with results in the low normal range or results below average, preoperative cSO2 and early postoperative cSO2 were lower compared to remainder ($61 \pm 4\%$ vs. $65 \pm 3\%$, $p = 0.013$ and $42 \pm 5\%$ vs. $49 \pm 7\%$, $p = 0.015$). The duration of cSO2 below 40% was not different (40 (0-290) vs. 180 (0-400) minutes, $p = 0.385$). Preoperative cSO2 correlated with the verbal ($r = 0.46$, $p = 0.033$) and full scale IQ ($r = 0.46$, $p = 0.030$) and with the global ($r = 0.59$, $p = 0.005$), verbal ($r = 0.55$, $p = 0.010$) and nonverbal ($r = 0.45$, $p = 0.039$) scale of the KET-KID.
Conclusions: Overall, HAWIVA-III and KET-KID results of HLHS patients after Fontan completion were in the normal range. Lower preoperative and early postoperative cerebral tissue oxygen saturations were associated with worse test results.

O6-6
Acute Vasodilator Response in Paediatric Pulmonary Arterial Hypertension: current clinical practice from the TOPP-registry

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Introduction: Acute pulmonary vasodilator testing(AVT) in paediatric pulmonary arterial hypertension(PAH) is considered important to identify patients with favourable prognosis using calcium-channel-blocker-(CCB)-therapy. However criteria used to identify acute responders, the prevalence of responders and the use of CCB in paediatric PAH are insufficiently studied.
Methods: Consecutive PAH-patients, 3months-18years at diagnosis were enrolled in the Tracking- Outcomes-and-Practice-in-Paediatric-Pulmonary-Hypertension-(TOPP)-registry at 31 centres in 19 countries from Jan-2008 to May-2013. Patients who had valid AVT during diagnostic heart catheterization were included in this study. Agents used for this test were recorded. AVT-responder status was identified at the discretion of the treating physician and retrospectively compared to the response according to criteria proposed by Barst for paediatric patients (2012) and Sitbon for adult patients (2005).
Results: Of 529 PH-confirmed children in the registry, 382 were eligible for inclusion in this study. Of these, 212 had idiopathic/hereditary PAH(IPAH/HPAH) and 105 had PAH associated with congenital heart disease(PAH-CHD). In 70% of the patients,

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AVT was performed using inhaled NO (+/-O2-suppletion), a variety of other agents were used in the remaining patients. Acute responders were identified in 78 (37%) of IPAH/HPAH patients according to the treating physician, 62 (30%) according to Barst-criteria and 32 (15%) according to Sitbon-criteria. For PAH-CHD patients these percentages were 36%, 13% and 7% respectively. Correlation between assessment by treating physician and by published response criteria was poor.
Regardless of the criteria used, acute responders had favourable baseline hemodynamics compared to non-responders, with lower mean pulmonary arterial pressure, mean right atrial pressure, pulmonary vascular resistance, PVR/SVR and PAP/SAP. There was no association between acute response status and age, sex, 6-minute walking distance or NYHA Functional Class.
Of the IPAH/HPAH patients, judged by the treating physician to be acute responders, only 23% were treated with CCB without additional PAH-targeted therapy. This was 26% in the "Barst-responders" and 47% in the "Sitbon-responders".
Conclusions: The current practice of identifying responders to AVT in centres treating children with IPAH/HPAH shows large discrepancies with the assessment by response criteria proposed in literature. Furthermore, in current clinical practice, the majority of paediatric IPAH/HPAH acute responders are not treated with CCB-therapy.

O7-1

Executive functions development in children with transposition of the great arteries: A first longitudinal study

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Objectives: Executive functions (EF) impairments are part of the main neurocognitive morbidities after cyanotic congenital heart disease (CHD). EF are higher-order neurocognitive skills underlined by the progressive maturation of prefrontal brain networks. Few studies have focused on EF development in children with CHD despite their crucial influence on academic achievement and social adaptation. More importantly, current data does not allow to draw conclusions on the evolution of these deficits as children grow up. The aim of this study is to characterize the pattern and severity of EF impairments in children with transposition of the great arteries (TGA) at a key developmental period of transition from preschool to school-age.
Methods: 45 children with TGA with or without ventricular septal defect (VSD) (mean age at intake = 5 y 4 mo; 67% males) were compared to 45 control children (mean age at intake = 5 y 5 mo; 62% males) on formal neuropsychological evaluation once a year for three consecutive years. Assessments included a comprehensive evaluation of motor and attentional inhibition, short-term and working memory, cognitive flexibility as well as a general IQ and language measures. Demographic variables were controlled.
Results: All children with TGA had normal IQ scores and did not differ from controls in any demographic variable including parental socio-economic status and educational levels (ps > 0.05). No significant differences between the groups were found in working memory measures (verbal and visuo-spatial) at any time point ($p > 0.05$). At age 5, patients displayed significantly worse scores at all measures of motor and attentional inhibition and cognitive flexibility (all $p < 0.05$). However, a different pattern of evolution was observed according to the EF domain. Impairments at motor inhibition tests normalized at age 6 contrary to deficits in cognitive flexibility that tended to aggravate with age and remained clinically important at age 7 ($p < 0.05$).
Conclusions: TGA, as other types of cyanotic CHD, is associated with altered EF. However, all EF domains may not have the same risk of dysfunction. Some deficits catch-up and some tend to worsen with age. Further characterization of these morbidities is necessary to develop targeted cognitive prevention and intervention programs at different age periods.

O7-2

Clinical Presentation, Therapy and Prognosis of Pulmonary Arterial Hypertension in Patients with Congenital Heart Defects: An Analysis Based on the Data of the German National Register for Congenital Heart Defects

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Background: 5-10% of patients with CHD develop PAH. We provide an overview over the spectrum of disease, clinical presentation, therapy and outcome based on the data of the German National CHD Register.
Patients and Methods: We included patients >1 year of age with persistent PAH after shunt closure or unoperated/palliated patients (including Eisenmenger syndrome [ES]).
Patients with isolated postcapillary pulmonary hypertension, patients in whom pulmonary pressures normalized after timely surgical intervention, patients with idiopathic PAH or patients with persistent PAH of the newborn were excluded. We defined the presence of PAH in accordance with current guidelines. In the absence of recent right heart catheterization data an estimated systolic pulmonary pressure >40 mmHg measured on echocardiography was also accepted for the diagnosis. We collected data on clinical symptoms, exercise limitation, medical therapy and outcome.
We included 184 patients (mean age 24.6 ± 14.9 years, 58.7% females). Of these, 108 patients had ES. Eisenmenger patients were approximately 10 years older than the remaining patients (mean age 29.3 ± 12.8 years vs. 17.8 ± 15.2 years, $p < 0.001$). Overall, 61.9% of patients were in NYHA class III. The mean 6-minute walk test distance was 382 ± 122 m (ES 368 ± 118 m). Overall, 44% of patients (ES 51%) received advanced PAH specific therapies (69.9% Bosentan, 25.3% Sildenafil), 14% received dual medical therapy. 51% of patients received heart failure medication. 16% of patients were treated with oral anticoagulants (ES 15%), while 22% (ES 25%) of patients received Aspirin. The mean survival rate at 1, 3, 5 and 10 years of follow-up was 94%, 83%, 77% and 64% in the entire cohort, whereas survival was even worse in ES patients (93%, 76%, 66% and 49% respectively).
Conclusions: Despite the availability of widespread and timely surgical correction for CHD patients in Germany, we could identify a considerable number of PAH-CHD and especially ES patients. The majority of patients are symptomatic and have a reduced exercise capacity. A large proportion of patients received heart failure medication and especially Aspirin, which is not supported by current recommendations. Additionally, our data

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illustrate the poor prognosis of PAH-CHD patients despite the use of disease targeting therapies in 44% of patients.

O7-3
Longitudinal Development of Psychopathology and Subjective Health Status in Congenital Heart Disease Adults: a 30-43 year Follow-Up in a Unique Cohort

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Objective: To determine longitudinal changes in psychopathology in a cohort of patients 30-43 years after their first cardiac surgery for congenital heart disease (ConHD) in childhood, compare outcomes at the 30-year follow-up with normative data, and identify medical predictors for psychopathology.
Methods: This is the third follow-up (N = 266) of a cohort operated for congenital heart disease. The first and second follow-ups of this same cohort were conducted in 1990 respectively 2001. At all three follow-ups, psychopathology was assessed with standardized, parallel questionnaires. In 2011 subjective health status was assessed by the Short-Form 36. Medical predictor variables were derived form medical examination and medical records.
Results: Over a 30-year period, proportions of patients showing psychopathology decreased significantly (N = 152 complete cases). At the 30-year follow-up, overall outcomes on psychopathology for the ConHD sample were the same or even better than for normative groups. In addition, subjective health status was better compared with normative data.
No differences were found between cardiac diagnostic groups. Medical variables that predicted the course of psychopathology over time are: the scar (as judged by the patient), results of the first cardiac surgery, and the number of hospitalizations.
Conclusions: Over a 30-year period, psychopathology decreased in patients with ConHD. Levels of psychopathology in these patients, who are now aged between 30 and 54 years were comparable or even better than normative data.

O7-4
Percutaneous pulmonary valve implantation for free pulmonary regurgitation after conduit-free RVOT plasty

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Objectives: To evaluate in patients with severe PR the feasibility and safety of a strategy of pre-stenting the conduit-free dilated right ventricular outflow tracts before excessive dilation occurs, followed by percutaneous pulmonary valve implantation (PPVI).
Patients and methods: 27 patients with free PR (26 post repair Fallot, 1 post balloon PS; age 12.1 years (6.0–44.9), weight 39.0 kg (range: 20–88)) were selected by echocardiography (RVOT < 21 mm) which predicted an adequate retention zone (size ≤24 mm defined by semi-compliant balloon interrogation of the RVOT with 23-25 mm Tyshak™ balloon). RVOT pre-stenting was done with open cell bare metal stent (Andrastent™ XXL range 30-57 mm at 20-25 mm); PPVI 2 months later.
Results: 26 patients had successful prestenting and proceeded to PPVI a median of 2.2 months (range: 1.4–3.4) after initial prestent placement. 25 Melody™ valves at 20-23 mm and one 26 mm Sapien™ valve were implanted. Complications consisted of immediate embolization of prestent (n = 1) and mild stent dislocation (n = 2) at initial procedure, crumpling (n = 4) at second procedure. At 3 months, RV volume had decreased from 188 ± 38 to 158±35 ml ($p < 0.01$) and LV volume increased from 77 ± 8 < 88 ± 8 ml/m2 ($p < 0.05$). During follow-up 0.2-3 years, no stent fractures were observed, and valve function remained perfect.
Conclusions: Post-surgical conduit-free RVOT with free pulmonary regurgitation can be stented and re-valvulated percutaneously if anatomical (predominantly size) criteria are met. In experienced hands, the technique is safe with acceptable morbidity.

O7-5
Can NT-ProBNP indicate the timing of the defect closure in children with ASD and VSD?

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Objective: The aim of this study was to investigate the potential role of NT-ProBNP in the assessment of shunt severity and invasive hemodynamic parameters in children with ASD/VSD.
Methods: This is prospective, controlled (n = 62), double-blind study. NT-ProBNP levels were correlated with various hemodynamic measurements (the ratio of pulmonary blood flow to systemic blood flow (Qp/Qs), pulmonary artery $pressure_{mean\text{-}peak}$ ($PAP_{mean\text{-}peak}$), pulmonary vascular resistance (PVR), ratio of pulmonary vascular resistance to systemic vascular resistance (PVR/SVR), the ratio of systemic peak pressure to pulmonary peak pressure (Sp/Pp), left ventricle end-diastolic pressure (LVEDp), right ventricle end-diastolic pressure (RVEDp)) which were obtained during cardiac catheterization of 127 pediatric patients (VSD = 64, ASD = 63; Table). A Qp/Qs ratio of ⩾1.5 was considered to indicate a significant shunt and the NT-proBNP cut-off points were determined for this Qp/Qs value in both defect types. A ROC analysis was carried out for the cut-off levels of NT-ProBNP.
Results: Statistically significant relationship was found between the mean NT-ProBNP values of the patients with Qp/Qs of ⩾1.5 in both defect types and that of the control group. A NT-ProBNP level of ⩾113.5 pg/ml was found to be associated with high specificity and sensitivity in determining VSD patients with a significant shunt. NT-ProBNP cut-off point of 57.9 pg/ml was found to determine a significant shunt in patients with ASD. A significant positive correlation was found between the all of invasive hemodynamic parameters with NT-ProBNP levels in patients with VSD, also a significant positive correlation was found only between mean pulmonary artery pressure, right ventricle end-diastolic pressure and the ratio of systemic pressure to pulmonary pressure with NT-ProBNP levels in patients with ASD.
Conclusion: Our study demonstrated that the NT-ProBNP measurements could be used as a supporting parameter by clinicians in determining the significance of shunt and the timing of VSD and ASD closure.

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Variables	Control ($n = 62$)	VSD ($n = 64$)	ASD ($n = 63$)	p
Age (months)	84.0 (24.0-130.0)[a]	12.0 (5.5-56.0)[b]	60.0 (40.0-96.0)[a]	**<0.001**
Gender (male/female)	33 (53.2)/29 (46.8)	36 (56.2)/28(43.8)	35 (55.6)/28 (44.4)	0.370
BMI (kg/m^2)	17.3 (15.4-18.8)	9.0 (5.7-18.0)	16.6 (15.0-19.5)	0.126
Qp/Qs	-	2.1 (1.5-3.6)	2.0 (1.5-2.8)	0.349
Sp (peak, mmHg)	-	96.0 (89.0-103.0)	100.0 (93.0-100.0)	0.457
Sp (mean, mmHg)	-	75.5 (67.0-81.0)	76.0 (68.0-80.0)	0.847
RAp (mean, mmHg)	-	7.0 (6.0-8.0)	8.0 (7.0-9.0)	0.070
PAP (peak, mmHg)	-	33.0 (27.0-51.5)	28.0 (25.0-32.0)	**0.001**
PAP (mean, mmHg)	-	21.0 (17.5-31.5)	19.0 (17.0-23.0)	**0.005**
PVR (odds)	-	1.1 (0.8-1.8)	0.9 (0.5-1.2)	**<0.001**
PVR/SVR	-	0.08 (0.05-0.11)	0.07 (0.04-0.09)	**0.042**
NT-ProBNP (pg/mL)	55.5 (31.6-72.8)[a]	182.5 (77.5-952.0)[b]	109.0 (51.4-201.0)[c]	**<0.001**
LVEDp (mmHg)	-	8.0 (7.0-9.5)	-	-
RVEDp (mmHg)	-	-	8.0 (6.0-10.0)	-

Values are expressed as n(%) or median(1st-3rd quartiles). Different superscripts in a column indicate a statistically significant difference between groups. BMI: Body mass index, Sp: Systemic arterial pressure, RAp: Right atrial pressure, PAP: Pulmonary arterial pressure, PVR: pulmonary vascular resistance, PVR/SVR: ratio of pulmonary vascular resistance to systemic vascular resistance, LVEDp: Left ventricular end-diastolic pressure, RVEDp: Right ventricular end-diastolic pressure.

O7-6
Long-term experience with Heart Transplantation in children and patients with congenital heart disease

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This study is to assess the long-term outcome of heart (HTx) and heart-lung transplantation (HLTx) in patients with congenital heart disease (CHD) and children with non-congenital cardiac or pulmonary disease.

Methods: The study is a retrospective single-centre analysis of long-term outcome after heart transplantation of children and adults with CHD, and children with non-congenital cardiac disease.

Results: From 1984 to 2013, 111 first-HTx, 5 HLTx and 6 second-HTx were performed (62 males), in patients aged 11.7 ± 8.2y (med12y): 96 (79%) were aged <18y at transplant. Cardiac diseases included 61 cardiomyopathies (50.8%), 50 CHD (41.7%), 6 retransplants (5%). HLTx included 1 Eisenmenger syndrom, 1 primitive pulmonary hypertension, and 2 pulmonary diseases. Patients with cardiomyopathy were younger than those with CHD (8.7y vs 14.9y). Seventeen (14%) patients were under circulatory mechanical support as a bridge to transplant. Immunosuppression was achieved by tritherapy in the majority of cases. Acute rejection occurred more frequently within the first year post-transplant (>5th year) or late after transplant in teenagers because of non-compliance. Overall mortality was 27%: 33 patients died, 3.5 ± 4.6y post-HTx (1 day to 16.4 y, med 1.5 months), due to : early multivisceral failure in 6 (18%), Pulmonary hypertension in 3 (9%), acute rejection in 7 (21%), graft coronary disease in 6 (18%), sepsis in 5 (15%) and miscellaneous in 6. Graft coronary disease occurred in 15 cases (12.4%), of them 4 had 2nd HTx, 6 died and 5 are alive. Five posttransplant lymphoma occurred, 4months to 14 y after HTx and were successfully cured in 4 (1died). Patients survival was 85% at 1 y, 81% at 5 y, 70% at 10 y and 61% at 20 y post-transplant. Graft survival rates were respectively 82%, 68% and 52% at 5 y, 10 y and 20 y post-transplant. Survival did not differ with pretransplant disease, age, gender, pretransplant mechanical support. Mortality was higher in patients with coronary disease (40%) than those free from (25%). The majority of survivors (97%) are in NYHA class I.

Conclusion: Our experience with HTx and HLTx is favourable with acceptable long-term prognosis in patients with end-stage disease. Graft coronary disease is the main cause of failure, less frequent than in the adult non-CHD heart-transplanted population.

O8-1
Aortic and carotid stiffening in patients after successful coarctation repair and its impact on left ventricular diastolic function

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Introduction: Arterial hypertension and accelerated arteriosclerosis are late complications even after successful coarctation (CoA) repair, which may clinically affect left ventricular (LV) function in the long-term follow-up. However, only few data exist on the occurrence of abnormal vascular bioelasticity and its effect on LV diastolic function in CoA patients with no or only mild arterial hypertension during long-term follow up. Therefore, we studied children and young adults after successful CoA repair using magnetic resonance imaging (MRI).

Methods: Fifty-two patients (18.9 ± 10.7 years), 14.6 ± 9.2 years after CoA repair, and 54 controls (19.0 ± 7.9 years) underwent 3.0-Tesla MRI. The data were used to calculate distensibility and pulse wave velocity (PWV) at different aortic locations. Furthermore, in a subgroup of patients and controls common carotid artery distensibility, PWV, wall thickness and wall area were measured. LV ejection fraction (EF), volumes (LVEDV, LVESV) and mass were assessed from short axis views. Axial cine images were used to measure left atrial (LA) volumes and parameter for LA diastolic function ($LAEF_{Passive}$, $LAEF_{Contractile}$, $LAEF_{Reservoir}$).

Results: In patients aortic distensibility was reduced at all positions of the thoracic aorta (aortic root: 5.5 ± 3.8 vs. 7.4 ± 3.0 10^{-3} $mmHg^{-1}$, ascending aorta: 5.8 ± 3.1 vs. 8.1 ± 3.6 10^{-3} $mmHg^{-1}$, descending aorta at the isthmus: 5.6 ± 3.0 vs. 6.8 ± 2.3 10^{-3} $mmHg^{-1}$, descending aorta at the diaphragm: 6.7 ± 2.8 vs. 8.0 ± 2.8 10^{-3} $mmHg^{-1}$; $p < 0.05$) and PWV in the aortic arch (4.7 ± 1.8 vs. 3.8 ± 0.8 m/s, $p < 0.01$) was elevated. In addition, the subgroup analysis showed an increased carotid PWV, wall thickness and wall area ($p < 0.05$). The LA volume before atrial contraction ($LA\text{-}Vol_{ac}$), and the minimal and maximal LA volume ($LA\text{-}Vol_{min}$, $LA\text{-}Vol_{max}$) were higher in patients ($LA\text{-}Vol_{ac}$: 33.2 ± 9.8 vs. 27.4 ± 5.9 ml/m^2, $LA\text{-}Vol_{min}$: 25.3 ± 7.6 vs. 20.9 ± 5.1 ml/m^2, $LA\text{-}Vol_{max}$: 48.4 ± 11.4 vs. 43.2 ± 8.7 ml/m^2; $p < 0.05$). $LAEF_{Passive}$ and $LAEF_{Reservoir}$ were reduced ($LAEF_{Passive}$: 31.7 ± 8.4 vs. 36.9 ± 6.6%, $LAEF_{Reservoir}$: 48.0 ± 7.2 vs. 51.9 ± 6.8%; $p < 0.01$) and correlated negatively with aortic arch PWV ($p < 0.05$).

LVEF, LVEDV, LVESV and LV mass were not changed compared with controls.

Conclusions: Patients after CoA repair have reduced bioelasticity of the entire thoracic aorta and the common carotid artery as well as an abnormal carotid wall structure. The impaired aortic bioelastic function likely contributes to LV diastolic dysfunction.

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O8-2

Importance of haemodynamic right and left ventricular parameters and CPET-results in patients with Tetralogy of Fallot

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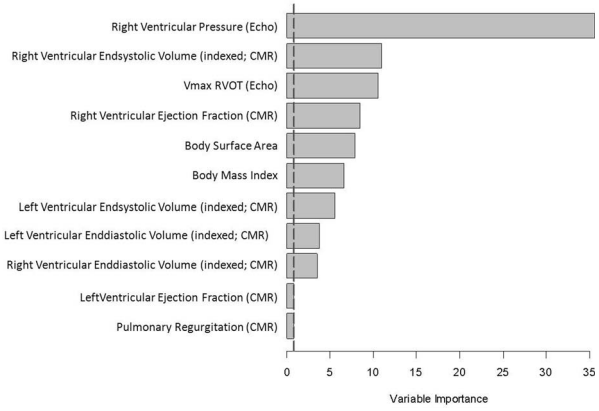
Objectives: Good quality of life correlates with a good exercise capacity in daily life in patients with Tetralogy of Fallot (TOF). Patients after correction of TOF usually develop residual defects as pulmonary regurgitation or pulmonary stenosis of different severity. We investigated the impact of several haemodynamic parameters measured by cardiovascular magnetic resonance (CMR) and echocardiography and analysed these data together with results of cardiopulmonary exercise testing (CPET) of these patients.
Methods: 136 consecutive patients with TOF were tested during routine follow-up with CMR, echocardiography und CPET. Right and left ventricular volume data, ventricular ejection fraction, pulmonary regurgitation were evaluated by CMR. Echocardiographic pressure gradients in the right ventricular outflow tract (RVOT) and through the tricuspid valve area were measured.
All data were classified and correlated with the results of CPET evaluations of these patients. The analysis was performed using the Random Forest model (classification and regression model with measurement of variable importance through permutation). In this way we calculated the importance of the different haemodynamic variables related to the maximal oxygen uptake in CPET (figure).
Results: Right ventricular pressure showed the most important influence on maximal oxygen uptake, whereas pulmonary regurgitation and right ventricular end-diastolic volume were not important haemodynamic variables to predict maximal oxygen uptake in CPET.
Conclusions: Patients with TOF and elevated right ventricular pressure showed a reduced exercise capacity. The maximal exercise capacity did not correlate with the percentage of pulmonary regurgitation or the right ventricular end-diastolic volume. Complex multivariate analysis can be performed in a reliable way by the Random Forest model.



O8-3

Initial shunt type for the Norwood procedure affects long-term myocardial function in children with HLHS

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Introduction: Long-term effects of initial shunt types on myocardial function are poorly understood in hypoplastic left heart syndrome (HLHS). We studied the effect of initial shunt type, Blalock–Taussig (BT) and Sano, on myocardial function at different stages of the treatment protocol.
Methods: Population based cohort of Finnish children with HLHS (n = 63) born between 2003 and 2010 was studied with echocardiography at four time points: before stages one, two and three and 0.5-2 years after stage three. 23 children were palliated with a BT shunt and 40 with a Sano shunt. For comparison, we utilized the fractional area change (FAC) derived from velocity-vector-imaging (Syngo, Siemens). This method has been validated with MRI.
Results: There were no differences between groups in demographics, HLHS morphology or mortality (BT 26.1% vs Sano 15.0%, p = 0.2) during study period. Among the survivors, an increase in FAC was observed after stage two in the BT shunt group but not in the Sano shunt group (+8.8 ± 9.4%units vs +1.8 ± 8.5% units, p = 0.01) (Figure). After stage three, those palliated with a BT shunt had higher FAC values (33.0 ± 6.4% vs 25.6 ± 6.1%) (Figure). In multiple regression analysis, shunt type and stage of palliation had an impact on myocardial function.
Conclusions: In patients palliated with a BT-shunt, increase in FAC during treatment protocol was higher and myocardial function after stage three better than in those palliated with a Sano shunt. This may be due to long term effect of myocardial scarring caused by a Sano shunt.

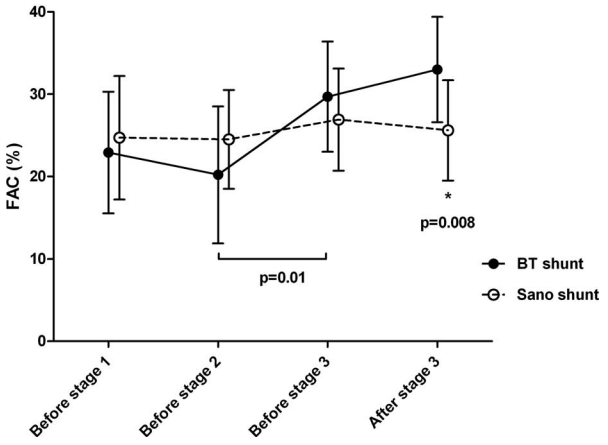

Figure. Fractional area change (FAC) in HLHS patients in different stages of treatment protocol initially palliated with either a Blalock–Taussig (BT) or a Sano shunt.

O8-4

Left ventricular preload is reduced in patients with tetralogy of Fallot and chronic pulmonary regurgitation

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Introduction: Dysfunction of the right ventricle in patients with TOF and significant pulmonary regurgitation (PR) leads to systolic dysfunction of the left ventricle (LV) due to altered ventricular interaction. In addition, abnormal diastolic function of the right ventricle (RV) affects emptying of the right atrium due to elevated RV end diastolic pressure. We wanted to measure the volumes of both atria by using cardiac magnetic resonance imaging in paediatric patients with TOF. We were specifically interested whether chronic pulmonary regurgitation affects preload of the LV.
Methods: The study subjects were 45 TOF patients with surgical repair between 1990-2003. All patients had pulmonary regurgitation. In addition, forty-four healthy volunteers were recruited. The average ages of patients and controls were 13.1 ± 3.2 vs.14.1 ± 3.4 years (ns). The mean post-operative follow-up time of the patients was 11.8 ± 3.1 years.
For atrial and ventricular volume measurements, the end diastolic and systolic cine images were manually planimetered (Philips ViewForum workstation). The conduit flow through the atria, arterial outflow and pulmonary (PR) and aortic (AR) regurgitation volumes were assessed using a phase contrast flow measurement. The volumetric data were indexed to BSA per cardiac cycle.
Results: The PR-volume of TOF patients was 16.0 ± 11.7 exceeding 30 ml/m^2 in seven subjects. The PR of control subjects and AR in all subjects was negligible.
The ventricular preload volumes derived from the atria were significantly reduced in TOF patients. The volume from the right atrium in patients and controls was 41.1 ± 7.0 vs. 50.7 ± 6.8; p = 0.001, respectively, and that from the left atrium 44.8 ± 7.3 vs. 50.3 ± 5.7 ml/m2; p = 0.002. Due to PR, the end diastolic RV–volumes, however, were significantly higher in TOF patients (115 ± 26.5 vs. 83.6 ± 14.8 ml/m^2; p < 0.0001).
The ejection fraction of the RV was significantly reduced in TOF-patients (50.1 ± 8.6 vs. 60.0+4.8%; p < 0.0001), whereas LV ejection fractions were preserved (61.1 ± 8.8 vs. 63.1 ± 4.7%; ns.)
Conclusions: In TOF patients with pulmonary regurgitation, volume flow through LA is decreased. Reduction in LV preload volume might be an important factor contributing to disturbed ventricular interaction and LV dysfunction.

O8-5

Outcomes and 3D Echocardiographic Predictors of Mitral Valve Reconstruction for Congenital Mitral Valve Disease

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Background: Congenital mitral valve disease (CMVD) is a significant challenge for medical and surgical management. Recent improvements in imaging, including three-dimensional echocardiography (3DE), and enhanced surgery seem to have improved outcomes. The aim of this study was to evaluate recent results of reconstructive surgery for CMVD, and to determine 3DE predictors of freedom from reintervention
Methods And Results: 106 patients with CMVD that underwent surgery from 2001 to 2011 were included. Median age and weight at operation was 16.8 months (0.1–216) and 8.6 kg (2.4-74), respectively. 41 patients (48%) had mitral stenosis (MS), 25 (29%) had mitral regurgitation (MR), and 20 (23%) had mixed disease (MD). Median follow-up was 38 months (0.5 months-12 years). For all patients, survival was 94.3 ± 2.5% at 1 year and 91.6 ± 3.6% at 5 years (see also Figure 1). Freedom from reoperation was 85.4 ± 5.5% at 1 year and 77.8 ± 7.2% at 5 years. Freedom from valve replacement was 93.0 ± 2.8% at 1 year and 89.4 ± 3.7% at 5 years. MS, MR and MD patients did not differ with respect to survival, freedom from reoperation or valve replacement.
36 patients had 2 and 3DE data sets 3DE effective orifice area (EOA), vena contracta regurgitant area (VCRA) and mean gradients (MG) were determined preoperatively and before discharge. EOA for MS and MD patients increased from 0.56 to 0.78 cm^2 (p = 0.025); VCRA for MR and MD patients decreased from 1.08 to 0.44 cm^2 (p = 0.007). An increase of the EOA > 30% (p = 0.006) and a decrease of the VCRA > 100% (p = 0.011) was associated with improved freedom of reoperation, in contrast to any changes in MG.
Conclusions: Reconstructive surgery for CMVD can be performed with favorable survival, and good 5 year freedom from reoperation and valve replacement. 3DE parameters, including increase in EOA for stenotic and decrease in VCRA for regurgitant valves in contrast to changes in MG are associated with improved freedom from reoperation. We postulate that MG did not change significantly due to high left ventricular filling pressures, emphasizing the importance of analysis of the EOA to determine outcomes. These parameters will help to guide medical and surgical management.

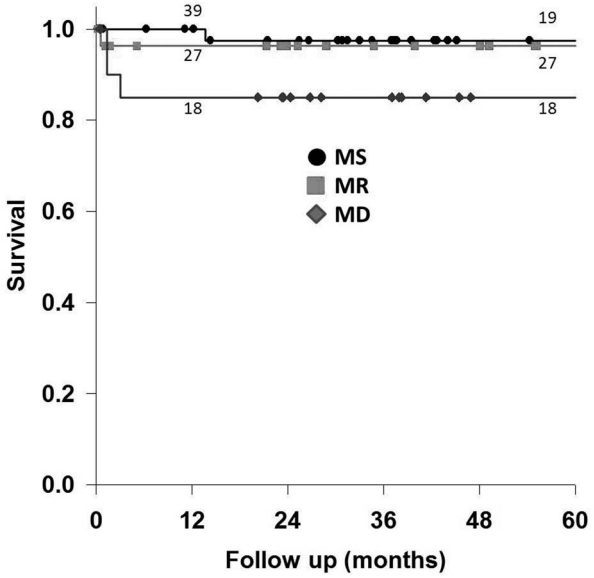

Figure 1. Survival of the three different patient groups.

O8-6

Determinants of pulmonary valve replacement indication in repaired Tetralogy of Fallot patients: a multicenter experience

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Introduction: Repaired Tetralogy of Fallot (rToF) patients often need pulmonary valve replacement (PVR) due to progressive right ventricular (RV) dilation and biventricular dysfunction. Cardiac magnetic resonance (CMR) represents the gold standard

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to evaluate RV size and function aiding definition of PVR timing. Determinants of PVR have not yet been described.
Methods: We enrolled patients who underwent hemodynamic evaluation by CMR late after ToF repair (by either transanular or infundibular RV outflow tract reconstruction) in five paediatric cardiology centers, between March 2003 and March 2013. Surgical data (time and types of repair) and CMR parameters were collected. Indications to PVR change to single center.

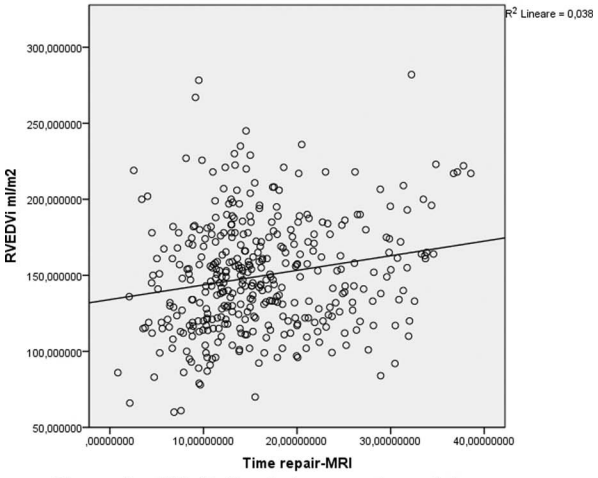

Figure 1a: RV dilation in transanular patch group

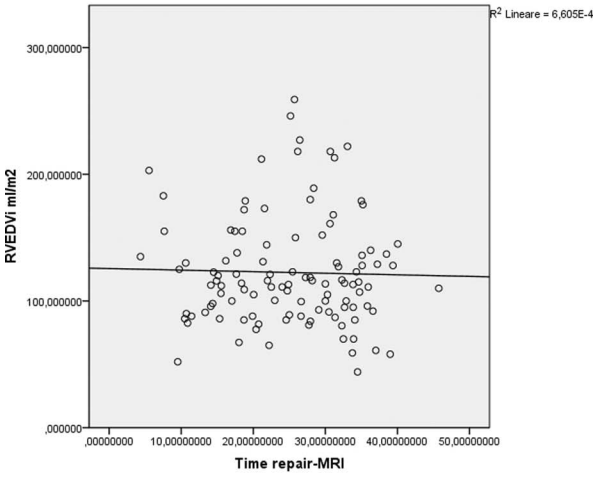

Figure 1b: RV dilation in infundibular patch group

Results: 495 patients (60% males) aged 21 ± 11 (range 2-67) years were included. Mean age at repair was 30.4 ± 54.2 (range 0.2-574.8) months. In the majority of cases (77%) transanular patch was the type of surgical correction performed. Among the study population 222 patients (47%) had RV end-diastolic volume indexed for bsa (RVEDVi) >150 mL/m2 and/or RVEF <47%, whose 70% were males and 86% with transanular patch (TP) repair. In multivariate logistic regression, in TP group, presence of RVEDVi >150 mL/m2 and/or RVEF <47% was significantly correlated with time elapsed from correction to MRI examination ($p = 0.017$) (figure 1a) and to grade of pulmonary regurgitation (PR) ($p = 0.008$). In addition, mean RV EF and LV EF were mildly lower in TP as compared to infundibular patch (IP) group (both $p < 0.05$). In IP group RV dilation was not related to time from surgical correction ($p = NS$) (figure 1b), but only to grade of PR and to the presence of significant tricuspid regurgitation ($p = 0.022$ and $p = 0.032$, respectively).

Conclusions: The mechanism underling RV dilation in rToF patients appears to have significant differences in the two groups. Indeed, time from surgical repair seem to have a significant impact only in the TP group while tricuspid regurgitation seem to be crucial in the other. Longitudinal follow-up studies are needed to define the role of each factors involved in RV dilation in predicting the adverse outcomes of rToF patients.

O9-1
Main pulmonary artery area limits exercise capacity in patients long-term after arterial switch operation for Transposition of the Great Arteries (TGA)

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Introduction: Right ventricular outflow tract obstruction (RVOTO) is the most frequent residual lesion in patients after arterial switch operation (ASO) for TGA. This study compares outflow tract area, distensibility and pulmonary blood flow (PBF) distribution of ASO patients to healthy controls and correlates these measurements to right ventricular (RV) function and exercise capacity.

Table 1. ASO patients (without homografts) compared to healthy controls. Symbols: #Median [range] and Mann-Whitney U test, other mean (±SD) and Student T-test. *p-value compared to healthy controls <0.05; **<0.001.

	ASO (n = 48)	Healthy (n = 21)
Age #	21.3 [12.2-35.3]**	26.4 [21.0-35.2]
Male (%)	32 (67%)	10 (48%)
RV function:		
RVEDV (ml/m^2)	101.5 (13.2)*	109.2 (18.0)
RVESV (ml/m^2)	50.3 (8.5)	55.2 (10.8)
RVEF (%)	50.4 (5.2)	49.7 (3.8)
RVmass (gr/m^2)	30.7 (5.4)**	22.2 (3.8)
RVOT areas:		
MPA (mm/m^2)#	184.6 [57.6-345.6]**	270.2 [197.2-537.5]
LPA (mm/m^2)#	58.9 [15.4-253.6]**	156.5 [117.5-261.6]
RPA (mm/m^2)	110.0 (53.2)**	151.2 (32.8)
LPA distensibility(%)	29.8 (8.9)	28.2 (6.8)
RPA distensibility(%)	32.2 (9.1)	33.5 (4.5)
Flow		
LPA: RPA(%)	46.2: 53.8 (10.8)	44.9: 55.1 (4.7)
Smallest branch flow	42.1 (8.2)	43.9 (3.2)

Methods: Participants underwent cardiac magnetic resonance imaging including angiography and flow measurements and cardio-pulmonary exercise testing. RV volumes and function, PBF distribution, smallest cross-sectional area of the main (MPA), left (LPA) and right pulmonary artery (RPA), LPA and RPA distensibility (relative area change) and percentage of predicted peak oxygen uptake (VO_2max/kg%) were measured. ASO patients were compared to healthy controls.
Results: Fifty patients were included (median follow-up 21.2 [12.1-32.8] yrs). Reintervention for RVOTO was performed in 9/50 (18%) of patients. ASO patients are compared to controls in table 1. MPA area was the only factor significantly correlated with RV stroke volume ($r^2 = 0.168$, $p < 0.01$) and VO_2max/kg% ($r^2 = 0.194$, $p < 0.01$) in multivariate analysis.
Conclusions: In our cohort MPA, LPA and RPA areas were smaller compared to healthy controls. Only MPA area independently

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correlated to stroke volume and VO_2max/kg%, likely because distensibility and PBF distribution were comparable to controls. This should be considered when selecting patients for reintervention.

O9-2
Right Ventricular Myocardial Deformation and Ventricular-Ventricular Interaction in Adults with Corrected Tetralogy of Fallot

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Objectives: Due to pulmonary regurgitation, most adult patients with corrected tetralogy of Fallot (cToF) have right ventricular (RV) volume overload. Using speckle-tracking echocardiography, we evaluated regional RV and left ventricular (LV) deformation in patients with cToF and in healthy controls.
Methods: Echocardiograms including the standard apical views were acquired in adult patients with cToF and in healthy controls. With speckle-tracking echocardiography, we analyzed longitudinal strain of the RV lateral wall, LV septum, and LV lateral wall.
Results: We included 148 subjects: 95 patients with cToF (61% male, age 33.0 ± 9.6 years, age at correction 3.7 ± 4.4 years) and 53 healthy controls (49% male, age 29.7 ± 6.8 years). RV global longitudinal strain (GLS) of the lateral wall was significantly lower in cToF patients than in controls (Figure 1). Of all three RV segments, the apical segment had the lowest strain in cToF patients, whereas it had the highest in controls. LV GLS was also significantly lower in cToF patients (-17.5 ± 2.5%) than in controls (-19.8 ± 2.2%, $p < 0.001$), mainly due to reduced longitudinal strain of the interventricular septum. LV GLS was positively correlated with RV GLS ($r = 0.44$, $p < 0.001$).
Conclusions: RV longitudinal strain is reduced in cToF patients, especially in the apical segment. This suggests that apical function is more affected in these volume overloaded RVs. With regard to the LV, particularly the strain of the septum is reduced which suggests that there is a negative influence on LV function due to the mechanical coupling of the ventricles.

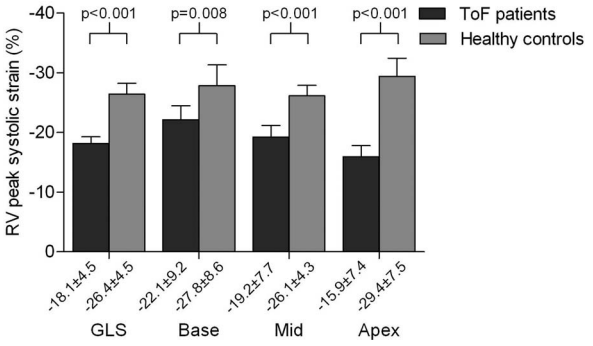

Figure. 1. RV global and segmental longitudinal strain of the lateral wall.

O9-3
Long-term outcome and late morbidity of adolescent and adult patients with Fontan circulation

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Objectives: Outcomes of the Fontan palliation for univentricular heart defects have substantially improved and most patients will now reach adulthood. However, given the significant intrinsic limitations of the Fontan circulation, we face a continuously growing population of adolescent and adult Fontan patients at risk of experiencing severe complications. We assessed long-term outcome of these patients in our institutional follow-up program aiming at early detection of complications and prompt interventions to optimize Fontan hemodynamics.
Methods: From 1990-2013, 205 patients underwent a modified Fontan operation at our institution; patients having reached ado-lescence or adulthood (n = 78) were included. Follow-up, including annual clinical exams, echocardiography and 24h-ECG-monitoring continued for a median of 14.6 years. Moreover follow-up was complemented by exercise capacity testing and patients were encouraged to complete standardized questionnaires exploring their quality of life (SF-36). MRI and cardiac catheterizations were performed according to clinical findings.
Results: There were 5 late deaths, one patient received heart transplantation due to ventricular failure. 15-year survival estimate was 94%. Severe complications included late Fontan failure (n = 8), dysrhythmias (n = 29) and thromboembolic events (n = 8). 34 patients (47.2%) received catheter interventions and 15 (20.8%) re-operations including 5 Fontan conversions. Overall late he-modynamic results were good (median pulmonary artery pressure 11 mmHg and transpulmonary gradient 6 mmHg). Median maximal oxygen consumption was 21 ml/min/kg (51% of reference). Severely decreased oxygen consumption (<40% of reference) and chronotropic competence (peak heart rate <120/min) were present in 19.6% and 30.4%, respectively. Ventricular function and Fontan flow examined by MRI were satisfactory (median ejection fraction 53%, cardiac index 2,5 l/min/m^2). Overall quality of life assessed by the SF-36 was good.
Conclusions: Stable long-term results demonstrate adequate patient selection and follow-up monitoring correlating with a good quality of life. Nevertheless, long-term morbidity remains a significant issue in adolescent and adult Fontan patients. A considerable fraction of patients displays severely decreased oxygen consumption and chronotropic competence which have recently been shown to be significant risk factors for late mortality. While inadequate Fontan hemodynamics due to increased pulmonary vascular resistance may be improved by medical, surgical or catheter intervention, in ventricular failure therapeutic options are limited and transplantation may be ultima ratio.

O9-4
Coronary anatomy and lumen area in patients after the arterial switch operation: a long term follow-up study

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Introduction: Re-implantation of the coronary arteries is a crucial part of the arterial switch operation (ASO) for transposition of the great arteries. The current study investigates coronary obstruction and anatomic characteristics of the coronaries in patients (pts) after ASO compared to controls.
Methods: Subjects underwent prospective triggered coronary CT-angiography (cCTA) using a 256-slice scanner. CCTAs were analyzed for coronary branching pattern (normal = 1LCx-2R or variant), presence of coronary stenosis and course between great

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arteries or contact with pulmonary artery was noted as 'wedged position'. Cross-sectional areas (CSA) were measured at predefined points in right (RCA) and left coronary artery (LCA) using validated software (Intellispace; Philips). Distance between lower aortic annulus and LCA/RCA coronary ostium was measured with 3Mensio (Pie Medical).
Results: 58 asymptomatic pts were included, median follow-up 20.1 yrs (range: 12.1-32.8). Variant coronary branching patterns, wedged position of a coronary artery and ostial stenosis were present in 14 (24%), 9 (15.5%) and 1 (1.7%) of pts, respectively. Thirty-nine pts (age: 21.3 ± 5.6 yrs) with 1LCx-2R (= normal branching pattern) were complety analyzed and compared to controls (age 22.9 ± 4.6 yrs) using Student T-test (figure using Student T-test (figure).
Conclusions: In our cohort, up to 33 years after ASO, ostial stenosis is present in 1 (1.7%) patient and in 9 (15.5%) patients a coronary is wedged between the great arteries. Coronary arteries in pts after ASO take off from the aortic root at a higher level and show small but significant differences in CSA compared to healthy controls.

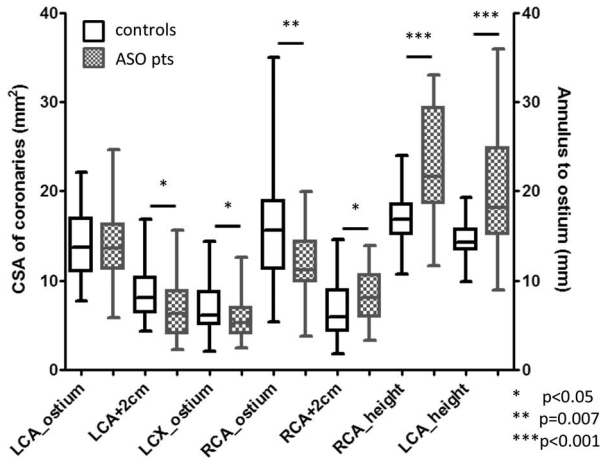

Figure 1: CSA lumen areas and angle between root and LCA/RCA of ASO patients compared to healthy controls using Student T-test.

O9-5
Causes of Hemoptysis in Eisenmenger Syndrome – A CT Angiography Study

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Introduction: Hemoptysis is a common cause of morbidity in Eisenmenger syndrome, but the causes of hemoptysis are not well defined. We analyzed the clinical predictors and causes of hemoptysis in a cohort of patients with Eisenmenger syndrome using computerized tomographic pulmonary angiography (CTPA).
Materials and methods: Of the 95 patients of Eisenmenger syndrome studied (mean age 23.7 ± 7.7 years; 57 male), 38 patients (40%) had presented with hemoptysis, and all of them underwent a CTPA within two weeks of index bleed.
Results: Patients with hemoptysis had a reduced 6 minute walk distance (356.2 ± 92.5 meters) as compared to patients without hemoptysis (395.1 ± 126.9 meters) (p = 0.03). However, other baseline demographic characteristics including diagnosis, complexity of lesion, functional class, and symptoms did not differ among patients with and without hempotysis. Of the 38 patients, 17 had a treatable cause of hemoptysis and received appropriate treatment. The identifiable causes included aorto-pulomanray collaterals, pulmonary thrombosis (2 patients), pulmonary tuberculosis (2 patients), pulmonary artery dissection (1 patient). Treating an identifiable cause reduced the risk of recurrence of hemoptysis by 0.46 (95% CI 0.28 – 0.64).
Conclusion: Hemoptysis remains a major cause of morbidity in patients with Eisenmenger syndrome. Hemoptysis occurs more frequently in patients with greater exercise limitation. CT pulmonary angiogram immediately following an episode of hemoptysis could identify a potentially treatable cause in nearly half of the patients and such treatment results in lesser recurrence of hemoptysis.

O9-6
Quantitative Assessment of Systolic Right Ventricular Function and its Relationship with NT-proBNP in Patients with a Systemic Right Ventricle

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Objectives: In patients with transposition of the great arteries (TGA) corrected by an atrial switch operation (D-TGA) and in patients with congenitally corrected TGA (L-TGA) dysfunction of the systemic right ventricle (RV) is a major concern. We evaluated RV longitudinal strain (LS) using speckle-tracking echocardiography (STE) in these patients, and assessed its relationship with conventional echocardiography and NT-proBNP. RV LS in patients was compared to RV LS in healthy controls.

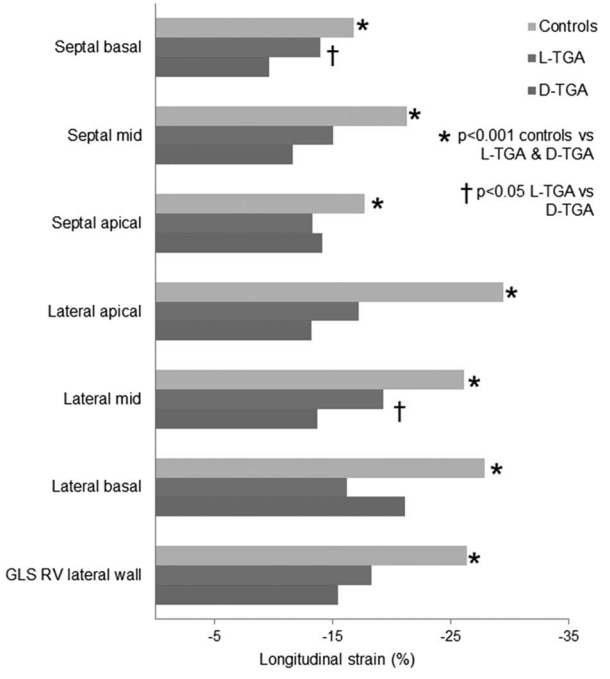

Figure 1.

Methods: Echocardiography, electrocardiography and NT-proBNP measurements were performed in consecutive patients with D-TGA (corrected by Mustard surgery) or L-TGA on the same day. Healthy controls. With STE, we analyzed longitudinal strain of the RV lateral wall and septal wall.
Results: Of the 40 patients with a systemic RV, 31 had a D-TGA and 9 patients had an L-TGA. The mean age was 36 ± 7 years,

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73% was male (34 ± 4 years after corrective surgery). The 26 healthy controls had a mean age of 31 ± 7 years and 46% was male. Longitudinal strain of the RV lateral wall tended to be lower in patients with D-TGA (-15.5 ± 3.5%) than in patients with L-TGA (-16.1 ± 3.6%, $p = 0.052$) and was significantly reduced compared to healthy controls (-26.4 ± 4.5%, $p < 0.001$). The reduced strain was most prominent in the apical segment (Figure 1). Median NT-proBNP level in the patients was 27.2 [IQR 17.6–53.9] pmol/l. RV LS correlated with RV apex-base diameter ($r = 0.54$, $p = 0.001$), RV fractional area change ($r = -0.36$, $p = 0.039$), QRS duration (0.43, $p = 0.012$) and NT-proBNP ($r = 0.53$, $p < 0.001$). No correlation between RV LS and TAPSE was observed.

Conclusions: RV longitudinal strain is significantly reduced in the systemic RV of patients with D-TGA and L-TGA, especially in the apical RV segment. RV longitudinal strain is related to RV function and dimension, and shows a negative correlation with NT-proBNP, which indicates a possible prognostic value of strain in patients with a systemic RV.

O10-1
Long-term results of complex treatment of middle aortic syndrome in children

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Middle aortic syndrome (MAS) characterized by segmental narrowing of the thoracic and/or abdominal aorta, often associated with stenoses of renal and visceral arteries, is an uncommon cause of arterial hypertension. Management strategy depends on the center experience and the anatomical forms of MAS.

The aim: presentation of single center experience of complex interventional and surgical therapy in children with MAS.

Material/Methods: Between 1994 and 2013, 26pts (aged 3-17 yrs, median 11.6) with severe arterial hypertension (AH) resistant to multidrug therapy and diagnosis of MAS underwent interventional and surgical treatment. Twenty five pts had narrowing of thoracic and/or abdominal aorta (length of stenosis 2–12 cm, minimum diameter 1.5–6 mm), one aortic atresia below the origin of stenosed left renal artery. Renal arteries stenosis coexisted in 12pts, coeliac trunk stenosis in 11pts, superior mesenteric artery in 10pts. Aotic narrowing was treated with stents in 24pts, aortic thoraco-abdominal bypass in one. Additional transcatheter (balloon angioplasty of renal arteries – 7pts, coeliac trunk – 2pts, mesenteric artery – 3pts, stenting of coeliac trunk – 1pt) and surgical (kidney autotransplantation – 12pts) procedures for primary treatment of MAS were performed.

Results: There was significant improvement ($p < 0.001$) in pre versus post stent aortic narrowing diameters (4.2 ± 2.8 mm vs 12.4 ± 3.2 mm) and systolic gradient (45 ± 5.2 mmHg vs 12 ± 5.1 mmHg). Renal function after autotransplantations were normal. During mean 9.2 ± 2.5 years follow-up additional procedures—elective stent redilation of stents implanted to aorta (8pts), additional stent implantation to aorta due to small in-stent aneurysm formation (pts) and progression of narrowing (2pts), balloon angioplasty of aorta due to neoitimal hyperplasia (7pts), balloon renal artery angioplasty (1pt), redilation of stent in truncus coeliacus (1pt) were performed.

At the latest clinical follow-up 5pts had no antyhypertensive treatment, all other had better control of AH on the lower doses of medications.

Conclusions:

1. Complex interventional and surgical treatment of children with middle aortic syndrome allows for better control of arterial hypertension.
2. Aortic obstruction related to disease can be treated successfully with the stent implantation.
3. Continuous follow-up of patients with middle laortic syndrome is required for recognition of indications for additional interventions.

O10-2
Significance of 3DRA in the cath lab for congenital heart disease

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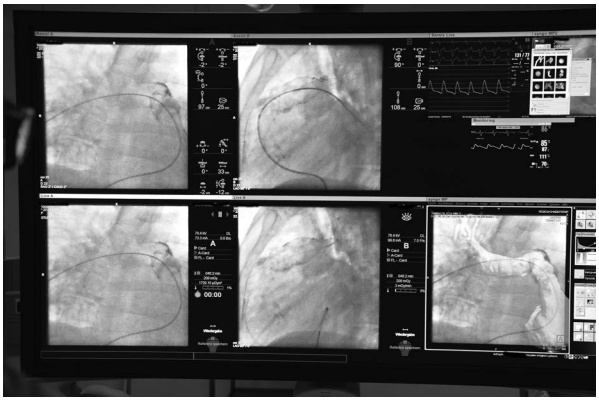
Objectives: 3DRA seems to be advantageous in the cath lab for congenital heart disease. Beneath its use as a comprehensive diagnostic tool, it promises faster and safer intervention with reduced radiation and contrast dye consumption and additionally better results.



Methods: Assessment of our 4 years' experience with 3DRA and 3D guidance in our cath lab and review of literature.

Results: More than 360 cases with 3DRA (29% of all investigations, 74% interventions), median 1.8 ml contrast/kg, 83 μGym^2, 12.4 kg [2.3-98]. 15% 3D-3D-fusion with MRI or MDCT images. Reduced fluoroscopy time in stenting aortic arch and percutaneous pulmonary valve replacement. Radiation dose lower than 0.7 mSv (biological and physical measurement). Valuable measurement of vessels with a systematic error of 4% in 3D-reconstructions. Application protocols are available, promising reproducible results for most anatomic regions. 3DRA is more than a 3D-angiography; it provides a complete 3D-CT dataset with valuable extravascular anatomic information.

Conclusions: Nowadays, a major indication for cardiac catheterization is the need of intervention. 3DRA facilitates the procedures in complex cases. 3D-models and 3D-Guidance from 3DRA or implementation of MRI or MDCT reconstructions enable a clearly demonstrating anatomy, accelerating complex interventions and improving patients' safety, particularly for less experienced investigators. Further evolvement of the detectors will reduce radiation, further development of the soft- and hardware will facilitate implementation of MRI- and CT images or allow even implementation of (4D) echo. 3DRA emerges to a standard imaging tool in catheter based cardiac interventions in congenital heart disease.

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O10-3

The Melody valved stent is more vulnerable for endocarditis than homografts or Contegra conduits in RVOT

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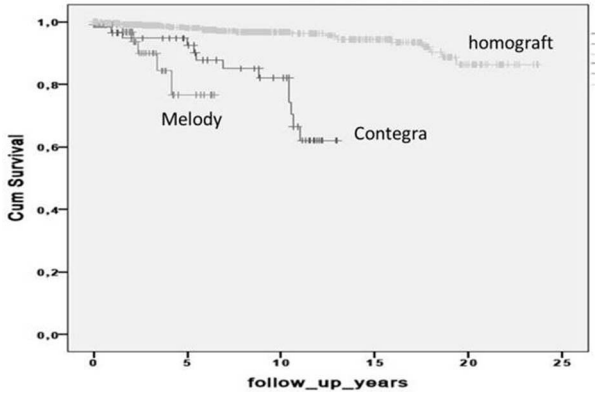
Background: All RVOT conduits are vulnerable for infective endocarditis (IE) which influences conduit longevity and clinical outcome. The incidence of IE of the Melody valved stent needs to be compared with other RVOT conduits.
Patients and methods: Retrospective study including all patients in the database of a tertiary center with an implantation of a homograft (European Homograft Bank), Contegra™ graft or Melody™ conduit in RVOT.
Results: 827 conduits were implanted in 657 patients. Between 1989 and 2013, 660 homografts were implanted in 579 patients (age 15.7 ± 12.8 y, range 3d-66 y); IE occurred in 23 pts during follow-up of 7.6 ± 6.3 y (0-23.7 y). 59 Contegra™ grafts were implanted in 58 patients between 2000 and 2003 (9.2 ± 8.6 y, range 3d-47 y); 13 (22%) had IE during follow-up of 7.5 ± 3.9 y (range 0.3-12.9 y). 109 Melody™ valved stents were implanted in 108 pts (18.3 ± 12.2 y, range 4-80y) in 2006-2013; IE occurred in 6 (5.5%)pts during 1.5 ± 1.9 y (0-6.4y). The bacteria in the Melody group were Corynebacterium pseudodiphtheriticum (1), HACEK(1), Haemophilus Aphrophilus(1), Streptococcus Viridans (2), Streptococcus Sanguinis(1); inadequate profylaxis had been present in at least 2 patients.
Survival free of endocarditis by Kaplan-Meier was for homografts 98% at 5 y and 86% at 20 y; Contegra 92% at 5 y and 62% at 10 y; Melody 77% at 5 y ($p < 0.001$).
The Melody conduit was sterilized successfully after 4-6 weeks IV antibiotics; 2 valves were obstructive at presentation: 1 valve was overstented at presentation (SBE then not diagnosed) and 10months later revalvulated with a new Melody; 1 valve had PS 42 mmHg, the other 4 valves functioned well after medical cure (PS <25 mmHg, PR < 2/4).
Conclusions: The Melody valved stent is significantly more vulnerable for endocarditis than a homograft or Contegra conduit. Optimal profylaxis might reduce the incidence, but adapted strategies will be required to obtain adequate longevity.



O10-4

Experience with the Cook Formula balloon expandable stent in congenital heart disease

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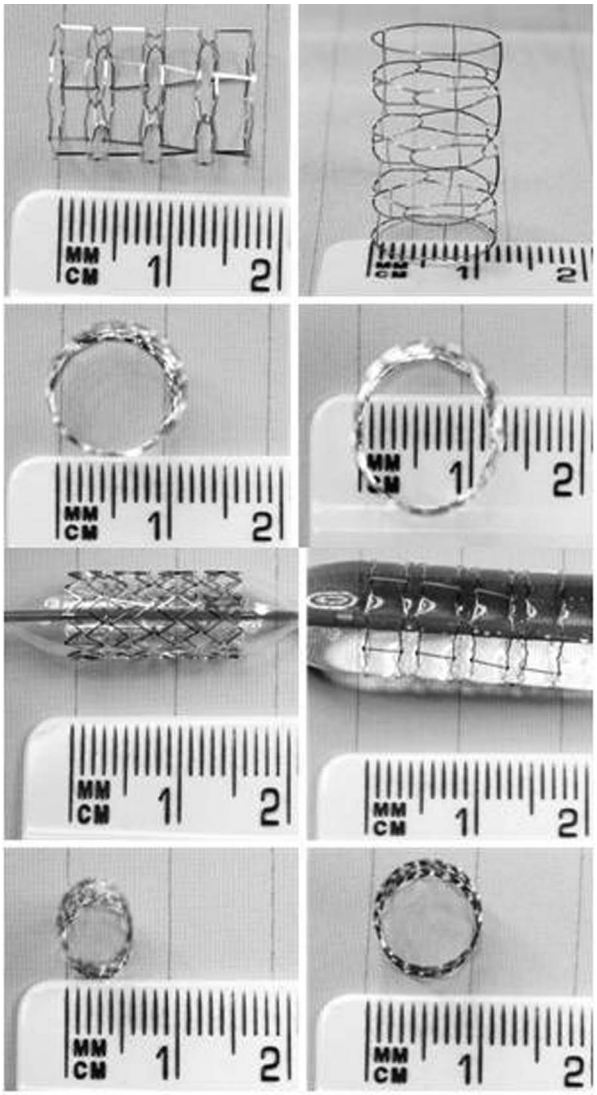
Introduction: Balloon expandable stents are an integral part in the catheter treatment of congenital heart disease. In the growing child, stents require dilatation to greater diameters over time. The Cook Formula stent is a recent 316 stainless steel open-cell design licensed for peripheral vascular work.
Methods: Following extensive ex-vivo studies, 95 stents were implanted in 84 children [median age 3.7 (0.02–16.6) years; median weight 13.4 (2.4-62.8) kg] over a two year period.



Results: Bench testing revealed that there was no stent shortening for dilatation to nominal diameter and beyond. The 5 mm stents could be dilated up to 10 mm, and the 10 mm stents to 20 mm. Stents were implanted through 4–7F sheaths or guidecaths over appropriate wires. Stent tracking and delivery was excellent.
Twenty-two stents were implanted in the right ventricular outflow tract in Fallot-type lesions, 47 for branch pulmonary artery stenosis (17 post cavopulmonary shunt/Fontan), 10 conduit stenosis, 8 Fontan fenestrations, 3 PDA in hybrid stage I Norwood, 4 in coarctation and 1 for SVC obstruction. 51 stents (54%) were overdilated. There were no stent fractures. Radial strength was very good, whereas stent conformability was limited.
Conclusion: The Cook Formula stent is a versatile pre-mounted balloon-expandable stent that can be significantly overdilated

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with virtually no shortening allowing for precise placement and minimal protrusion into adjacent vessels. This stent design is a great addition to the range of stents for use in the catheter treatment of complex congenital heart disease in children.

O10-5

One Year Follow-Up of the PREMIER Multicenter Registry for the Edwards SAPIEN Pulmonic Transcatheter Heart Valve: An Interim Report

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German Heart Center Munich, Germany (1); King Faisal Specialist Hospital, Riyadh, Saudi Arabia (2); Institute of Cardiology, Warsaw, Poland (3); Ospedale Pediatrico Bambino Gesù, Rome, Italy (4); Heart and Diabetes Centre North-Rhine Westphalia, Bad Oeynhausen, Germany (5); Royal Brompton Hospital, London, UK (6); Deutsches Herzzentrum Berlin, Germany (7); Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Hospital, Istanbul, Turkey (8); Children's Medical Center of Israel, Petah Tikva, Israel (9); Edwards Lifesciences, Irvine, USA (10); University Hospital Gasthuisberg, Leuven, Belgium (11)

Introduction: PREMIER is a single arm, multi-center registry assessing the safety and efficacy of the commercially available Edwards SAPIEN™ 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. One year follow-up has been completed for the first 99 patients and an interim analysis was carried out.
Methods: The Edwards SAPIEN™ Pulmonic 23 mm and 26 mm valves were implanted in the pulmonary position in 131 patients with a dysfunctional RVOT. Enrolment has been completed and patients are being followed up annually throughout 5 years. If valves were implanted before the registry initiation, data were recorded retrospectively starting with the first commercial implant at the site.
Results: The mean patient age was 27.3 ± 12.9 years, and 35.9% of patients were female. A total of 46 patients (35.4%) underwent prior conduit implantation, 12 patients (9.2%) underwent the Ross procedure, 41 patients (31.5%) underwent prior pulmonary valve repair. NYHA class was ⩾II in 63.0% of patients, pulmonary regurgitation was grade 3+ or 4+ in 75.2% of patients and the mean RVOT gradient was 39.5 ± 22.2 mm Hg. The procedural success rate was 92.4%. The mean procedure and fluoroscopy time was 179.0 ± 71.3 min and 38.1 ± 26.4 min, respectively. The survival at 1 year after the valve implantation was 100%. There were no valve stent fractures, no re-interventions or reoperations. Adverse events occurred in 19.1% of patients. Of the 40 adverse events reported, only 6 were considered serious with right pulmonary artery rupture treated with surgery being the most significant one. At 1-year, 68.9% of subjects with complete echocardiographic evaluation (51/74) had none/trace pulmonary regurgitation, 25.7% had mild and 5.5% (4/74) had moderate (n = 3) or severe (n = 1) pulmonary regurgitation. At 1-year, follow up the RVOT gradient decreased significantly by 14.9 ± 16.8 mmHg ($p < 0.0001$).
Conclusions: This interim reports suggests that the Edwards SAPIEN™ valve can be implanted safely in the pulmonary position with very low risk and significantly improve pulmonary regurgitation and RVOT gradient at 1-year. Further evaluation and long term follow-up is in progress to validate the clinical implications of this promising treatment for patients with dysfunctional RVOT.

O10-6

Pre and post operative characteristics of tetralogy of Fallot patients undergoing primary repair versus initial RVOT stent palliation

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Background: Repair of tetralogy of Fallot (TOF) after 3 months of age is associated with good post-operative outcomes. Right ventricular outflow tract (RVOT) stenting of the symptomatic or duct-dependent infant offers an alternative option to early repair or palliative shunt.

	Group 1A	Group 1B	Group 2A	Group 2B	Group 3
Number of patients	23	9	38	44	40
Age at surgery (days)	195 (160, 233)	188 (136, 241)	50 (41, 59)	22 (16, 28)	182 (170, 196)
Weight at surgery (kg)	6.4 (5.7, 7.1)	6.8 (5.5, 8.2)	4.1 (3.7, 4.5)	3.0 (2.8, 3.1)	6.8 (6.5, 7.2.)
Nakata Index (mean)	158 (137, 181)	144 (113, 177)	145 (128, 163)	148 (134, 164)	189 (165, 219)
Length of CICU stay (after surgery, days)	6.9 (3.3, 11.3)	7.7 (4.8, 10.4)	7.3 (5.7, 9.1)	14.0 (8.3, 23.3)	2.7 (2.2, 3.4)
Length of hospital stay (after surgery, days)	14.5 (8.7, 22.9)	19.8 (12.3, 30.1)	16.9 (12.2, 23.3)	23.5 (16.5, 35.6)	8.3 (7.0, 9.8)

Objective: To describe pre-operative characteristics and compare post-operative outcomes in patients after repair of TOF undergoing three different management pathways: palliative RVOT stent implantation, primary (early) repair at ⩽3 months of age and patients undergoing primary (conventional) repair ⩾3months of age.
Methods: This is a retrospective, single institution review between January 2000 and December 2013. Comparative groups were: TOF with pulmonary valve stenosis (TOF-PS-stent, group 1A), pulmonary atresia (TOF-PA-stent, group 1B) that underwent RVOT stenting; TOF-PS (group 2A) and TOF-PA (group 2B) that underwent early repair; conventional repair (group 3). Indications for stent implantation included infants with prostaglandin dependency or cyanotic spells deemed to be at higher risk for primary repair based on low weight, significant non-cardiac comorbidity and pulmonary artery hypoplasia.
Results: Thirty five patients underwent RVOT stenting with 58 stents being placed during 50 procedures. Patient characteristics at time of surgery, length of postoperative cardiac intensive care unit (CICU) and total hospital stay are outlined in the table (mean values and 95% CI are shown).
There was no operative or procedural mortality in any of the groups. In group 1A one patient died 10 days after surgery due to persistent biventricular failure, and two patients died from non-cardiac related causes after prolonged hospital stay. In group 2B two patients died on postoperative day 54 and 55 due to persistent cardiac failure. Acute complications in the early post-operative period were present in 53% in group 1A and 1B, 50% in group 2A, 63% in group 2B, and 35% in group 3. Rate of catheter re-interventions in the first 18 months after surgery was 23% in groups 1A and 1B, 26% in group 2A, 30% in group 2B and 2.5% in group 3.
Conclusion: Postoperative morbidity, mortality and follow up interventions are similar in pre-operatively RVOT stent placement patients and those with early primary repair.

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O11-1

Non-invasive measurements of energy expenditure and respiratory quotient in children on extracorporeal membrane oxygenation by respiratory mass spectrometry

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Objective: Extracorporeal membrane oxygenation (ECMO) provides temporary life-saving support for pediatric patients with severe cardiac failure, but causes metabolic disturbances and altered nutritional requirements. However, few studies have addressed the optimal energy supply to meet demand of these children largely due to technical difficulties with their invasive nature. We have adapted respiratory mass spectrometry to continuously measure O2 consumption and CO2 production across the ECMO oxygenator. This study aimed to assess energy expenditure (EE) and respiratory quotient (RQ) in children on ECMO.
Methods: Five children (age 0.3-36 months, median 20) were studied in day 1 to 6 on ECMO. EE and RQ were measured in sequential fashion at the child's native lungs and ECMO oxygenator using respiratory mass spectrometry. Measurements were collected at 4-hr intervals, with the means in 24 hours representing the values of each day. Each child's caloric and protein intakes were recorded for each day.
Results: In ECMO day 1-6, there was a small but significant increase in EE from 40 to 46 kcal/kg/d($p = 0.03$). In comparison, the caloric intake significantly increased by twice as much as the increase in EE from 30 to 61 kcal/kg/day ($p = 0.017$). As a result, RQ significantly increased from 0.6 to 1.0 ($p < 0.0001$). Protein intake significantly increased during ECMO day 1-6 from 0.5 to 1.5 g/kg/day ($p = 0.04$).
Conclusions: Respiratory mass spectrometry provides a unique, safe and precise technique to measure EE and RQ in patients on ECMO. Without this knowledge, both underfeeding and overfeeding would occur. Clinical nutritional treatment should be guided by directly measured energy and protein demands in this special group of critically ill children.

O11-2

Noninvasive measurement of cerebral hemodynamics and oxygen metabolism in neonates and infants with congenital heart defects

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Objectives: As a result of abnormal hemodynamics and/or reduced arterial oxygen saturation (SaO2) infants with complex congenital heart defects carry an increased risk of cerebral injury due to hypoxia and ischemia. Therefore protection of cerebral perfusion in the neonatal period, during and following surgical procedures is a major issue in the treatment of these children. A novel device O2C (Oxygen to see, LEA Medizintechnik) based on combined laser-Doppler flowmetry and photo-spectrometry allows for noninvasive transcranial monitoring of the regional cerebral oxygen saturation (rcSO2), relative amount of hemoglobin (rcHb) and relative capillary venous blood flow (rCBF). The aim of this study was to analyse whether online measurements of cerebral oxygen consumption are feasable and whether there are significant differences in cerebral oxygen supply in different subgroups of infants with complex congenital heart disease.
Methods: In 50 infants with acyanotic, biventricular cyanotic and univentricular congenital heart defects, SaO2, Hb, rcSO2, rcHb and rCBF were measured preoperatively. Cerebral fractional tissue oxygen extraction (cFTOE), arterio-cerebral difference in oxygen content (acDO2) and approximated cerebral metabolic rate of oxygen (aCMRO2) were determined.
Results: Infants with cyanotic or acyanotic congenital heart defects did not differ in rcSO2. In infants with biventricular acyanotic heart defects cFTOE was significantly higher than in infants with biventricular cyanotic and univentricular heart disease ($p < 0.05$). In infants with univentricular circulation rCBF was significantly lower as compared to patients with biventricular hearts ($p < 0.05$), while aCMRO2 was equal in all groups.
Conclusions: Under resting conditions regional oxygen saturation did not differ between acyanotic biventricular, cyanotic biventricular and univentricular congenital heart defects. Although cFTOE was increased among patients with biventricular acyanotic defects, higher levels of hemoglobin in infants with univentricular hearts and cyanotic biventricular heart defects resulted in aCMRO2, that did not differ significantly among the 3 groups. Nevertheless infants with univentricular circulation had the lowest rCBF and highest acDO2. According to these findings neonates with functionally univentricular hearts would be more vulnerable to minor changes in their hemodynamics. These findings provide an explanation for their increased cerebral vulnerability. According to our experience O2C offers interesting additional informations concerning cerebral perfusion as compared to NIRS.

O11-3

Tracheal reconstruction in patients with aortic arch anomalies and tracheomalacia

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Introduction: Developmental anomalies of the aortic arch, like double or right-sided aortic arch, may result in compression of the trachea and esophagus. Patients may present with neonatal stridor, respiratory distress and feeding difficulties. The treatment of choice is vascular decompression surgery.
We report our experience in the management of aortic arch anomalies using an adaptive approach of vascular decompression and tracheal reconstruction.
Methods: A retrospective analysis of data from 24 pediatric patients with aortic arch anomalies who underwent tracheal decompression surgery in our institution between 2008 and 2013 was conducted. Age, tracheal pathology, surgical technique, associated diseases, and follow-up data were analyzed.
Results: The patients were aged between 9 days and 16 years at the time of the surgery (median 3.2 yr), weight range was 2.3 to 64 kg (median 12.8 kg). Vascular pathologies included double aortic arch (complete vascular ring formation) in 12 patients, right-sided aortic arch in 10 patients and an aberrant subclavian artery in 2 patients. Tracheomalacia was diagnosed by tracheobronchoscopy in 8 patients. Accompanying heart defects comprised ASD, VSD and valvular anomalies. One patient was diagnosed with 22q11.2 microdeletion syndrome.

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All patients underwent vascular decompression surgery and, in the case of a congenital heart defect, corrective heart surgery. Based upon the findings of severe tracheomalacia during intraoperative tracheobronchoscopy, four patients underwent resection of the malacic segment and tracheal reconstruction during the same surgery.
Follow-up data were available for 17 patients for up to 5 years (median 17 months) and proved a clinically stable outcome. One patient died 15 months after the surgery due to complications of a congenital heart defect.
Conclusions: Although vascular decompression surgery is the current standard of care in patients with aortic arch anomalies and tracheal compression, this approach might not be sufficient in patients with concomitant tracheomalacia. We therefore recommend an intraoperative evaluation of the tracheal pathology. Patients with localized tracheomalacia may benefit from resection of the malacic tracheal segments with subsequent tracheal reconstruction during the same surgery.

O11-4

Reoperation for Right Ventricular Outflow Tract Obstruction After Arterial Switch Operation for TGA and Aortic Arch Obstruction

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Objective: Right ventricular outflow tract obstruction (RVOTO) is one of the reasons for late re-interventions after repair of transposition of the great arteries (TGA) with aortic arch obstruction (AAO). Aim of the present study was to identify predictors of reoperation for RVOTO in patients who underwent ASO and arch repair for TGA with AAO.
Methods: Since 1977, 394 ASOs were performed: TGA/IVS (243), TGA/VSD (119), Taussig-Bing (32). TGA and AAO was repaired in 42 patients (TGA/IVS 5, TGA/VSD 13, Taussig-Bing 24) with (coarctation 20, arch hypoplasia 5, coarctation and hypoplasia 10, aortic arch interruption 7). In these 42 patients operation reports and 2D-echocardiographic follow-up data were reviewed. We evaluated position of the great arteries, coronary artery anatomy, and diameters of RVOT, aortic valve annulus, aortic sinotubular junction, pulmonary valve annulus, and transverse aortic arch previous to ASO. Four patients were lost to follow-up, reliable echo data were available in 21. Cox proportional hazard models were performed to examine predictors of reoperation. Reinterventions for solitary supravalvular RVOTO were excluded.
Results: Median age at ASO was 19 (range: 1-4627) days. The RCA crossed the RVOT in 5 patients. AAO was repaired concomitant with ASO in 37 patients; in 5 coarctation was repaired at a median of 605 (range: 15-1093) days after ASO. Early mortality occurred in 6 patients, late in 4. Nine (TGA/VSD 2, Taussig-Bing 7) had 12 reoperations for (sub)valvular RVOTO. One patient died after reoperation. Taussig-Bing anomaly was a significant predictor of in-hospital mortality ($p = 0.029$) and reoperation ($p = 0.041$). Higher aortic valve Z-score significantly decreased the reoperation risk ($p = 0.021$).
Conclusions: After ASO and AAO repair, Taussig-Bing anomaly predicts a higher chance of mortality and reoperation for RVOTO. A higher aortic valve Z-score reduces the chance of reoperation for RVOTO. RCA crossing the RVOT complicates RVOTO relief.

O11-5

Decreased airway epithelial sodium transport in chronically hypoxemic patients with cyanotic congenital heart defect

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Introduction: Recovery from congenital cardiac surgery may be interfered by pulmonary complications such as pulmonary edema. Transepithelial lung edema reabsorption rests on airway epithelial sodium transport from the lung lumen towards the blood space through sodium channels (ENaC). ENaC comprises three homologous subunits (α, β, γ), and is inhibited by the diuretic agent amiloride. ENaC activity is impaired by hypoxia, which has been shown in vitro and in humans in high altitude pulmonary edema. The effect of chronic hypoxemia on airway epithelial ion transport, however, has not been studied.

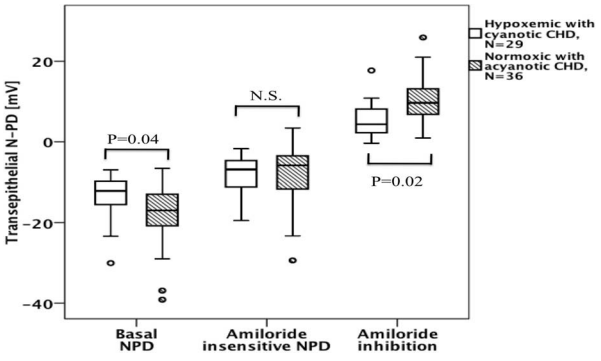

Figure 1.

Material and methods: We studied 26 hypoxemic (SpO2 ⩽85%) patients with cyanotic congenital heart defect (Cy-CHD) and 21 normoxemic (SpO2 ⩾95%) patients with acyanotic CHD (Acy-CHD). The activity of airway epithelial sodium transport was assessed by transepithelial nasal potential difference (N-PD) during locally perfused saline-solution ($N = 37$). After recording of the maximal basal N-PD, the contribution of ENaC to this resting N-PD was tested by inhibiting sodium transport with amiloride perfusion (10^{-4} M). Relative α-, β- and γ-ENaC mRNA expressions were determined by real time RT-qPCR in samples taken from nasal respiratory epithelium ($N = 41$). Data are given as mean ± SD or median with range, and statistical analyses were performed with T-Test or Mann-Whitney U Test, as appropriate ($P < 0.05$ considered statistically significant).
Results: Basal N-PD was significantly less negative in Cy-CHD (-12, range -21 – -7 mV) than in Acy-CHD (-15, range -24 – -7 mV) (Figure 1). Amiloride inhibitable sodium transport was significantly lower in Cy-CHD (4, range 0–11 mV) than in Acy-CHD (10, range 1–17 mV) (Figure. 1). Relative expressions of α-, β- and γ-ENaC, however, did not differ significantly between Cy-CHD (α 1.29 ± 0.35, β 0.90 ± 0.48, γ 1.21 ± 0.86) and Acy-CHD (α 1.13 ± 0.33, β 0.82 ± 0.34, γ 0.99 ± 0.50).
Conclusions: Our findings provide evidence that chronic hypoxemia impairs amiloride sensitive airway epithelial ion transport activity without significant effect on ENaC-subtype expression. Defective lung fluid reabsorption may predispose patients with cyanotic CHD and chronic hypoxemia to postoperative lung edema.

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O11-6

Long-term results of Ross procedure in a population-based follow-up

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Introduction: Ross procedure is commonly used in children requiring aortic valve replacement. The growth potential of the autograft and possibility to avoid anticoagulation therapy makes this procedure preferable for small patients. The purpose of this study was to evaluate long-term outcomes of Ross procedure in a nationwide follow-up.
Patients and methods: This retrospective study involved all children treated with Ross procedure in Finland between 1994 and 2009. The clinical records were reviewed for demographic and anatomical characteristics, Ross operation data, surgical history, and status at the latest follow-up. The median follow-up time was 11.5 (range 2.4-19.2) years.
Results: Fifty-one patients underwent either Ross (n = 37) or Ross-Konno (n = 14) procedure at a median age of 4.8 (range 0.02-16.3, 13 infants <1 year) years. Concomitant procedures (e.g. mitral valve surgery, VSD closure or aortic arch reconstruction) were performed in 17 (33%) patients. Before Ross operation, 28 (55%) patients had undergone aortic valve procedure and 4 patients had endocarditis. The indication for Ross procedure was aortic valve stenosis, regurgitation or mixed in 29%, 24% and 47% of patients, respectively. Early (<30 d) mortality was 5.9% (15.4% in infants) and late mortality 7.8% (30.8% in infants). During the follow-up, 20 reinterventions were needed in 16 (31%) patients. Two (3.9%) patients underwent heart transplantation. The most common (50%) cause for reintervention was pulmonary homograft stenosis, which was treated with valvuloplasty (n = 1), transcatheter pulmonary valve replacement (n = 3) or surgical homograft replacement (n = 6). At the last follow-up, mild to moderate aortic root dilatation was reported in 52% of patients, and two (5%) patients had undergone reconstruction of the ascending aorta. Trivial autograft valve regurgitation was commonly seen, but only one patient developed severe autograft regurgitation requiring mechanical valve replacement 15.9 years after Ross operation. Overall freedom from reintervention was 88% at 5 years and 80% at 10 years.
Conclusions: In long-term follow-up after Ross procedure, the most common reason for reintervention is pulmonary homograft stenosis. Aortic root dilatation and autograft valve regurgitation are relatively common but rarely lead to reinterventions before adulthood. In our center, Ross procedure has provided good long-term results in this challenging group of pediatric patients.

MP1-1

Fibrillin 1 gene mutations in development of Tetralogy of Fallot

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Objectives: The transforming growth factor β (TGF- β) signaling has an essential role in promoting development of the heart. Fibrillin-1 (FBN1) protein interacts with latent TGF-β binding protein (LTBP-1), which is the only form expressed during embryonic development, to control TGF-β activity. The genetic cause of Tetralogy of Fallot (TOF) is heterogenous. In this report we suggested that mutations in FBN1 gene play role in development of disease in patients with TOF.
Methods: We have performed mutation detection on 26 patients with TOF from the Pediatric Cardiology Department in Dr. Sami Ulus Children's Hospital. Total RNA extracted from peripheral blood lymphocytes with the High PureRNA Isolation kit (Roche Applied Science, Indianapolis, IN) was reverse-transcribed using Transcriptor First Strand cDNA Synthesis kit (Roche Applied Science, Indianapolis, IN), according to the manufacturer's recommendations. All coding exons of *FBN1* genewere amplified with the primers described previously [10]. The polymerase chain reaction products were purified and sequenced on an ABI PRISM 3130 automated DNA sequencer (Applied Biosystems).
Results: The median age of patients was 5.8 years (range between 9 months and 17 years) and male to female ratio was 4.2. We found an FBN1 mutation detection rate of 38.4% (n:10) in patients with TOF. Seven mutations were identified, of which 6 are reported here for the first time. The detected mutation types were 1536_1538ins3,1561_1563del3 (n:1), c.6314del66ins57 (n:3, 2 heterozygotes and 1 homozygote), heterozygote deletion of exon 64(n:2), heterozygote deletion of exon 57 (n:2), heterozygotep.K595E (c.1783A > G) (n:1) and heterozygote exon 13-14 deletion (n:1)
Conclusions: The mutations in FBN1 gene may be responsible for development of TOF in embryological period of cardiac development by effecting the function of long form of LTBP1L.

MP1-2

Left Ventricular Functions are Affected in Patients with Familial Mediterranean Fever

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Objectives: Familial Mediterranean fever (FMF) is an autosomal recessive disease that is prevalent among populations surrounding the Mediterranean Sea. The aim of this study was to assess early changes in regional and/or global systolic or diastolic myocardial functions of FMF patients without any cardiovascular symptoms using tissue dopplerechocardiography (TDE) and strain and strain rate echocardiography, and to compare them with the results of the control group.
Methods: This study was performed between May 2012–September 2012 at Pediatric Cardiology department of Dr Sami Ulus Obstetrics and Children Health and Diseases Training and Research Hospital. 45 patients with diagnosis of FMF and followed-up by Pediatric Nephrology Clinic were included.
Results: Among 45 patients with FMF, 24 (55.3%) were female and 21 (46.7%) were male, and age of them varied 2-18 years (mean 11.3 ± 3.7 years). There was no significant difference in terms of age, gender, height and weight between patient and control groups (p > 0.05). Mean disease durations were 4.6 ± 2.4 years (range 6 months–10 years). In the patient group, homozygote M694 V mutation was most common (64.4%) mutation type. All of the patients were using colchicine therapy. Mitral valve early filling velocity wave was higher in patients than

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the control group ($p < 0.05$). Sm wave of septal mitral annulus was lower in the patient group than the control group ($p < 0.05$). Septal MPI was higher in the patients than the control group ($p < 0.05$). Septal mitral annular IVCT and Am wave was lower in the patient group than the control group ($p < 0.05$). Patients with FMF were found to have statistically lower longitudinal global strain (GS), radial GS and strain rates (%-14.44 ± 4.77, % 14.80 ± 6.29 and 0.59 ± 0.24 srespectively) than those of the controls (%-17.40 ± 1.79, % 17.53 ± 4.63, 0.83 ± 0.51 s-) ($p < 0.05$). Circumferential GS did not differ significantly between groups.
Conclusions: As a result, subclinical FMF patients with normal cardiac functions may have normal left ventricular function measured with conventional echocardiography, miyocardial tissue were affected because of the disease (FMF), and this patients should be follow-up because of this situation.

MP1-3

Does vitamin D level affect cardiac ventricular functions in healthy children?

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Objective: We aimed to study the effects of vitamin D levels on right and left ventricular functions in healthy subjects.
Methods: We enrolled fifty-four healthy children who were admitted to our outpatient clinic between January 2013 and June 2013. Subjects were 3 to 24 months old. We chose subjects for whom vitamin D testing and complete transthoracic echocardiography with tissue Doppler imaging (TDI) for the current analysis. We then excluded patient with following features: septal hypertrophy, valvular disease, hypertension, chronic disease such as diabetes mellitus, chronic renal failure and chronic liver disease, as these finding could influence both vitamin D status and echocardiographic data. We also did not enroll subjects with rickets. We divided our study population into three groups according to the levels of vitamin D: subjects with >30 ng/ml 25 (OH) D were grouped as sufficient, subjects with 20-30 ng/ml 25 (OH) D were grouped as insufficient, and subjects with <20 ng/ml 25 (OH) D were grouped as deficient.
Results: Vitamin D deficiency was observed in 16 patients, and vitamin insufficiency was observed in 16 cases. The vitamin D level was within normal limits in 22 cases. As for the levels of vitamin D there was a significant difference among median values of sufficient group, insufficient group, and deficient group. Medial mitral annular Sm and Am velocities measured by TDI echocardiography was significantly higher in subjects with insufficient and deficient vitamin D level than those who has sufficient vitamin D. Right ventricle TDI echocardiography revealed that tricuspid isovolumic contraction time (IVCT) was significantly lower in subjects with insufficient and deficient vitamin D than those who had sufficient vitamin D ($p < 0.024$).
Conclusion: We have shown that left and right ventricular functions did not change at early stages of vitamin D deficiency. However, we have found that tricuspid IVCT, early marker of diastolic functions, was significantly decreased in subjects with insufficient and deficient vitamin D. To our knowledge, this is the first such study investigating the impact of vitamin D on left and right ventricular functions in healthy children.

MP1-4

Assessment of Left Ventricular Functions With Strain and Strain Rate Echocardiography in Children with Duchenne Muscular Dystrophy

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Introduction: Duchenne muscular dystrophy (DMD) is the most common hereditary neuromuscular disorder affecting individuals from all races and ethnic origins. Although the cardiac involvement may be subclinical, it may also present as cardiomyopathy (CMP) and heart failure. It has been reported that in spite of the preserved ejection fraction (EF) values in early stages of the disease; when the response to treatment is better, there might be actually occult systolic dysfunction
Methods: The aim of this study was to assess the myocardial functions of DMD patients using PW-DDI and S/SR echocardiography who had normal results in conventional echocardiography before, and to compare them with the results of the control group.
Results: In our study, 32 male patients with DMD whose mean age was 85.2 ± 38.4 months were compared with 31 healthy boys whose mean age was 89.0 ± 38.9 months. The EF and shortening fraction (SF) values of both DMD and control group subjects were within normal ranges. A statistically significant difference was found regarding the heart rate between the two groups ($p < 0.001$). Patients with DMD were found to have higher heart rate. In the measurements performed from the base of the interventricular septum, statistically significant differences were found between the Em, S amplitude and isovolumetric relaxation time (IVRT), myocard performance index (MPI) values of the two groups ($p < 0.05$). Besides, in the measurements made from the base of the left ventricular free wall, Em, S amplitude and IVRT, MPI values were shown to be significantly different ($p < 0.05$). In addition, the results of the S and SR measurements done from the base of the left ventricular free wall were significantly different between patients and control group ($p < 0.001$), and in the global strain measurement performed from the apical 4-chamber position, a significant difference was noted between the two groups ($p < 0.001$).
Conclusion: We suggest that when evaluating left ventricular functions in these cases detection of the subclinical dysfunction using techniques like S/SR echocardiography apart from the conventional echocardiography would be useful for the timing of treatment and follow up of the patients.

MP1-5

The impact of vitamin D status on arterial functions and carotid intima-media thickness in healthy children

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Introduction: Vitamin D deficiency is accepted as an independent risk factor for atherosclerosis. Several studies addressing the

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possible role of vitamin D deficiency in the pathogenesis of endothelial dysfunction and vascular stiffness. Arterial stiffening is an important cardiovascular risk factor and an independent predictor of all-cause and cardiovascular death. Increased aortic stiffness index (AoSI) or elastic modulus (AoEM) and/or decreased aortic distensibility (AoD) may suggest the widespread nature of the atherosclerotic process. Vitamin D deficiency is associated with increased carotid intima-media thickness (CIMT) and reported as an independent risk factor for atherosclerosis. In this study, we aimed to study the effects of vitamin D levels on CIMT, elastic properties of aortic artery and carotid artery in healthy subjects.

Table 1. CIMT and elastic properties of the carotid artery in three groups

	Vitamin D level (ng/ml)			
Variables	⩽ 20	21-29	>30	p Value
CIMT	0.44 ± 0.06	0.43 ± 0.06	0.44 ± 0.08	NS
Systolic carotid diameter (mm)	4.92 ± 0.57	4.87 ± 0.53	5.07 ± 0.60	NS
Diastolic carotid diameter (mm)	4.32 ± 0.52	4.25 ± 0.48	4.38 ± 0.55	NS
Carotid strain (%)	14.1 ± 6.3	15 ± 6.7	16 ± 7.3	NS
Carotid distensibility ($cm^2\ dyne^{-1}\times10^{-6}$)	8.2 ± 3.97	9 ± 5.5	9.2 ± 5	NS
Carotid stiffness index	4.3 ± 2.35	4 ± 2	3.9 ± 1.8	NS
Carotid elastic modulus ($cm^2\ dyne^{-1}\times10^{-6}$)	3.08 ± 1.6	2.88 ± 1.5	2.7 ± 1.2	NS

Data are shown as mean ± standard deviation. NS: Not significant.

Table 2. Elastic properties of the aortic artery in three groups

	Vitamin D level (ng/ml)			
Variables	⩽ 20	21-29	>30	p Value
Systolic aortic diameter (mm)	13.2 ± 1.97	12.8 ± 2	12.8 ± 1.6	NS
Diastolic aortic diameter (mm)	12 ± 0.2	11.7 ± 1.9	11.6 ± 1.7	NS
Aortic strain (%)	10.4 ± 4.5	10.3 ± 4.6	11 ± 6.2	NS
Aortic distensibility ($cm^2\ dyne^{-1}\times10^{-6}$)	6 ± 2.5	6 ± 2.7	6.5 ± 4.4	NS
Aortic stiffness index	5.31 ± 1.64	5.27 ± 1.57	5.5 ± 1.73	NS
Aortic elastic modulus ($cm^2\ dyne^{-1}\times10^{-6}$)	3.82 ± 1.33	3.81 ± 1.3	3.85 ± 1.4	NS

Data are shown as mean±standard deviation. NS: Not significant.

Methods: We enrolled fifty-four healthy children who were 3 to 24 months old. We chose subjects for whom vitamin D testing and complete transthoracic echocardiography with carotid artery imaging for the current analysis. We divided our study population into three groups according to the levels of vitamin D: subjects with >30 ng/ml 25 (OH) D were grouped as sufficient, subjects with 20-30 ng/ml 25 (OH) D were grouped as insufficient, and subjects with < 20 ng/ml 25 (OH) D were grouped as deficient.
Results: Vitamin D deficiency was observed in 16 patients, and vitamin insufficiency was observed in 16 cases. The vitamin D level was within normal limits in 22 cases. No statistically significant difference was found in the three groups regarding to CIMT, the elastic properties of the aortic and carotid arteries (Table 1 and Table 2).
Conclusion: In the present study, we have shown that CIMT and the elastic properties of the aorta and carotid artery did not change at early stages of vitamin D deficiency. To our knowledge, this is the first such study investigating the impact of vitamin D on carotid artery and aortic vascular functions in healthy children.

MP1-6

Treatment of Pulmonary Arterial Hypertension Using Sildenafil in Neonates with Bronchopulmonary Dysplasia

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Introduction: Sildenafil citrate, a phosphodiesterase-5-inhibitor, is a controversial treatment option for pulmonary arterial hypertension (PHT), a significant complication of bronchopulmonary dysplasia (BPD). There is scarce data in premature infants.
Objectives: Evaluate the use of sildenafil citrate in infants with BPD complicated by PHT.
Methods: Retrospective cohort study in a level 3 NICU (CHU mere-enfant Sainte Justine). All medical records of premature infants with PHT secondary to BPD treated with sildenafil citrate between January 2009 and May 2013 were reviewed. Primary outcomes were: 1-clinical response with 20% decrease in respiratory support score or FiO2 requirements; 2-echocardiographic response with decrease in tricuspid regurgitation gradient (at least 20%) or septal flattening (at least 1 degree).

Pharmacologic data (Median [IQR])	
Corrected age (weeks) at introduction of treatment	40 [28-54]
Dose (mg/kg/day)	4,4 [1-8]
Length (days) of sildenafil treatment	68 [2-857]
Time (days) to reach maximum dose	9 [2-39]
Benefits (n (%) or median [IQR]))	
Clinical response	8 (35)
Clinical response in first 48 hours	6 (26)
Clinical response in first 7 days	2 (9)
Echocardiographic response	16 (73)
Time (days) to echocardiograhic response	19 [2-312]
Side effects (n (%))	
Significant hypotension related to sildenafil	10 (44)
Other side effects (priapism and progression of ROP)	0 (0)
Other data (n (%))	
Survival	15 (65)
Death during treatment	5 (23)

Results: Twenty-three infants (61% male) with a median [IQR] gestational age of 26 wks [23-30] and birth weight of 710 g [480-1170] were included. Antenatal data is marked by presence of chorioamnionitis (17%), oligohydramnios (22%), intrauterine growth retardation (39%) and premature rupture of membranes (39%). Neonatal course included: confirmed sepsis (48%), grade 2 or more necrotizing enterocolitis (32%), stage 3 retinopathy of prematurity (ROP) (56%) and patent ductus arteriosus (78%).
Pharmacologic data, benefits and side effects are represented in the table.
Conclusions: Sildenafil citrate treatment for patients with PHT associated with BPD improves echocardiographic measurements in, more than 2/3 (73%) of patients despite only 1/3 (35%) of clinical response. Significant side effects are present (44% had hypotension). Further prospective studies are required to better assess efficacy of this treatment.

MP1-7

The efficacy and reliability of biological markers ın pediatric pulmonary hypertension

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Objective: The present study aims to determine the efficacy and reliability of serum markers such as brain natriuretic peptide (BNP), uric acid, troponin T, C-reactive protein, D-dimer, von Willebrand factor (vWF) and CA-125 in clinical assessment of pediatric pulmonary hypertension (PH).

Table 1. Serum Markers and Six Minute Walk Distance with respect to Functional Classification

	Class 1 (n = 2)	Class 2 (n = 18)	Class 3 (n = 17)	Class 4 (n = 3)	P
BNP (mg/L)	90.0 ± 42.4	513.5 ± 468.7	1190.2 ± 651.9	4666.7 ± 1527.5	0.001†*
Uric acid (mg/dl)	3.4 ± 0.4	4.6 ± 2.3	5.9 ± 2.3	7.0 ± 1.0	0.124
Troponin T (ng/ml)	0.003 ± 0.001	0.004 ± 0.003	0.007 ± 0.005	0.206 ± 0.131	0.003†*
CRP (pg/ml)	2.3 ± 1.0	4.3 ± 3.5	2.7 ± 1.9	4.9 ± 3.4	0.717
D-dimer (ng/ml)	17.5 ± 2.1	144.1 ± 140.1	115.0 ± 66.8	293.2 ± 240.0	0.408
vWF(%)	108.0 ± 10.0	113.4 ± 30.0	117.3 ± 26.0	143.3 ± 15.3	0.360
CA-125 (IU/L)	19.3 ± 10.9	22.6 ± 9.2	25.4 ± 21.2	215.1 ± 150.7	0.005†*
6 MWD (m)	460.0 ± 0.0	484.1 ± 91.6	376.3 ± 104.4	120.0 ± 14.1	0.001†*

*$p<0.05$ was accepted to be statistically significant.
†Statistically significant difference was detected between Class 1 and Class 4.

Table 2. Serum Markers and Six Minute Walk Distance with respect to Treatment Modality

Variables	Monotherapy (n = 20)	Combination treatment (n = 20)	P
Brain natriuretic peptide (mg/L)	511.6 ± 490.5	556.0 ± 432.1	0.046*
Uric acid (mg/dl)	5.2 ± 2.5	5.2 ± 2.1	0.990
Troponin T (ng/ml)	0.006 ± 0.005	0.081 ± 0.023	0.041*
C-reactive protein (pg/ml)	4.0 ± 3.2	3.2 ± 2.7	0.978
D-dimer (ng/ml)	133.5 ± 111.9	135.5 ± 104.4	0.361
Von Willebrand factor (%)	113.7 ± 21.0	118.5 ± 33.2	0.966
CA-125 (IU/L)	22.9 ± 17.7	84.4 ± 43.7	0.037*
Six minute walk distance (m)	405.0 ± 118.8	294.7 ± 145.8	0.044*

*$p<0.05$ was accepted to be statistically significant.

Methods: This is a retrospective analysis of 40 children with PH in aspect of serum markers. The diagnosis of PH was made when mean pulmonary artery pressure was measured ⩾25 mmHg by catheter angiography.
Results: Congenital heart diseases were the most frequently encountered etiological factor for PH during childhood. As functional class advanced, serum concentrations of BNP, troponin T and CA-125 increased significantly and six minute walk distance decreased significantly (Table 1). There was a significant and negative correlation between six minute walk test and each of the variables including serum BNP, CA-125 and vWF levels ($p = 0.008$, $p = 0.016$ and $p = 0.019$ respectively). When compared with children receiving monotherapy for PH, the children who were on combination treatment had significantly shorter six minute walk distance ($p = 0.044$) as well as significantly higher concentrations of BNP, troponin T and CA-125.
Conclusion: Serum markers including BNP, troponin T, vWF and CA-125 are related with six minute walk distance and functional class which reflect the severity of PH at pediatric age. The combination of these serum markers may stand for a simple, cheap and non-invasive tool for clinical workup of PH during childhood.

Keywords: biological marker; child; functional class; pulmonary hypertension

MP1-8

In vitro analysis of the mechanisms of intravenous immunoglobulin and prednisolone for the prevention of coronary artery abnormalities in Kawasaki disease

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Objectives: Kawasaki disease (KD) is an acute febrile vasculitis of childhood. Coronary artery abnormalities (CAA) are a major problem and high dose intravenous immunoglobulin (IVIG) plus prednisolone (PSL) are effective and reducing the occurrence of CAA. However, the pathogenic mechanism of CAA remains to be well elucidated. We investigate the relationships between CAA and endothelial cells' proliferation or apoptosis, and the effects of IVIG and/or PSL using Human umbilical vein endothelial cells (HUVECs) stimulated with sera of KD patients obtained before and after IVIG treatment.
Methods: 32 KD patients and 10 controls with bacterial infections were enrolled. CAA z-score was measured by two-dimensional echocardiography, using the Japanese normal values of coronary artery dimensions as previously described. Third passaged HUVECs were cultured with KD patient's sera before IVIG treatment or control's sera for 24 h. Subsequently HUVECs cultured with KD patient's sera were left untreated or treated with IVIG and/or PSL for next 24 h. Co-cultured HUVECs were assessed by MTT assay and analyzed Akt phosphorylation by Western blotting.
Results: MTT assay indicated loss of viability with exposure to sera from KD patients before IVIG treatment compared to controls (0.21 ± 0.09 vs. 0.30 ± 0.04, $p < 0.05$). MTT assay also indicated cell's multiply after IVIG treatment in KD patient compared to those of before IVIG treatment. The decrease in absorbance value in MTT-assay showed a negative correlation with the CAA z-score ($r = 0.593$, $p < 0.05$). The decrease in the phosphorylation of Akt was present in KD patient's sera with CAA. Treatment of stimulated cells with sera from KD patients before IVIG treatment, Treatment of IVIG or PSL alone led to improvement in cell viability compared to untreated group (IVIG 0.27 ± 0.12, PSL 0.26 ± 0.14, untreated 0.21 ± 0.09, $p < 0.05$). In addition, the combination treatment with IVIG plus PSL had synergistic suppressive effect of loss of viability in MTT assay (IVIG + PSL 0.32 ± 0.14, $p < 0.05$).
Conclusions: Impaired endothelial cells might affect the subsequent impaired activation of PI3K/Akt pathway and endothelial dysfunction those are thought to be causally related to development of CAA. Our results suggest that IVIG and/or PSL effect cell viability and reduce the coronary outcomes in KD patient.

MP1-9

GH Therapy Increases the Prevalence of Aortic Dilation in Patients with Turner's Syndrome

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Introduction: Dilatation of the ascending aorta (AoD) is described in Turner's syndrome (TS) with variable prevalence according to different sources (6.8%-32%). Reported series typically include patients with associated cardiac anomalies, e.g. aortic coarctation, left outflow tract obstruction and bicuspid aortic valve.

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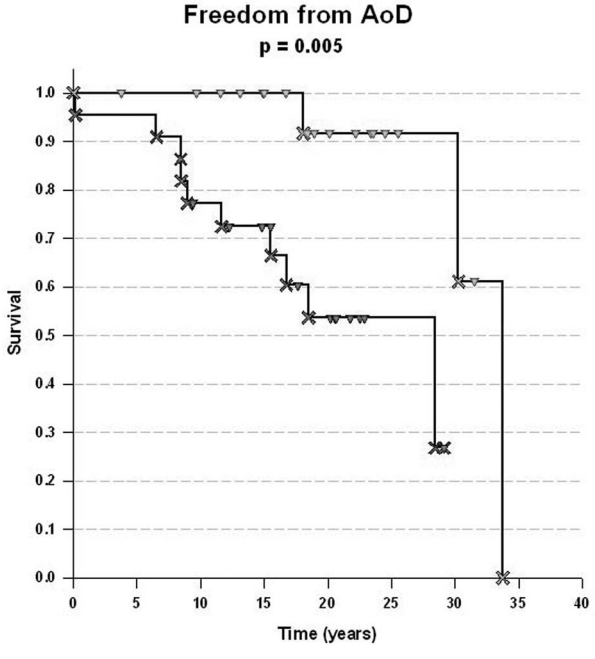
Objective: To characterize the prevalence, age of onset and the progress of AoD in TS patients free of structural cardiac anomalies. Potential risk factors, such as karyotype and growth hormone (GH) therapy, were analyzed for correlation with AoD.
Methods: Retrospective study with data collected from medical records and echocardiography studies. TS patients followed between 1992 and 2010, free of structural cardiac malformations were eligible when they had at least 2 echocardiography studies. Patients with previous cardiac surgery or those under anti-hypertensive medication were excluded. Ascending aorta diameter measurements were collected for all patients and adjusted for body surface area based. Regression equation was derived from our institution's echo laboratory from 1300 healthy children. AoD was defined as a Z-score >2.
Results: From 120 patients, 33 were excluded due to associated cardiac anomaly, and 15 due to anti-HTA medication. Another, 28 patients were excluded due to incomplete data.
The study population consisted of 44 patients, aged 11.9 ± 7.4 years at the first echocardiogram and 17.9 ± 7.3 years at last follow-up, with a follow-up duration of 6.0 ± 3.7 years. 13 (29.5%) patients exhibited an AoD during the follow-up period suggesting an actuarial estimate of the freedom from AoD dropping from 86%, 70%, and then 37% at 10, 20 and 30 years old respectively. Increased prevalence of AoD was associated with GH therapy (n = 23) compared to non treated patients (n = 21) 39.1% vs 9.5% (p = 0.036) and decreased survival with freedom of AoD (p = 0.005). In contrast, there was no statistically significant impact of karyotype (X0 v.s. mosaic; p = 0.38).
Conclusions: The prevalence of AoD in TS free of structural aortic anomalies is comparable to published data with associated lesions. GH therapy increases significantly the likelihood of AoD irrespective of karyotype.



MP1-10
Pulmonary hypertension in the preterm infant with bronchodysplasia can be caused by pulmonary vein stenosis: a must-know entity
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Background: Pulmonary vein stenosis is a rare cardiac defect. It has been described associated to other congenital heart defects and to prematurity. Haemodynamically it typically causes postcapillary pulmonary hypertension but a precapillary component can be observed.
Objective: Retrospective analysis from 1998 till 2012 in two French pediatric congenital cardiac centers of all premature infants with pulmonary vein stenosis. Diagnostic modalities, haemodynamics, pulmonary vein anatomy and outcome are described.
Results: 15 premature infants <37 weeks of gestation with the diagnosis of pulmonary vein stenosis were identified. Median gestational age at birth was 28 + 5 weeks (range: 25 + 5–35 weeks). Median birth weight was 790 g (range: 585-1500 g). Nearly all the infants (86%) had bronchodysplasia. 27% (n = 4) had associated cardiac defects other than persistent arterial duct or secundum atrial septal defect. Six infants (40%) had a first cardiac catheterization for exploration of pulmonary hypertension without visualization of pulmonary vein stenosis. In 73% of infants the diagnosis of pulmonary vein stenosis was suspected by echocardiography during follow-up. The remaining infants were diagnosed during cardiac catheterization, by pulmonary computer tomography and one during cardiac surgery. Median age at diagnosis was 6.8 months (range: 1.5-71 months). The majority of infants (60%) had initially unilateral pulmonary vein stenosis affecting in 89% one of the left pulmonary veins. Median initial mean pulmonary artery pressure (PAP) at diagnosis was 40 mmHg (range: 24–70 mmHg). Treatment modalities included: surgical intervention (pulmonary venoplasty, sutureless, lobectomy, heart and lung transplantation) for 6 patients, decision of non-intervention in 6 patients, interventional cardiac catheter (percutaneous pulmonary vein dilatation) in 2 patients and additional medical treatment for pulmonary hypertension in three patients. Overall mortality was high with only 46% of patients still alive at latest follow-up (median: 6 years; range: 1.2-10.9). Median follow-up until death was 7.2 months (range: 3.6-12.1 months).
Conclusion: Pulmonary vein stenosis is an unusual cause of pulmonary hypertension in the premature infant with bronchodysplasia. Diagnosis can be difficult since initial echocardiography can be normal and the disease progressive. The diagnostic method of choice is cardiac catheterization. Treatment options are numerous by surgical or interventional means but prognosis remains extremely poor.

MP1-11
Pulmonary hypoplasia associated to congenital heart diseases: a foetal study
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Background: Abnormalities of the fetal pulmonary vasculature may alter lung morphogenesis. Postnatal studies have suggested that

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pulmonary hypoplasia (PH) may be associated to congenital heart diseases (CHDs). However, no fetal study have been carried out to determine whether these abnormalities have a common developmental origin, early in morphogenesis, or wether they gradually become established after birth.
Objective: To determine the prevalence of PH associated with CHDs, and to identify the types of CHDs associated with the highest risk for lung growth impairment.
Methods: Between January 2006 and December 2010, fetuses with CHD obtained following the termination of pregnancies due to fetal abnormalities were examined in a prospective manner for the detection of heart and lung defects. CHDs were classified into five pathophysiological groups. PH was defined as a fetal lung weight to body weight ratio <0.015 before 28 weeks, and <0.012 after 28 weeks. The expression of CD31 and VEGF in lungs was evaluated by immunohistochemistry.
Results: PH was detected in 15 of the 119 fetuses analyzed (13%). It was significantly associated with CHD with right outflow obstruction, independently of chromosomal abnormality and associated extra-cardiac anomaly ($p < 0.03$). Right outflow obstruction was present in 60% of fetuses with CHD and PH, but in only 32% of those with CHD but no PH. In fetuses with right outflow obstruction, no difference was observed between those with PH and those without PH, in terms of the ratio of the pulmonary artery diameter to the aortic diameter, lung CD31 expression, or lung VEGF expression.
Conclusion: CHDs with right outflow obstruction are a significant risk factor for prenatally acquired PH. The occurrence of fetal PH is not correlated to abnormalities of the pulmonary vasculature, suggesting the involvement of perfusion-independent mechanisms.

MP1-12
Hybrid procedure to postpone succesful biventricular repair in left heart obstructive lesions with borderline left ventricle
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Background: In neonates with small left heart lesions, the decision of single ventricle vs biventricular circulation is difficult. Critical morphologic criteria (Rhodes score/LV adequacy score <0.35, aortic annulus <6 mm, mitral annulus <9 mm, LV volume <25 ml/mm2) or physiologic considerations (antegrade flow in ascending aorta, bidirectional shunt at ductal level) were defined to predict risk/success for biventricular repair. To postpone this decision and use the growth potential of left heart structures, neonatal hybrid palliation (bilateral PA banding, stenting duct) can be performed in high risk neonates.
Aims: Evaluate results of biventricular repair in high risk neonates after neonatal hybrid palliation for left heart obstructive lesion with small left heart.
Patients and Methods: Retrospective study, n = 7, mean birthweight 3,3 kg. Diagnoses: critical coarctation, arch hypoplasia n = 5, +VSD in 2, parachute MV and mild MS in 1; critical aortic stenosis n = 2,+coarctation in 1. All had duct dependent systemic circulation. Mean aortic annulus 4.6 mm (4-6 mm), mitral annulus 6.7 mm (3,7-10 mm), LV volume 20.2 ml/m2 (14-33 ml/m2), LV adequacy Rhodes score -2.8, precluding biventricular repair. Hybrid palliation was performed: bilateral PA banding and stenting of the arterial duct, +/- atrial septostomy (n = 3).
Results: Neonatal hybrid palliation was successful in all 7 patients obtaining adequately balanced circulations. In 2 patients with critical AS a balloon valvuloplasty was performed. At a mean age of 6.2 mths left heart structures had grown: mean aortic annulus 8,7 mm, mean mitral annulus 9,8 mm, LV volume 25 ml/m2. Biventricular repair was achieved in all patients: aortic arch reconstruction, PA debanding, duct closure (n = 7)+VSD closure in 2 pts.
Conclusions: Hybrid procedure allows to postpone the decision of uni- versus biventricular repair in small left left heart lesions. Loading conditions after birth with increased LA en LV filling may alter LV volume and size of LVOT and MV, so that successful biventricular circulation can be achieved at an older age with lower risk.

MP1-13
Toxic Metals in Children's Heart Tissue
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Objectives: investigation of toxic metals content in heart tissues of children with and without congenital heart diseases (CHD).
Methods: We had divided children in 2 groups. In first group we had included 46 children (aged from 14 days to 17 years) and 2 fetus with various cardiovascular malformations. The second group were 19 children and fetuses without CHD, died from different reasons. All patients were examined by the spectral analysis of Al, Cd, Pb, Hg, Be, Ba, Tl, Bi, As, Ni, Sb, Sn, Sr, Ti, W, Zr, Ag, Li, B, Co, Si, V in intraoperative and autopsy biopsies of endocardium, myocardium, pericardium, aorta, pulmonary artery, ductus arteriosus by methods of the atomic emission spectrometry in the inductively coupled plasma and atomic absorption spectrometry with electrothermal atomization.
Results: We revealed the presence of toxic metals in heart tissue in $85,4 \pm 5,1\%$ ($p < 0.05$) of patients with cardiovascular malformations and in $47,4 \pm 11,5\%$ children without CHD. Patient of first group had exceed of acceptable level of barium, aluminum, lithium, nickel, strontium, arsenic, zirconium, lead; patients of second group—barium, aluminum, nickel. We had determined that children with cardiovascular malformation significantly ($p < 0.05$) more frequently in comparison with patients without CHD had Ba, Al, Li and Sr in heart. In children with CHD we discovered dependence of the toxic metals level concentration and the topography of biopsy: at the location of the malformation – in locus of aortic coarctation, valve atresia, in septum of septal defects open ductus arteriosus it concentration was significantly ($p < 0.05$) higher than in other heart and vessels areas. Children with elevated toxic metals level in heart tissue significantly frequently ($p < 0.05$) had combined CHD.
Conclusions: The above mentioned is the basis for suggestion about possible barium, aluminum, lithium, and strontium, role in cardiogenesis violation in humans. Future development of this methods may possible serve as addition diagnostic tool in detection of congenital heart diseases in fetus and kids.

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MP1-14

Plastic bronchitis after extracardiac total cavopulmonary connection

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Introduction: Plastic bronchitis (PB) is a severe complication in patients with Fontan circulation. The incidence of PB is low and only few reports are available. The aim of our study was to investigate the perioperative factors at the time of total cavopulmonary connection (TCPC), which might prove predictive for the later onset of PB; as well as the clinical status and the medical course of the patients with PB.
Method: A retrospective cohort study included all patients (Jan 2000–Dec 2012) who received TCPC, using an extracardiac conduit for a functionally univentricular heart at the German Heart Centre Munich. Patients who developend PB during follow-up were included in "PB group" while the remaining patients constituted the "non-PB" group.
Results: The prevalence of PB at our institution was 2,4% (8/333). The median time interval between the TCPC and the first diagnosis of PB was 29 months (range 2 weeks—113 months). At the time of TCPC, there was no significant difference in preoperative weight, age, central venous pressure, transpulmonary gradient and arterial—or venous oxygen saturation between the two groups. The occurrence of post-operative chylothorax was significantly higher in the PB group (7/8 in the PB group vs. 64/325; $p < 0.001$), as well as the occurrence of diaphragm paresis (4/8 in the PB group vs. 25/325; $p < 0.001$). On hospital admission for symptomatic PB 6/8 patients had an underlying problem leading to impaired hemodynamics: severe aortic valve regurgitation in two patients, severe left pulmonary artery (LPA) -stenosis in four patients. One patient developed cardiac decompensation after pneumonia and in one patient no detectable cause for PB was found. One patient received and aortic valve replacement, four were treated interventionally by LPA-stenting, while the remaining patients received a conservative treatment. During follow-up (median time 18 months (range 5-32 months), six patients are free of PB symptoms, one patient died and one patient is symptomatic with PB.
Conclusions: Post-operative chylothorax and diaphragm paresis at the time of TCPC were significantly more frequent in patients who later developed symptomatic PB. Compromised hemodynamics was found in nearly all symptomatic patients.

MP1-15

Long standing cyanosis in congenital heart disease does not cause diffuse myocardial fibrosis

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Introduction: The assumption of the presence of diffuse myocardial fibrosis in long standing cyanotic congenital heart disease (CHD) inspired us to noninvasively determine the myocardial extracellular volume (ECV) using contrast CMR (T1 mapping).
Methods: T1 maps were measured pre and 3-10 minutes after the infusion of 0.15 mmol/kg of gadolinium on 25 subjects. Seven adult patients with longstanding cyanotic CHD and no previous surgical history (aged 16-53 yrs and oxygen saturations of 69-90%), nine normal subjects (aged 14-49 yrs), and nine patients with previously cyanotic CHD after total repair during which a heart lung machine was used (aged 2 months-58 yrs). Images were obtained in a mid-ventricular short axis plane. Late gadolinium enhancement using the phase sensitive inversion recovery (PSIR) sequence was performed to exclude scar areas. The T1 values were measured in two areas of the myocardium, in the septum and in the left ventricular posterior or inferior wall, such that same areas were assessed in every patient in the pre and post contrast T1 scan. ECV was calculated according to (1-hematocrit)*(ΔR1myocardium/ΔR1blood).
Results: Patients with cyanosis had significantly lower ECV percentage than the previously cyanotic patients after total repair (septum: $22 \pm 2\%$ vs $35 \pm 12\%$, $p = 0.01$; LV wall: $22 \pm 2\%$ vs $30 \pm 7\%$, $p = 0.02$, respectively). No significant differences were found between patients with cyanosis and normal controls (septum: $22 \pm 2\%$ vs $24 \pm 1\%$, $p = 0.14$; LV wall: $22 \pm 2\%$ vs $24 \pm 2\%$, $p = 0.21$, respectively). ECV values were significantly different between the three groups in both septum and LV wall ($f = 0.007$ and $f = 0.02$, respectively).
Conclusions: Long standing cyanosis in congenital heart disease without cardiac surgery does not cause diffuse myocardial fibrosis or expansion of the myocardial extra cellular volume.

MP1-16

Preoperative dilatation of the right pulmonary artery is a risk factor for refractory respiratory complications late after the definitive repair of Tetralogy of Fallot with absent pulmonary valve

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Background: Refractory respiratory complication is not a rare condition late after definitive surgery of Tetralogy of Fallot with absent pulmonary valve (TOF/PVA). Here we evaluated perioperative risk factors for the post-operative respiratory complications in TOF/PVA patients.
Methods: Twenty-one patients with TOF/PVA who had underwent the definitive repair (1987-2012) were enrolled in this study. Plication of the pulmonary artery (PA) was performed in 19 patients. Cardiac catheterization was examined before surgery in 15 patients. We evaluated preoperative conditions of the patients as follows; age at surgery, sex, hemodynamic data in cardiac catheterization and those in echocardiography. We also evaluated the right and left pulmonary arterial area index (RPAI, LPAI) using cine-angiography; π*(right or left pulmonary diameter /2)^2/body surface area. Clinically significant respiratory complications are defined as; 1) repeated admission due to respiratory infection and 2) mechanical ventilation over one month.
Result: One patient died during perioperative period and one died late after the operation. Overall survival rate was 86% in 20 years. Eight patients (38.1%) including two mortalities developed significant respiratory complication. One patient underwent tracheostomy and mechanical ventilation, seven patients needed unscheduled hospitalization due to repeated respiratory infections. These all eight patients had preoperative respiratory failure and required definitive surgery in the first year of life.

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Preoperative respiratory failure was a risk factor ($p = 0.04$). Larger RPAI was also a significant risk factor of postoperative respiratory complication compare with those who had no respiratory complication (RPAI: 1371 ± 616 vs. 856 ± 298; $p = 0.044$). Post-operative PAI did not show any significant difference in patients with or without post-operative respiratory complication ($p = 0.657$). None of other perioperative factors including type of reconstruction of right ventricular outflow tract, preoperative LPAI and postoperative RPAI, data in echocardiography had a significant correlation with the post-operative respiratory complication.
Conclusions: Survival after the definitive repair for TOF/PVA was satisfactory. The postoperative respiratory complications were correlated to the preoperative respiratory failure and the dilatation of the right pulmonary artery. The post-operative outcomes could be improved by earlier surgical interventions before marked dilatation of the right pulmonary artery and severe respiratory disturbance develop.

MP1-17

Does exercise training improve cardio-respiratory fitness and daily physical activity in adolescents with corrected tetralogy of Fallot or Fontan circulation? A randomized controlled trial

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Introduction: Current public-health guidelines suggest >60 minutes of moderate-to-vigorous daily physical activity for adolescents. Many adolescents with congenital heart disease do not meet these guidelines. We hypothesized that exercise training would increase cardio-respiratory fitness and daily physical activity. We assessed the effects of an exercise-training program on cardio-respiratory fitness and daily physical activity in patients with corrected tetralogy of Fallot (ToF) or Fontan circulation.
Methods: A multi-center prospective, randomized controlled trial design was used. Patients with ToF or Fontan circulation (age range 10-25 years) were randomized. The intervention group participated in a 12 week standardized aerobic exercise-training program. The control group did not change their lifestyle. Cardiopulmonary exercise-testing and activity measurements were performed before and after a 12 week period.
Results: Nine-three patients participated, 56 in the exercise group. 37 in the control group. PeakVO2 increased in the exercise-group by 5.0% (1.7 ± 4.2 ml/kg/min, $p = 0.011$), in the control-group by 2.7% (0.9 ± 5.2 ml/kg/min, p = not significant) (p = not significant between the groups). Increase in peak workload was significant (6.9 ± 11.8 Watt vs. 0.8 ± 13.9 Watt; $p = 0.047$) Time spent in moderate to very vigorous activity at baseline was 13.6 ± 8.6%, which did not significantly change after training.
Subgroup analysis showed a significant increase in the pre-to-post peakVO2 in ToF, not in Fontan patients.
Conclusions: Aerobic exercise training improved cardio-respiratory fitness in patients with tetralogy of Fallot but not in patients with a Fontan circulation. Exercise training did not significantly change daily physical activity.
Funding: N Duppen was supported by research grant from the Dutch Heart Foundation, (grant 2008B026)

MP1-18

Clinical Evaluation of Intracardiac and Major Vessel Thrombosis in a Pediatric Cardiology Unit: A Single-Center Experience from Turkey

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Introduction: Experience in managing intracardiac and large vessel thrombosis in pediatric cardiology and cardiovascular surgery is limited, particularly concerning r-TPA usage. In this study, we evaluate the data gathered in our center on patients with intracardiac and major vessel thrombosis.

Table 1. Locations of cardiovascular thrombi (*Some patients had multiple locations)

Thrombus Location	n*
Total	27
Right heart	11
- Atrium/Ventricle	9/2
Left heart	7
-Atrium/ Ventricle	1/6
Located in the vessels	9
Major vessels associated with the heart	5
- Pulmonary artery bifurcation	3
- Pulmonary vein opening	1
- Inferior vena cava	1
Peripheral major vessels	4
- Popliteal/Brachial artery	1/3

Methods: Enrolled in the study were 20 patients that were admitted to our hospital between January 2010-December 2013 and diagnosed with intracardiac and large vessel thrombosis. We evaluated demographic findings, primary pathologies, treatment and complications.
Results: We assessed 25 thrombus attacks that occurred at different times in 20 patients (8 female, 12 male). Mean age was 7.4 ± 7.3 years (1.2 months-22 years). Thrombus locations are summarized in Table 1. Four patients had sequential thrombosis. Regarding concomitant cardiac pathologies, 10 patients (50%) had complex congenital heart disease, 7 (35%) dilated cardiomyopathies (4 of them secondary to noncompaction), 2 (10%) arrhythmogenic right ventricular dysplasia, and 1 (5%) hypertrophic non-obstructive cardiomyopathy. At diagnosis, left ventricular function was reduced in 7 patients (35%). Fractional shortening was reduced all of the patients (n = 6)

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with left ventricular thrombi; mean fractional shortening was 17.8%(10-24%). In 7 patients (28%), thrombosis was associated with cardiac surgery. Three of them (12%) (supracardiac TAPVD, cor triatriatum sinister, Taussig-Bing anomaly) developed thrombus in the early postoperative period and 4(16%) in the late period.
Regarding treatment, r-TPA was used in 9 patients (36%) with thrombi in the left heart (n = 7) or in the peripheral artery (n = 2). An unfraxioned or low molecular weight heparin (LMWH) was used in the rest. Two patients that developed cerebral thromboembolism during acute treatment later recovered without major sequelae by maintaining appropriate anticoagulation. Warfarin, aspirin, clopidogrel and LMWH were used in maintenance treatment. Mean treatment duration was 83 ± 107 days (7-510); mean follow-up period was 265 ± 220 days (6-948 days). Treatment was successful and thrombi regressed in 92% of cases. The patient with brachial artery thrombus underwent distal upper extremity amputation. None of the patients died.
Conclusions: Intracardiac thrombi in major vessels associated with the heart can be treated successfully with anticoagulants and thrombolytics. Potential embolic complications can be recognized early and long-term sequelae can be reduced with close monitoring throughout the treatment period.

Table 1. Classification of LVNC Patients

LVNC Category	n (%)
● Total	50 (100)
● Isolated	29 (58)
● With Congenital Heart Disease	21 (42)

*Acyanotic	13	*Cyanotic	8
– VSD	4	– TA, VA concordance, VSD	2
– MVP, myxomatous MV, Cleft, MI	2	– c-TGA, VSD, PS	1
– VSD, ASD	2	– Critical PS, PDA	1
– PDA	2	– SSD, PA, ARV/PA, VSD	1
– BAV, PDA	1	– TA, RVH, VSD, ASD, PDA	1
– BAV, AS, AI	1	– PA/IVS	1
– ASD, Ebstenaoid TV, TI	1	– Unguarded TV, severe PI,	1

BAV: bicuspid aortic valve, **AI**: aortic insufficiency; **AS**: aortic stenosis; **VSD**: ventricular septal defect; **MI**: mitral insufficiency; **PA**: pulmonary atresia; **PS**: pulmonary stenosis; **PH**: pulmonary hypertension; **PI**: pulmonary insufficiency; **TV**: tricuspid valve; **TI**: tricuspid insufficiency; **TA**: tricuspid atresia; **c-TGA**: corrected transposition of great artery; **RVH**: right ventricular hypoplasia; **VA**: ventriculoarterial; **SSD**: situs solitus dextrocardia; **ARV/PA**: aorta arising from right ventricle with pulmonary atresia, **PA/IVS**: pulmonary atresia with intact ventricular septum

MP1-19
Left Ventricular Noncompaction in Childhood: Four Years Single Center Experience from Turkey

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Introduction: Left ventricular noncompaction (LVNC) is a specific cardiomyopathy that occurs following a disruption of endomyocardial morphogenesis. This study presents clinical findings, diagnostic features, treatment and follow-up of pediatric LVNC patients.
Methods: Enrolled in the study were patients that applied to our clinic between January 2010 and December 2013 with isolated LVNC or LVNC with congenital heart disease (CHD). We evaluated patients for rhythm disorders, and in cases with CHD, for surgery and complications.
Results: Study population was comprised of 50 patients, 24 male (48%). LVNC was isolated in 29 (58%) and with CHD in 23 (46%). Clinical features are summarized in Table 1. Mean age at diagnosis was 4.35 years ± 88 months (4 days-19 years). Findings at admission were heart murmur in 28 (56%), congestive heart failure in 10 (20%), dyspnea in 5 (10%), cyanosis in 4 (8%), growth and developmental retardation and pneumonia in 3 (6%). Echocardiography revealed a shortening fraction of 29 ± 11% (9–50%), ejection fraction of 52.7 ± 19.3 (14-80), and LVDD of 34 ± 13 mm (17-64 mm). Noncompaction was apical in 9 patients (18%), free wall in 14 (28%), apical and free wall in 20 (40%), biventricular in 2 (4%) and less typically located in 5 (10%). Electrocardiographic abnormalities were present in 35 patients (70%). Typical findings were ST-T changes 22%(n = 11), LV hypertrophy 10%(n = 5) and total AV block 6%(n = 3). The more rare findings included left axis deviation 8%(n = 4), first-degree AV block 6%(n = 3), low voltage 4%(n = 2), VES 4%(n = 2), nonspecific intraventricular conduction delay 2%(n = 1), VT/VF 2%(n = 1), biventricular hypertrophy (1), RBBB 2%(n = 1) and LBBB 2%(n = 1). Mean follow-up period was 13 months ± 220 days (range, 1-46 months). During follow-up, 10 patients developed cardiac insufficiency, 7 rhythm disoders and 3 intracardiac thrombosis. One of these underwent transvenous ICD implantation due to VF. No patients died; 3 are currently on transplant waiting lists.
Conclusions: LVNC is a primary myocardial disease that presents with varied clinical, electrocardiographic and echocardiographic findings in childhood. Concomitant LVNC must especially be considered in CHD accompanied by LV dysfunction. Early diagnosis can prevent possible life-threatening complications.

MP1-20
Genetic markers of early onset of arterial hypertension

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Introduction: Familial history of hypertension is known to increase the risk of hypertension in the offsprings. Numerous studies confirming genetic predisposition to arterial hypertension have been performed during the last years, however data on heritability of particular genes from parents to.
Objective: To investigate the heritability of gene polymorphisms associated with essential hypertension in children. We examined whether gene polymorphisms associated with hypertension identified in parents are associated with a high blood pressure in offsprings.
Methods: The study subjects consisted of 13 essential hypertensive children (aged 6 to 18 years old) and their hypertensive parents with onset of disease before 35 years old (n = 13) and 18 normotensive children. 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 subjects. Gene DNA was extracted from blood samples and amplified by polymerase chain reaction (PCR).
Results: The study showed an association of TT genotype of the aldosterone synthase (CYP11B2) C-344T polymorphism with

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hypertension in children and early onset of disease in adults ($p < 0,05$). T allele/TT genotype were identified in 11 adults and it were 100% inherited by their offsprings. AGT T704C gene polymorphism and AGTR 2 G1675A gene polymorphism were inherited by 80% offsprings.
Conclusion: Family history of arterial hypertension is the main risk marker of high blood pressure in children. An early and a regular screening of the children of hypertensive parents is necessary to prevent any future cardiovascular diseases.

MP1-21
Impact of Cardiac MRI on Decision-making and Outcomes in Patients with a Borderline Left Ventricle
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Introduction: It is oftern challening to determine whether a 'borderline' left heart is sufficient to support the systemic circulation. In the last decade, cardiac MRI has emerged as a method to quantify left ventricular (LV) size and function. The goal of this study was to assess the outcomes of patients with HLHS and 'borderline' left ventricles in the current era.
Methods: All neonates with HLHS who underwent cardiac MRI to assess LV volume between Jan 2003–Jan 2012 were identified. Patients with valvar atresias and AVSD were excluded. Medical charts and imaging data were reviewed.
Results: 42 patients were identified. One underwent single ventricle palliation (Norwood). 3/42 patients received palliative care (genetic abnormalities or neurologic injury). 27/42 patients underwent primary biventricular management ($n = 2$ aortic valve dilation, $n = 22$ CoA repair, $n = 2$ CoA repair + VSD closure, $n = 1$ CoA repair and PAB). 11/42 patients had a stage one Hybrid procedure (H1, PDA stent and pulmonary artery banding) as an initial intervention, with 7/11 having subsequent biventricular repair (arch reconstruction, PA debanding +/- VSD closure +/- ASD fenestration). Mean birthweight 3.3 kg (range 2.9-4.2 kg). Mean age at first intervention 10 days (range 2-35).
LVEDVi measured by MRI was 27.9 +/- 6.9 ml/m2 (17.4-44.8 ml/m2), which was significantly larger compared to echo (monoplane Simpson's) 12.9 +/- 3.6 ml/m2 (4.9-24.3 ml/m2) ($p < 0.05$, $r = 0.58$) and echo (5/6 area-length) 22.8 +/- 5.8 ml/m2 (7.7-36.5 ml/m2) ($p < 0.05$, $r = 0.6$).
9/27 (33%) patients required re-intervention ($n = 1$ Ross-Konno, $n = 1$ AoV dilation, $n = 1$ SubAS resection, $n = 5$ MV repair (3/5 with SubAS resection). 2/9 patients required 3 and 4 re-operations each. 3/7 patients who had a BiV repair after H1 required re-intervention ($n = 1$ RPA plasty, $n = 2$ MV repair). There were 2 deaths in patients with severe pulmonary hypertension and one patient underwent heart transplant. At last follow-up, one patient was awaiting MV surgery for mitral stenosis. The other patients were well without significant residual lesions.
Conclusions: Measurement of LV volume is underestimated when assessed by echo compared to MRI. Additional data from MRI volume measurements is useful in decision making for patients with HLHS and a borderline LV. The H1 procedure can be a useful bridge to biventricular repair in patients who are truly 'borderline'.

MP1-22
Palivizumab prophylaxis in the infants with congenital heart disease: A controlled study from Turkey
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Background and Aim: Lower respiratory tract infections (LRTI) multiply morbidity and mortality within patients with significant congenital heart disease (CHD). For respiratory syncytial virus (RSV), one of the most important pathogens, immunoprophylaxis with palivizumab has successfully been introduced. In this retrospective case controlled study, we aimed to investigate efficacy of palivizumab with comparing the patients who had palivizumab prophylaxis with the ones who did not have.
Material and Method: In this retrospective case controlled study demographic, clinical and household characteristics, respiratory tract infections, hospitalization, morbidity and mortality rates of patients who had RSV prophylaxis (Group 1, n:92) between 2010-2012 period, were compared with the patients who did not take prophylaxis (Group 2, n:95), 2009-2010 period in 2 RSV seasons.

Characteristics and hospitalization features of the patients who had LRTI admissions in RSV season		Group 1 (n = 92)	Group 2 (n = 95)	P	Relative Reduction (%)
The rate of LRTI in the viral season	1x	14, %15.2	42, %44.7	**<0.001**	66
	2x	1, %1.1	6, %6.4	**<0.001**	83
	3x	1, %1.1	-	-	
Hospitalization rate associated with LRTI in the RSV season	1x	9, %9.8	37, %39.4	**<0.001**	75
	2x	1, %1.1	2, %2.2	**0.999**	50
Complicated LRTI		3, %3.2	11, %11.6	**0.029**	72
ICU admission associated with LRTI		3, %3.2	10, %10.5	**0.046**	70
Exitus associated with LRTI		4, %4.3	8, %8.4	0.256	49
Days of Hospitalization due to LRTI		6.5 (5.0-18.0)	8.0 (5.0-13.0)	0.951	19
Days of ICU stay		4.0 (2.0-6.0)	8.0 (1.0-11.0)	0.727	50

Values are expressed as n (%) or median, LRTI: Lower respiratuary tract infection, ICU: Intensive care unit.

Results: LRTI rates of patients who had prophylaxis compared to ones who did not have (15.2% vs 44.7%; $p < 0.001$), LRTI related hospitalization (9.8% vs 39.4%; $p < 0.001$), complicated LRTI (3.2% vs 11.6%; p:0.029), ICU need for LRTI (3.2% vs 10.5%; p:0.046) was statistically significant in Group 1. Most common cause LRTI was found to be RSV. Mortality rate was low in Group 1 (4.3% vs 8.4%) but not statistically significant (p:0.254). Independent risk factors for hospital admission in Group 1 was detected as under the weight centile <10rd (5.8 times) and associated chromosomal anomaly (Down syndrome etc) (4 times); in group 2 was congestive heart failure (8.6 times) and number of siblings at home (<11 years of age, 3.4 times).
Conclusion: This report is the first infant palivizumab efficacy report of Turkey. It was shown that palivizumab prophylaxis decreased hospitalization of patients with CHD, related complications and ICU admission rate. But further prospective, multicentered randomized, pharmacoeconomic studies were required.

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MP1-23

Are general paediatricians able to accurately distinguish innocent from pathological murmurs in children?

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Introduction: Up to 80% of children have heart murmurs but less than 1% have confirmed organic heart disease (1). Correct identification of pathological murmurs is important to avoid inappropriate referrals to paediatric cardiology (along with unnecessary patient and parental anxiety) or missed disease. This study investigated how accurately general paediatricians can distinguish between innocent and pathological murmurs.
Method: Sixty-two general paediatricians were recruited from across the Yorkshire region of England. They were played eight anonymised digital heart recordings obtained from asymptomatic children with an otherwise normal cardiovascular examination. The murmurs included a ventricular septal defect, atrial septal defect, patent ductus arteriosus, Still's murmur and pulmonary flow murmur. Participants stated whether each recording was normal heart sounds, an innocent or pathological murmur as well as the likely diagnosis and necessity for referral. Demographic data including age, gender, level of experience and any speciality training in cardiology was also collected.
Results: Of those recruited, 35 were consultants and 27 trainees. The sensitivity of distinguishing innocent from pathological murmurs was 89.9%, specificity 66.5% and diagnostic accuracy 78.3%. The false positive rate for referrals to paediatric cardiology was 43.2% with a false negative rate of 8.7%. On sub-group analysis the only variable of statistical significance was level of experience. The difference between specificity for consultants (74.3%) and trainees (51.2%) was 23.1% (95%CI = 10.1 to 36.1%).
Conclusion: These results raise concern about the ability of general paediatricians to accurately distinguish innocent from pathological murmurs. Although inappropriate cardiology referrals place extra pressure on already stretched services, potential complications from missed disease are clearly far more worrying. Given that there is clearly neither the time or resources available for paediatric cardiologists to assess every child with a murmur, this research provides further evidence of the need for increased numbers of paediatricians with cardiology expertise in the UK.
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MP2-1

Knowledge-based 3D reconstruction compared to MRI for evaluation of right ventricular volumes and function in congenital heart diseases affecting the right ventricle

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Background: Right ventricular (RV) volume and ejection fraction (RVEF) measurements are essential in the management of children with congenital heart disease. Cardiac Magnetic resonance imaging (MRI) is considered the reference method for RV volumes and RVEF measurements. Three-dimensional knowledge-based reconstruction (3D-KR) derived from two-dimensional echocardiographic imaging is a novel technique. It has proved to provide accurate and reproducible measurements of RV volumes in patients with Tetralogy of Fallot or pulmonary arterial hypertension. The aim of this study was to assess the feasibility and reliability of this novel echocardiographic technique in children (age 3 months to 18 years) with diverse CHD involving the right ventricle.
Methods: 75 children (mean age 9.2 +/- 2.3 years) referred for cardiac MRI, were included. Among them, 25 patients had barometric overload, 32 patients had volumetric overload, and 18 patients had mixed overload. Echocardiographic image acquisition was performed using a standard ultrasound scanner linked to a Ventripoint Medical Systems unit. Parameters analyzed were end-diastolic volume (EDV), end-systolic volume (ESV), and RVEF. The method of disks was used for CMR RV volumes. Intra-observer, inter-observer, and inter-technique variability was assessed using Pearson's correlation coefficient (CC), coefficients of variation (COV), and Bland-Altman analysis.
Results: Feasibility of 3D-KR was 100%. Echocardiographic RV volumes correlated well with CMR (EDV, CC = 0.96; ESV, ICC = 0.93; RVEF, ICC = 0.75). For inter-observer analyses, COV were 8% for EDV, 15% for ESV, and 17% for EF. For intra-observer analyses, COV were 4% for EDV, 7% for ESV, and 9% for EF. The correlation of volumes and RVEF with MRI was slightly worse in the group with mixed overload compared with patients with volumetric or barometric overload. 3D-RVEF was overestimated compared with MRI whereas the volumes tend to be underestimated.
Conclusions: 3D-KR is feasible in children. It provides accurate and reproducible measurements of RV volumes. RVEF is less accurate and reproducible than volumes when compared with MRI but values are still comparable with both techniques. This new technique can be used as an accurate routine tool to assess RV function in CHD with pure barometric or volumetric overload.

MP2-2

Impaired Cardiac Mechanics in a Large Population of Children with Heart Transplantation: A Speckle Tracking and Three-Dimensional Echocardiography Study

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Background: Cardiac dysfunction is a life threatening condition in heart transplanted kids. The purpose of our study was to evaluate advanced echocardiographic indices of cardiac function in a sample of pediatric heart transplant.
Methods: 60 pediatric patients with stable cardiac transplantation and 60 matched healthy controls were included in the study. All individuals underwent transthoracic echocardiographic examination including tissue Doppler, 2D-speckle tracking and three-dimensional echocardiography. 2D-Speckle tracking analysis was used to obtain measures of left ventricular radial, circumferential and longitudinal strain, and to derive left ventricular twist and torsion. Three-dimensional echocardiography was used to measure left ventricular volumes, ejection fraction and to evaluate left ventricular systolic synchrony.
Results: No differences were observed between the two groups in left ventricular volumes, left ventricular ejection fraction, or right ventricular fractional area change. However, transplanted patients showed lower values of longitudinal systolic excursion of valvular planes at both the mitral and the tricuspid valve level, as well as higher mitral E/E ' ratio (all $p < 0.05$). Cardiac radial strain was similar between groups, while a significant net reduction in both global left and right ventricular longitudinal strain as well as in left ventricular global circumferential strain could be observed between the two groups (all $p < 0.05$). In addition reduced left

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ventricular twist and torsion was found in patients with cardiac transplantation as compared to normal subjects ($p < 0.01$) mainly due to a significant reduction in basal rotation ($p < 0.05$). Eventually, in 20% of our cardiac transplant patients—and none of the controls—overt systolic dyssynchrony was observed.
Conclusion: even in the absence of acute rejection and in the presence of a normal ejection fraction, children with transplanted heart show a significant reduction in subclinical markers of biventricular function. Additional prognostic studies are needed to establish whether these abnormalities predict the incidence of future development of clinically evident cardiac dysfunction.

MP2-3

Global longitudinal diastolic and systolic right ventricular function decreases during medium-term follow-up of children with Hypoplastic Left Heart Syndrome

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Basics: Serial assessment of right ventricular (RV) function in children with Hypoplastic Left Heart Syndrome (HLHS) is important after 3 stage palliation with total cavopulmonary connection (TCPC). For this purpose, echocardiographic two-dimensional Speckle Tracking (2DST) is a promising technique as limitations related to geometry or angle of insonation are lacking. Therefore, it was used to evaluate RV global longitudinal peak systolic strain (GS) and peak strain rate in systole (GSRs), early (GSRe) and late (GSRa) diastole in HLHS patients. During medium-term follow up global strain rate has been shown to be valid surrogate parameter of myocardial elastance.
Methods: RV-GS and -GSRs, -GSRe, and -GSRa of 42 HLHS patients (median age at TCPC 31.5 (28.1 to 35.2) months) were obtained at two times: early after TCPC at a median interval of 18.8 (15.3 to 25.6) months after TCPC, and at a median interval of 61.2 (50.4 to 83.5) months after TCPC. GS and GSR values were compared using the non-parametric Wilcoxon-test.
Results: GS did not change between both examinations (median -18.5 (-16.2 to -22.3)% vs. -17.9 (-15.9 to -20.5)%, $p = 0.49$), however GSR in systole and diastole decreased significantly (GSRs: median -1.5 (-1.4 to -1.7)1/s vs. -1.35 (-1.13 to -1.5)1/s, $p = 0.003$; GSRe: median 2.1 (1.9 to 2.6)1/s vs. 1.9 (1.7 to 2.28)1/s, $p = 0.033$; GSRa: median 1.1 (0.9 to 1.4)1/s vs. 0.9 (0.61 to 1.1)1/s, $p = 0.01$).
Conclusions: In HLHS patients global systolic and diastolic strain rate decrease within a median period of 42.4 months. These findings indicate a decline of systemic right ventricular function in children with HLHS. The fact that in a previous study strain rate has been identified as a valid surrogate parameter of myocardial elastance, underlines the importance of our findings. Therefore, regular 2DST examinations should be part of echocardiographic follow up in all patients with HLHS after surgical palliation.

MP2-4

Aortopulmonary collateral flow in patients with cavopulmonary bypass is underestimated by fluoroscopic angiography compared to magnetic resonance flow measurements

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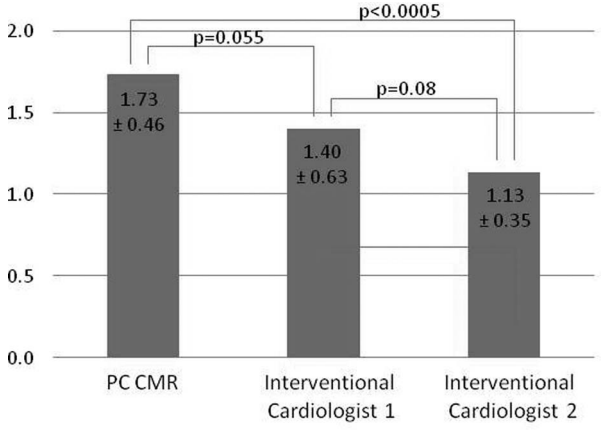
Introduction: Aortopulmonary collaterals (APCs) in patients with functionally single ventricle after creation of a bidirectional cavopulmonary connection (BCPC) are a predictor of adverse outcome after the Fontan completion. While phase contrast flow velocity mapping (PC) by cardiac magnetic resonance (CMR) is now the gold standard for a quantitative assessment of collateral flow, fluoroscopic angiography is the most frequently used tool in clinical practice. The aim of this study was to compare the accuracy of collateral flow assessment using fluoroscopic angiography to CMR.
Methods: Fluoroscopic angiographies in 15 patients (age 28 ± 8 months) with BCPC were reviewed and compared to APC flow by CMR, obtained under the same anaesthesia. The studies were reviewed by two experienced interventional cardiologists blinded towards subsequent coil occlusion of collaterals and CMR results. APC flow was graded as mild, moderate or severe on the basis of arterial injections into the ascending aorta and its branches (direct visualization of APCs) and venous injections into the superior vena cava (contrast wash-out from unopacified APC flow). APC flow was quantified by PC CMR as the difference of pulmonary venous flow (Qpv) and pulmonary arterial flow, expressed as the percentage of Qpv and classified into mild (<20%), moderate (20-40%) and severe (>40%) collateral flow.
Results: Grading of the APC flow differed significantly between the CMR and fluoroscopic method ($p < 0.0005$ and $p = 0.055$ for interventional cardiologist 1 and 2, respectively, Kappa agreement 40% for both). The agreement between the two catheter angiography readers ($p = 0.08$, Kappa overall agreement 79%) was acceptable. There was no correlation between grading by fluoroscopic angiography and PC CMR ($r = 0.266$), with an overall lower grading of APC flow by fluoroscopic angiography (see Figure).
Conclusions: In patients with BCPC, fluoroscopic angiography, although reasonably reproducible, underestimates APC flow systematically as compared to PC CMR. The inaccuracy of fluoroscopic angiography is thought to be due to diffuse collateral flow which is not routinely visualized by fluoroscopic angiography, but captured by PC CMR. PC CMR can guide the cardiac catheterization and the decision to occlude APCs prior to the completion of the Fontan circulation.



Grading of Collateral Flow, mean values

MP2-5

Clinical significance of left and right ventricle echocardiographic parameters in children with idiopathic pulmonary arterial hypertension

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Background: Echocardiography is commonly used for assessment and serial follow-up of right (RV) and left ventricle (LV) function in children with idiopathic pulmonary arterial hypertension (iPAH). Clinical significance and predictive value of echo parameters have been scarcely characterized.
Objectives: to characterize RV function in children with iPAH in stable and worsening clinical status, and to assess value of echocardiographic indices to predict clinical worsening.
Methods and results: Clinical, biological and echocardiographic variables were prospectively collected in 38 children with iPAH. Patient's median age at inclusion was 6.3 years old, 95%IC [3.2-11.1]. Median follow-up was 15.4 months. Forty seven echo scans were performed in children at time of clinical worsening (TCW) defined as NYHA $\geqslant$ III and/or recent syncope and/or overt RV failure, and 222 echo scans were performed in children in stable clinical status (SC) defined by NYHA $\leqslant$ II, without syncope and without RV failure. Median delay between each visit was 54 days, 95%CI [45-71].
Patients at TCW were significantly younger (median age 5.5 years old 95% CI [2.8-8.6], $p = 0.001$), and had lower body mass index ($p = 0.002$). TAPSE, RV peak systolic myocardial velocity, pulmonary acceleration time, aortic and pulmonary velocity-time integral (VTI) were significantly reduced at TCW ($p \leqslant 0.001$). LV filling was also impaired at TCW: mitral early (E)/late(A) ratio and LV myocardial early diastolic velocities (mitral E') were significantly reduced (respectively $p = 0.03$ and $p = 0.01$). In univariate analysis, RV fraction area change and mitral E' were predictive of SC. In multivariate analysis, aortic VTI was predictive of TCW even after adding age and B-type natriuretic peptide into the model (HR = 0,7349, 95%CI [0,5616-0,9617], $p = 0,02$). ROC curve analysis confirmed the relationship between aortic VTI and TCW (area under curve 0,810, 95%CI [0,722-0,880]). An aortic VTI $\leqslant$ 16 cm (sensitivity 71%, specificity 83%) predicted TCW.
Conclusion: Echocardiographic parameters of RV function and LV filling were impaired in children with iPAH at TCW. Aortic VTI predicted TCW. Inter-ventricular interactions in this setting might provide further insights into the mechanisms leading to clinical worsening.

MP2-6
Surrogate echocardiographic parameters to assess right ventricular function in children with congenital heart disease or pulmonary hypertension

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Background: In congenital heart diseases (CHD) and pulmonary hypertension (PH), right ventricle (RV) dysfunction is associated with prognosis. Because of RV pyramidal shape, RV ejection fraction measured by standard 2D echocardiography is not accurate. Surrogate parameters of RV systolic function are used in routine. However, their dependence to RV loading conditions has not been tested. Moreover, studies have suggested that these parameters are altered by cardiac surgery.
The aim of our study was to assess the feasibility, reproducibility and relevance of surrogate echocardiographic parameters of RV systolic function in children with CHD involving the RV or pulmonary hypertension (PH).
Methods: We recruited 294 consecutive children with RV pathological loading conditions: 159 have barometric overload, 102 have volumetric overload, 33 have mixed overload. Among them 154 patients have history of cardiac surgery. The median age was 9.25 +/- 2.32.
We assessed RV global systolic function by measuring the Fractional area change (FAC). 31 patients had also assessment of RVEF by MRI and 168 had assessment of RVEF by the Ventripoint system (VMS).
We analyzed the feasibility, reproducibility and relevance of parameters of RV longitudinal systolic function: the Tricuspid Annular Peak Systole Excursion (TAPSE), RV 2 dimensional longitudinal strain and Tissue Doppler derived parameters: tricuspid systolic excursion velocity (Sa), myocardial acceleration during isovolumic contraction (IVA), and RV Tei index.
Results: TAPSE and Sa are the surrogate parameters with the best feasibility (>98%) and reproducibility (inter and intra observer variability <6%). 2D strain, IVA and Tei index have worse feasibility (78, 83 and 91% respectively) and reproducibility (inter observer variability of 10, 21 and 19%). TAPSE a nd Sa were correlated to RV loading conditions. In patients with history of cardiac surgery, no parameter of longitudinal function was correlated to FAC or RVEF. Only in PH, TAPSE and Sa were correlated to RV global systolic function assessed by FAC (respectively r 0.84 and r 0.54, $p < 0.001$).
Conclusion: Parameters of longitudinal RV systolic function correlate with loading conditions. They cannot be used to predict RVEF in children with CHD or in children who had undergone cardiac surgery. They can only be used to predict RVEF in PH.

MP2-7
Right Ventricular Myocardial Deformation in Adult Patients with a Repaired Atrial Septal Defect

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Objectives: Patients with atrial septal defect (ASD) have right ventricular (RV) volume overload caused by left to right shunting or anomalous venous return. Using speckle-tracking echocardiography, we evaluated regional RV and left ventricular (LV) deformation in adult patients with repaired ASD in childhood in comparison with healthy controls.
Methods: Echocardiogram including the standard apical views was acquired in adult patients with repaired ASD and in healthy controls. With speckle-tracking echocardiography, we analyzed longitudinal strain of the RV lateral wall, LV septum, and LV lateral wall. Cardiac magnetic resonance imaging (CMR) was performed in 47 (92%) ASD patients.
Results: We included 104 subjects: 51 with repaired ASD (39% male, age 43.3 $\pm$ 4.9 years, age at repair 7.9 $\pm$ 3.6 years) and 53 healthy controls (49% male, age 29.7 $\pm$ 6.8 years). RV global longitudinal strain (GLS) of the lateral wall was significantly lower in ASD patients than in controls, especially of the apical segment (Fig. 1). RV GLS correlated significantly with CMR derived RV and LV end-diastolic volume ($r = 0.49$, $p = 0.014$ and $r = 0.43$, $p = 0.034$), and with RV and LV end-systolic volume ($r = 0.53$, $p = 0.005$ and $r = 0.46$, $p = 0.019$). LV GLS was similar between the ASD patients and controls.
Conclusions: Although the ASD repair was performed in childhood, RV longitudinal strain is still significantly reduced in adult patients long after surgery. Especially the strain of the apical

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segment is reduced suggesting that apical function is more affected in these RV overloaded ventricles.

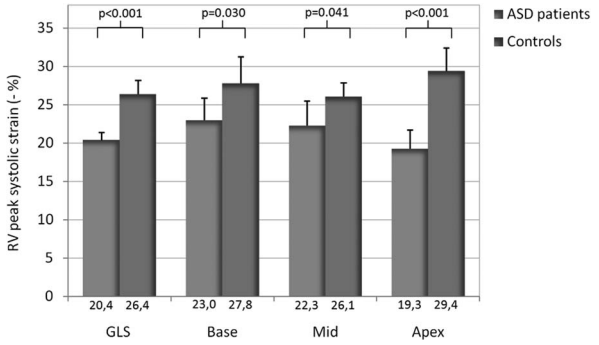

Fig. 1. Right ventricular global and segmental longitudinal strain of the lateral wall.

MP2-8

Speckle tracking strain echocardiography unmasks left ventricular systolic dysfunction in patients after neonatal coarctation repair

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Introduction: Patients with neonatal, critical coarctation of the aorta often suffer from ventricular dysfunction pre-operatively. This contrasts to patients in whom coarctation of the aorta becomes evident at an older age who usually have left ventricular hypertrophy with preserved systolic function. If these differences remain present during long-term follow-up is as yet unclear.
Methods: We compared the echocardiographic results of patients who underwent coarctectomy within the first 30 days of life (group A; N = 132) and those who underwent coarctectomy at an older age (Group B; N = 137) after a follow-up period of 7.5 ± 6.5 years. A commercially available echosystem was used (Vivid-7.0 General Electric Vingmed Ultrasound). Mitral inflow parameters (E, A and decelaration time) were measured in 4-chamber view, as well as measurement of the mitral valve annulus. Long axis view was used to measure LV end systolic and end diastolic dimensions as well as left ventricular thickness of the septum and posterior wall. Shortening fraction (SF) was defined as ((LVED-LVES)/LVED)*100%. Global longitudinal strain was measured from an apical 4-chamber view as the average of all six predefined LV segments.
Results: Age did not differ between the two groups (9 ± 7 versus 9 ± 7 years). There was no difference in mitral valve annulus (21 ± 6 versus 21 ± 5 mm) nor in residual flow velocity at the previous coarctation site (2.2 ± 0.6 versus 2.2 ± 0.5 m/sec). No difference was found in LV septal thickness or in posterior wall thickness. LV diastolic function did not differ between the two groups (MVE 1.2 ± 0.2 versus 1.3 ± 0.2 m/sec; MVA 0.7 ± 0.3 versus 0.7 ± 0.3 m/sec as did systolic function conventionally measured (LVED 41 ± 9 versus 42 ± 10 mm; LVES 25 ± 7 versus 25 ± 7 mm; SF 40 ± 7 versus 41 ± 7%). However, there was a small, but significant difference in global longitudinal strain (18 ± 3% versus 19 ± 4%; $p < 0.05$). Both groups differed significantly in strain from a group of 117 normal controls (global longitudinal strain 22 ± 2%; $p < 0.001$).
Conclusions: LV systolic performance as assessed by speckle tracking strain echocardiography shows a lower global longitudinal strain in patients after coarctectomy which is most prominent in those who undergo neonatal coarctation repair. This emphasizes careful follow-up of LV systolic performance, especially in neonatal coarctation patients.

MP2-9

Echocardiographic Nomograms for Chamber Diameters, Area and Volumes in Children: Preliminary Data

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Background: Despite a quantitative evaluation of cardiac chambers dimension in pediatric echocardiography is often required, nomograms for these structures are very limited or even absent.
Aim: to establish reliable pediatric echocardiographic nomograms for cardiac chamber dimensions.
Methods: we performed two-dimensional and M-mode measurements of 23 individual cardiac structures. These included: systolic and diastolic left ventricular (LV) volumes, area and length calculated in 4 and 2-chamber view by Simpson's method, LV diameters evaluated in M-mode and in 4 and 2-chamber views, right ventricular area and diameters and atrial diameters and area evaluated in 4-chamber view.
Statistical analysis: Models using linear, logarithmic, exponential, and square root relationships were tested. Heteroscedasticity was tested by White test and Breusch-Pagan test. Age, weight, height, and body surface area (BSA), calculated by seven different formulas, (i.e. Haycock, DuBois, Mosteller, Dreyer, Meban, Boyd and Gehan) were used as the independent variables in different analysis to predict the mean values of each echocardiographic measurement. Structured Z scores were then computed.
Results: 912 consecutive Caucasian Italian healthy children (age range 0 days-17 years; 44.8% females) with a BSA ranging from 0.12 to 1.8 m2 were prospectively enrolled.

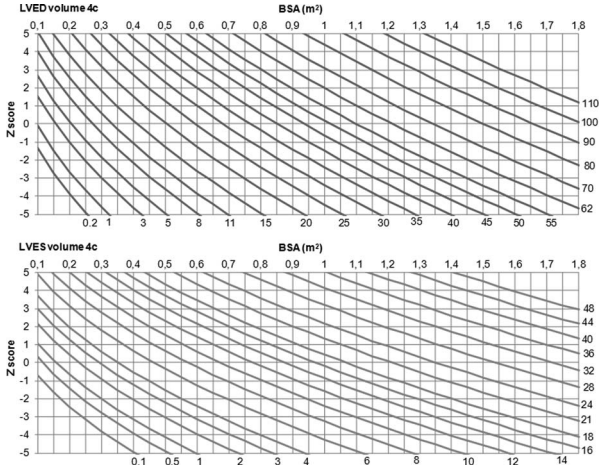

Figure 1: we report, as an example, percentile-charts for LV volumes (by Simpson's method).
LVED = left ventricular end diastolic, LVES = left.

The calculation of BSA using the Haycock formula provided the best results while other formulas either underestimated (DuBois,

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Mosteller, Dreyer, and Meban) or overestimated (Boyd and Gehan) BSA.
The Haycock formula has been used when presenting data. Data have been presented by the use of tables showing predicted values (mean ± 2SD) for a given BSA and by percentile charts. Equations relating echocardiographic measurements to BSA for every parameter were also provided. For all the measurements there was no significant intra/inter-observer variability.
Conclusions: We present echocardiographic normal values for chamber volumes, area and diameters derived from a large population of children. Our data partly cover an important gap of knowledge in actual pediatric echocardiographic nomograms and will allow for a more reliable calculation of chamber dimensions. Further studies are required to reinforce these data, as well to evaluate other parameters and ethnicity.

MP2-10
Long axis dysfunction and stiff arteries in children with severe acute malnutrition in Kenya
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Background: Annually, 2.1 million children die world wide as a consequence of severe acute malnutrition (SAM). There is conflicting evidence on cardiac function in SAM, which has influenced treatment guidelines. We wished to establish whether children with SAM have impaired ventricular function.
Methods: In the CArdiac Physiology in MALnutrition (CAPMAL) Study we recruited children with SAM (marasmus or kwashiorkor), and age and gender matched, equally sick non-malnourished controls who presented to a rural Kenyan hospital between March and November 2011. Echocardiograms, lactate and N-Terminal-proBNP (NT-proBNP) were recorded at days 0, 7 and 28.
Results: 88 SAM cases and 22 controls had similar baseline characteristics apart from anthropometry (weight for height z-score -3.2 for cases vs -1.1 for controls) and HIV status (22.7% vs 0.0%) ($p < 0.05$).
On admission, SAM children had reduced long axis function compared with controls in the right (TAPSE 13.9 vs 17.0 mm; $p < 0.0001$) and left ventricle (MAPSE 8.4 vs 10.5 mm; $p < 0.0001$), reduced systolic myocardial velocity (5.6 vs 7.9 cm/s, $p < 0.0001$) and early diastolic velocity (10.7 vs 13.7 cm/s, $p = 0003$). Systemic vascular resistance index (SVRI) was increased in SAM at all reviews (day 0: 1862 vs 1015, day 7: 1538 vs 966 and day 28: 1476 vs 973, all $p < 0.005$). Both TAPSE and MAPSE were negatively correlated with SVRI (Spearman's rho = -0.5863 and -0.5803 respectively, $p < 0.0001$). We found no difference on admission in fractional shortening (36.4 vs 37.8%), Tei Index (0.39 vs 0.34), E/A ratio (1.24 vs 1.16), cardiac index (CI) or left ventricular mass (LVm) (all $p > 0.05$). Similar differences were seen when the analysis was confined to HIV-negative children, apart from CI and LVm which were significantly lower in cases. No difference in lactate and NT-proBNP was observed at any time point.
Conclusions: This is the first detailed echocardiographic study of children with SAM, and the first to report a reduction in long axis function in SAM. This is associated with increased systemic vascular resistance and might help to explain why children fare badly on attempted re-feeding. It might also help explain why systemic hypertension is seen in adults with a history of childhood malnutrition.

MP2-11
Right Ventricular Systolic Function in HLHS: A Comparison of manual and automated software to measure fractional area change
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Introduction: Quantitative echocardiographic assessment of right ventricular function is important in children with hypoplastic left heart syndrome (HLHS). Previous work from our group has demonstrated that fractional area change (FAC) measured on echocardiography had the closest correlation with magnetic resonance imaging (MRI) derived ejection fraction (EF) when compared to other echocardiographic measures such as myocardial velocity or longitudinal strain. The aim of this study was to examine repeatability of different echocardiographic techniques, both manual and automated, to measure fractional area change (FAC) in HLHS patients and to compare these measurements to MRI.

Table. Inter and intraobserver correlation and comparison to MRI EF for FAC from VVI, QLAB and manual tracing.

Technique	Intraclass correlation coefficient Intra observer/Interobserver	Correlation to EF MRI
Automated technique		
VVI	0.997 (0.989-0.999)/0.976 (0.902-0.994)	0.7 ($p<0.0001$)
QLAB	0.976 (0.903-0.994)/0.891 (0.561-0.973)	0.6 ($p = 0.001$)
Manual technique	0.774 (0.427-0.922)/-0.205 (-0.498-0.269)	0.4 ($p = 0.003$)

Methods: Children with HLHS underwent transthoracic echocardiogram and cardiac MRI under the same general anesthetic as part of routine interstage assessment. FAC was measured from apical 4-chamber view using three different techniques: Velocity-vector-imaging (VVI) (Syngo USWP 3.0, Siemens Medical Solutions), 2D automated analyzing program (QLAB R 10.0, Philips) and manual endocardial contour tracing (Xcelera, Philips). Intra and interobserver variability was calculated using intraclass correlation coefficient. MRI EF was calculated in the standard way and compared to echocardiography derived FAC.
Results: Forty-nine patients had studies available for analysis. Intraobserver variability was good for all methods (Table). Interuser variability was good for all parameters except the manually computed FAC where it was poor, particularly with regards to the end systolic area. All techniques used to calculate FAC correlated with MRI derived EF (Table). The highest correlation was found with the automated techniques.
Conclusions: Automation improves reliability in measuring right ventriciular systolic function in HLHS and correlates better with MRI. There are some differences between automated softwares in terms of correlation with MRI derived EF. In clinical practice, a single automated technique appears optimal.

MP2-12
Effect of inhaled nitric oxide on blood flow dynamics in patients after the Fontan procedure using standard CMR flow measurements
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Background: Invasive hemodynamic studies have shown that nitric oxide (NO), a selective pulmonary vasodilator, can lower pulmonary vascular resistance (PVR) in Fontan patients although calculation of blood flow can be inaccurate in these patients. The aim of the study was to detect changes in blood flow within the Fontan circulation after inhalation of NO using cardiovascular magnetic resonance (CMR).
Methods: 29 patients (mean age 12.7 ± 6.7 years) after the Fontan procedure underwent CMR as part of their routine clinical assessment. Standard 2-dimensional blood flow measurements were performed in the inferior vena cava (IVC), superior vena cava (SVC) and ascending aorta (Ao) before and after inhalation of 40 ppm NO for 8-10 minutes. Aortopulmonary collateral (APC) flow was calculated as Ao − (SVC+IVC).
Results: Heart rate (83 ± 19 to 82 ± 18 bpm; p = 0.90) and transcutaneous oxygen saturations (93 ± 5 to 94 ± 4%; p = 0.65) did not change under NO inhalation. NO inhalation did not affect flow in the Ao (3.23 ± 0.72 to 3.07 ± 0.81 l/min/m2; p = 0.43), IVC (1.56 ± 0.41 to 1.64 ± 0.47 l/min/m2; p = 0.50) and SVC (1.03 ± 0.41 to 1.04 ± 0.42 l/min/m2; p = 0.88) resulting in unchanged total caval flow (2.59 ± 0.62 to 2.69 ± 0.72 l/min/m2; p = 0.50). APC flow decreased significantly from 0.64 ± 0.48 to 0.39 ± 0.45 l/min/m2 (p = 0.04).
Conclusions: Inhalation of NO does not improve pulmonary blood flow in Fontan patients while APC flow decreased significantly suggesting a beneficial effect of pulmonary vasodilators in unloading the single ventricle.

MP2-13
Coronary artery imaging in patients with Congenital Heart Disease: Improved image quality by the use of an intravascular contrast agent and specific MR sequence design

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Aim: In patients with congenital heart disease (CHD) imaging of coronary artery origin and course can be crucial for preoperative planning. In this study a novel intravascular contrast agent (gadofosveset trisodium-GdT) was injected and a standard commercially available T2-prepared steady-state-free-precession (SSFP) MR sequence and an inversion-recovery (IR) SSFP magnetic resonance (MR) sequence was used. Results were compared with a frequently used extravascular contrast agent (Gadopentetate dimeglumine-GdD) and a standard commercially available T2-prepared SSFP MR sequence.
Material and Methods: Ten patients with CHD (age range 22 to 40 years; mean 31 years) were scanned at a 1.5 T clinical MR scanner (Achieva, Philips Healthcare, Best, Netherlands) using a 32-element cardiac coil. An extravascular contrast agent was used first using a standard commercially available T2-prepared SSFP sequence. Within 72 hours patients were re-scanned using a novel intravascular contrast agent and IR SSFP additional to the standard T2-preapred SSFP MR sequence. Contrast-to-noise ratios (CNR) and image quality achieved by using the intravascular agent with an optimized scan protocol were compared to those achieved by using standard extravascular agent with a standard sequence.
Image quality was graded form 0 (non-diagnostic) to 4 (best image quality) for the left anterior descending (LAD), the left circumflex (LCx) and the right coronary artery system (RCA) using the extracelluar (T2-prepared SSFP) and intracellular (T2-prepared SSFP and IR-SSFP) contrast agent. Data were analyzed using Soap Bubble Tool and Osirix.
Results: Mean image quality was calculated for each vessel and sequence design, as well as mean CNR for each of the sequence. GdT showed to be superior in imaging coronaries, not only in image quality based on grading, but also better CNR when combined with an IR SSFP sequence. (See Fig. 1a to 1d)

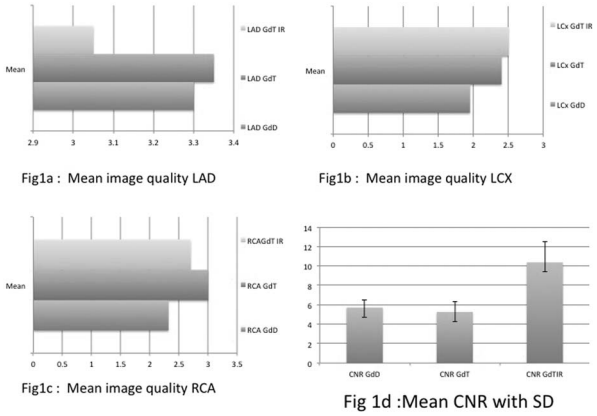

Fig1a : Mean image quality LAD

Fig1b : Mean image quality LCX

Fig1c : Mean image quality RCA

Fig 1d :Mean CNR with SD

For LAD and RCA GdT T2-prepared SSFP showed better diagnostic accuracy, whereas using GdT associated to an IR sequence LCx and CNR showed to be superior.
Conclusion: The combination of a novel intravascular contrast agent (GdT) and the addition of an adapted sequence design (IR SSFP) to the routine protocol provide superior imaging results compared to currently available contrast agents and sequence design.

MP2-14
The Effect of Albumin:Creatinine Ratio on Standard Echocardiographic Parameters in Adolescent Type 1 Diabetes.

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Introduction: Adolescents with type 1 diabetes (T1D) are at increased risk of early adult-onset cardiovascular disease. This study compared standard echocardiographic parameters in patients screened for the Adolescent Type 1 Diabetes Cardio-Renal Intervention Trial (AdDIT) with healthy controls.
Methods: Standard M-mode, B-mode and Doppler echocardiography was performed in all subjects. In subgroup analysis, T1D separated into tertiles according to urinary albumin:creatinine ratio as per the AdDIT protocol, were compared with the subgroup of healthy controls that underwent the same baseline clinical assessment including glycaemic measures and serum lipids. Between groups comparisons were performed using Student's t-tests, with p-values <0.05 considered significant.
Results: 188 T1D patients (M:F 93:95; age 14.4 ± 2.1 years; disease duration 7.0 [1.7-15.0] years) and 178 controls (M:F 84:94; age 14.5 ± 1.6 years) were studied. Sex, age and height were similar, but T1D were heavier. T1D had increased systolic (114 ± 10 vs. 110 ± 9 mmHg; p = 0.0001) and diastolic blood pressures (62 ± 7 vs. 58 ± 7 mmHg; <0.0001), but decreased resting heart rates (65 ± 9 vs. 68 ± 12 beats per minute; p = 0.0312). Left ventricular (LV) dimensions along with indexed LV mass were all similar, except for posterior wall

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thickness which was increased in T1D (0.66 ± 0.11 vs. 0.64 ± 0.11 cm; p = 0.0258), as has been previously reported. Systolic function indices, ejection fraction (68 ± 5 vs. 66 ± 5%; p = 0.0020) and mean velocity of circumferential shortening (1.18 ± 0.18 vs. 1.14 ± 0.16 circ/sec; p = 0.0161), were increased in T1D. Diastolic function indices, mitral valve E, A and E/A were all similar, but deceleration (154 ± 17 vs. 149 ± 20 msec; p = 0.0235) and isovolumetric relaxation times (74 ± 17 vs. 76 ± 8 msec; p = 0.0070) were increased. In subgroup analysis, comparing 53 high-risk, 71 medium-risk and 64 low-risk T1D vs. 59 controls, as expected all T1D had increased fasting blood glucose and HbA1c, and also HDL cholesterol. Total and LDL cholesterol and triglycerides were similar. Only LV posterior wall thickness remained increased when comparing the T1D subgroups vs. controls (0.67 ± 0.11 high-risk, 0.66 ± 0.11 medium-risk, 0.67 ± 0.13 low-risk vs. 0.62 ± 0.10 cm controls; p = 0.0172, p = 0.0327, p = 0.0182, respectively).
Conclusions: Adolescent T1D of short to intermediate disease duration, have early suggestion of blood pressure, diastolic dysfunction and left ventricular geometric changes, which may contribute to increased risk of early adult-onset cardiovascular disease.

MP2-15

Exploring Myocardial Function in Adolescent Type 1 Diabetes

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The Hospital for Sick Children, Toronto, Canada (1); University Health Network, Toronto, Canada (2); Addenbrooke's Hospital, Cambridge, United Kingdom (3); Department of Family and Community Medicine, University of Toronto, Toronto, Canada (4)

Introduction: Preclinical detection of myocardial dysfunction in adolescents with type 1 diabetes (T1D) may help identify individuals at increased risk of adult-onset cardiovascular disease, who would most benefit from early intervention strategies. This study compared echocardiographic assessment of myocardial function in patients screened for the Adolescent Type 1 Diabetes Cardio-Renal Intervention Trial (AdDIT) with healthy controls.
Methods: Myocardial function assessment included tissue Doppler, strain and strain rate imaging. In subgroup analysis, T1D separated into tertiles according to urinary albumin:creatinine ratio as per the AdDIT protocol, were compared with the subgroup of healthy controls that underwent the same baseline clinical assessment including glycemic measures and serum lipids. Between groups comparisons were performed using Student's t-tests, with p-values <0.05 considered significant.
Results: 188 T1D patients (M:F 93:95; age 14.4 ± 2.1 years; disease duration 7.0 [1.7-15.0] years) and 178 controls (M:F 84:94; age 14.5 ± 1.6 years) were studied. Sex, age and height were similar. T1D were heavier with increased systolic and diastolic blood pressures, but decreased resting heart rates. Lateral mitral valve annulus myocardial velocities, E' (17.5 ± 2.6 vs. 18.6 ± 2.6 cm/s; p = 0.0001), A' (5.4 ± 1.1 vs. 5.9 ± 1.1 cm/s; $p<0.0001$) and S (10.5 ± 1.8 vs. 11.1 ± 2.0 cm/s; p = 0.0017) were decreased and mitral valve E/E' (5.8 ± 1.1 vs. 5.4 ± 1.0; p = 0.0002) increased. Global left ventricular (LV) circumferential strain ((-20.4 ± 2.3 vs. -19.5 ± 1.7%; p = 0.0002) and longitudinal strain (-19.6 ± 1.7 vs. -18.9 ± 1.9%; p = 0.0003) were different, but systolic and diastolic global longitudinal strain rates were similar. Basal rotation was similar, but apical rotation (6.26 ± 2.97 vs. 5.28 ± 2.45 degrees; p = 0.0012) and LV twist (10.35 ± 3.58 vs. 9.33 ± 3.05 degrees; p = 0.0065) were increased. In sub-group analysis, comparing 53 high-risk, 71 medium-risk and 64 low-risk T1D vs. 59 controls, all T1D had increased fasting blood glucose and HbA1c as expected and also HDL cholesterol, but total and LDL cholesterol and triglycerides were similar. Only global LV circumferential strain in the medium-risk tertile remained different when comparing the smaller T1D subgroups vs. controls (-19.4 ± 1.6 vs. -18.8 ± 1.7%; p = 0.0312).
Conclusions: Significant changes in myocardial function are evident in adolescent T1D of short to intermediate disease duration, suggesting these may be clinically useful preclinical markers of deterioration in cardiac performance to guide early intervention.

MP2-16

Three-dimensional echocardiography in Ebstein's anomaly of the tricuspid valve; a comparison with MRI

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Introduction: Tricuspid valve is the most complex valve with variable geometry. Ebstein's anomaly further adds to this complexity. Small studies and case reports showed incremental benefits of 3D echocardiography over 2D echocardiography in the evaluation of Ebstein's anomaly. We aimed to study the utility of 3D trans-thoracic echocardiography in the evaluation of Ebstein's anomaly and correlate with findings on cardiac magnetic resonance (CMR) imaging.
Methods: We included 24 patients (with age range of 8–45 years) with a diagnosis of Ebstein's anomaly. 2D echocardiography followed by trans-thoracic 3D echocardiography and CMR were done.
Results: In all the 24 cases, we could accurately visualize the size, mobility, tethering and degree of displacement of the leaflets using 3D echocardiography techniques. These findings were confirmed by CMR. The morphological details of tricuspid valves were better seen with 3D echocardiography, while right ventricular (RV) volumes were better assessed with CMR. The atrialized RV [r = 0.7, (p = 0.05)] and functional RV size estimated by 3D echocardiography and CMR correlated well, but RV systolic volumes and ejection fraction by both the techniques did not correlate well with each other. The most interesting finding on CMR is the presence of late Gadolinium enhancement in septal tricuspid leaflet and interventricular septum, which correlated with the severity of Ebstein's anomaly as assessed by Carpentier classification.
Conclusion: Both 3D echocardiography and CMR complement each other for complete morphological and functional evaluation, which is important for planning the optimal treatment strategy in Ebstein's anomaly.

MP2-17

Contrast echocardiography for screening of pulmonary arterio-venous fistula in children with portal hypertension

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Introduction: The hepato-pulmonary syndrome consists of intrapulmonary shunting (IPS) secondary to pulmonary arterio-venous fistulas (PAVF) in the setting of chronic liver disease and portal hypertension. The objective of this study was to establish the diagnostic value of contrast echocardiography (CE) as a screening technique for IPS in children with portal hypertension using lung scintigraphy as the gold standard.
Methods: CE was performed in children with portal hypertension using an intravenous line inserted preferably in the antecubital region. CE was done using microbubbles created by hand-

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agitated saline solution. IPS positivity was defined as the appearance of microbubbles in the left atrium within <5 heart cycles after complete opacification of the right atrium. A lung scintigraphy was performed within 4 weeks with IPS presence graded from 1-4 according to the amount of shunt.
Results: 30 children with portal hypertension underwent CE and lung scintigraphy. The mean age was 9.9 ± 3.8 years (2.1-16.6), with no difference in gender distribution (18 girls, 60%). The origin of portal hypertension was pre-hepatic in 6 (20%), intra-hepatic in 22 (73%) and secondary to a congenital or surgical shunt in 2 (7%). Scintigraphy was negative in 24 (80%) and positive for IPS in 6 (20%). All patients with a negative lung scintigraphy had a negative CE. In the 6 patients with a positive lung scintigraphy, 5 had a positive CE while 1 had a negative CE (false negative). The sensitivity of CE was 83% (CI 36.5-99.1) with a specificity of 100% (CI 82.8-100). The negative likelihood ratio was 17% (CI 0.03-0.99) and the negative predictive value 96%.
Conclusion: CE is a simple and reliable screening tool for PAVF and IPS in children with portal hypertension, with a sensitivity of 83% and a negative predictive value of 96% Technical limitations may be an issue particularly in small children when rapid saline injection is limited by the small size of the intravenous catheter. Detection of IPS is of utmost importance because hepato-pulmonary syndrome is now considered an indication for liver transplantation since the shunting disappears following liver transplantation.

Table: Sensitivity, Specificity, Positive (LHR +) and Negative (LHR –) Likelihood Ratio Values, Positive (PPV) and Negative (NPV) Predictive Values of Contrast Echocardiography for Shunt Prediction

Sensitivity (% [95% CI])	Specificity (% [95% CI])	LHR + (% [95% CI])	LHR – (% [95% CI])	PPV (%)	NPV (%)
83.3 [36.5 – 99.1]	*100 [82.8 – 100]*	*∞*	*0.17 [0.03 – 0.99]*	*1*	*0.96*

MP2-18
From Studies to Clinical Practice: Screening for Anthracycline-Induced Cardiomyopathies with 2-dimensional Speckle-Tracking Derived Global Strain and Strain Rate – A Feasibility Study in Consecutive Outpatients
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Introduction: Studies showed that 2-dimensional speckle-tracking derived strain and strain rate can detect left ventricular systolic dysfunction earlier than ejection fraction or fractional shortening (FS). This study aimed to evaluate the potential value of different strain and strain rate parameters for the follow-up of anthracycline-treated patients in daily clinical practice.
Methods: We included all anthracycline-treated patients who attended our outpatient unit in a one-year period for routine echocardiographic follow-up examination. An age and sex-matched healthy control group was established. Global longitudinal strain and strain rate (GSL, GSrL) were derived from the apical four-chamber (4CH) view, and global circumferential and average radial strain and strain rate (GSC, GSrC, AvSR, AvSrR) from a parasternal short axis (SAX) view. Global values were given directly by the software; radial averages were calculated by the observer. Intra- and inter-observer variations were measured in 40 randomly selected subjects by the Bland-Altman method, the coefficient of variation (CoV) and intraclass correlation coefficient (ICC).
Results: 131 patients and 66 control subjects were included. The feasibility of tracking the whole ventricular wall was better in the 4CH than in the SAX (76% vs. 62.4%). In only 48% it was possible to study both axes. Most often inadequate tracking occurred in the 4CH's apical-lateral (17%) and the SAX's lateral segment (29%). GSL, GSrL, GSC, GSrC and FS were significantly lower in patients than controls (-20.3 ± 1.8% vs. -19.2 ± 1.9%; -1.11 ± 0.12/s vs. -1.02 ± 0.16/s; -19.8 ± 3.1% vs. -17.6 ± 3.0%; -1.27 ± 0.22/s vs. -1.13 ± 0.20/s; 35.5 ± 3.5% vs. 32.7 ± 4.1% ($p < 0.005$)). AvSR and AvSrR were not significantly lower in patients (51.2 ± 13.1% vs. 46.2 ± 14.1%; 1.94 ± 0.33/s vs. 1.85 ± 0.38/s ($p > 0.05$)). Only GSC and GSrC showed significant results throughout in subgroup-analyses (gender, diagnosis and cumulative anthracycline dose). GSL showed the best intra- and interobserver reproducibility without bias, CoV <5% and ICC >0.90.
Conclusions: For clinical practice GSL and GSrL provide the best combination of feasibility, sensibility and reproducibility. However GSC and GSrC seem to be the most sensitive measurements in patients with suitable echocardiographic windows. AvSR and AvSrR showed several disadvantages and failed to detect a difference between patients and controls.

MP2-19
The Effect of Gender and Heart Rate on 2-dimensional Speckle-Tracking derived Strain and Strain Rate in Children and Young Adults
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Introduction: Speckle Tracking is of great interest in research and is playing an increasing role in clinical practice. The aim of this study was to determine the influence of gender, age, body surface area (BSA) and heart rate (HR) on 2-dimensional speckle-tracking derived strain and strain rate in children and young adults.
Methods: 66 healthy subjects (52% female) with a mean age of 16.5 ± 6.40 years (range 5 to 28 years), mean BSA of 1.56 ± 0.38 m^2 and mean HR of 73.4 ± 12.5/min were recruited. Global longitudinal and global circumferential strain and strain rate (GSL, GSrL, GSC and GSrC) as well as segment-derived average radial strain and strain rate (AvSR, AvSrR) were measured in the apical four-chamber or parasternal short axis view with dedicated software (EchoPAC, GE).
Results: Measurements of longitudinal parameters were feasible in 58 subjects, whereas circumferential and radial parameters could be measured in 40 cases: GSL -20.3 ± 1.8%, GSC -19.8 ± 3.1%, AvSR 51.2 ± 13.1%, GSrL -1.11 ± 0.12/s, GSrC -1.27 ± 0.22/s and AvSrR 1.94 ± 0.33/s. GSL was significantly higher in female than male subjects (-21.0 ± 1.6% vs. 19.5 ± 1.6%; $p < 0.001$). This gender difference was present in minors (<18 years) and young adults (≥18 years) (-20.7 ± 1.5% vs. -19.3 ± 1.6% and -21.4 ± 1.8 vs. -19.9 ± 1.6%; $p < 0.05$). No other strain or strain rate was affected by gender. All Strain Rates (GSrL, GSrC and AvSrR) were influenced by heart rate; strain rate increased as heart rate increased. The influence was less in global strain rates obtained with Lagrangian Strain (GSrL and GSrC both $R^2 = 0.15$, $p < 0.05$) than in averaged segmental Strain Rate obtained with Natural Strain (AvSrR $R^2 = 0.26$; $p < 0.005$). Linear regression showed no influence of age or BSA on measurements ($p > 0.05$).
Conclusions: This is the first study showing that GSL is significantly higher in girls than boys. Until now this was only known for the adult population. These results underline the importance of

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sex-matched groups in clinical studies and the need for the use of gender specific reference values. The influence of HR on Strain Rate should be kept in mind when interpreting results.

MP2-20

Subclinical Anthracycline Induced Cardiotoxicity in The Long-term Follow-up of Asymptomatic Childhood Cancer Survivors: a Speckle Tracking Echocardiographic Study

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Objectives: The aim of this study was to assess ooccult cardiotoxicity in childhood cancer survivors exposed to anthracycline therapy in the long-term follow-up.
Methods: We studied 45 survivors (26 male, 19 female) 5-20 years old, (median 11 years). and 38 (22 male, 16 female) healthy controls. Blood samples were taken from survivors to determine BNP levels. Left ventricular ejection fraction, fractional shortening, diastolic functions, tissue Doppler and myocardial performance index were measured. All subjects were assessed with tissue tracking 2D strain echocardiography. Regional and global strain parameters of survivors were compared with those in healthy controls and were related to conventional echocardiographic parameters, brain natriuretic peptide (BNP) levels and clinical parameters.
Results: There were not any significant differences in left ventricular ejection fraction, fractional shortening, and diastolic functions, tissue Doppler and myocardial performance index between two groups. Myocardial strain in asymptomatic survivors of childhood cancer was significantly lower compared with healthy controls. While 39% of survivors had at least one segment, which was impaired, there was not any healthy controls who have impaired segment. Radial strain did not differ significantly between the two groups.
Conclusion: Strain echocardiography seems more useful to detect antracycline cardiotoxicity. Whether myocardial strain are superior to conventional echocardiography in the early detection of subclinical cardiac toxicity needs to be explored in further longitudinal prospective studies

MP2-21

Residual right ventricular outflow tract obstruction preserves right ventricular contractility in patients after repair of tetralogy of Fallot: a CMR feature tracking study

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Background: Residual right ventricular outflow tract obstruction (RVOTO) is considered beneficial in patients after repair of tetralogy of Fallot (TOF) although underlying mechanisms are unknown. The aim of our study was to elucidate differences in myocardial contractility parameters in patients after TOF repair with and without residual RVOTO using dedicated cardiovascular magnetic resonance (CMR) feature tracking (FT) analysis.
Methods: Fifty-four patients (mean age 16.4 ± 8.4 years) were assessed by CMR 14.2 ± 7.3 years after repair of TOF. Residual RVOTO on echocardiography was defined as a peak systolic RVOT gradient >25 mmHg. Right (RV) and left ventricular (LV) strain measurements were performed using CMR-FT software (TomTec, Unterschleissheim, Germany).
Results: The groups (n = 27, respectively) were well matched for age at CMR-scan, time and type of surgical repair. There was no difference in the degree of pulmonary regurgitation (PR) and RV enddiastolic volume (RVEDV). Patients with RVOTO showed significant higher circumferential (CS) (-15.7 ± 4.0 vs -12.3 ± 5.8%; p = 0.02) and radial strain (RS) (15.0 ± 4.8 vs 11.8 ± 5.3%; p = 0.02) values, whereas longitudinal strain (LS) did not differ between the two groups (-9.9 ± 5.4 vs -11.5 ± 5.9%; p = 0.39). The magnitude of RVOTO showed a significant correlation with RV-CS (r = 0.37; p = 0.006) and RV-RS (r = 0.30; p = 0.03) while RV-LS was unrelated to RVOTO (r = 0.06; p = 0.68). RV strain parameters were not related to PR and RVEDV. Significant relationships between RV and LV strain parameters were only found in the RVOTO group. LV-LS was significantly lower in the group with RVOTO (-9.2 ± 5.2 vs -12.4 ± 5.0%; p = 0.03).
Conclusions: Residual RVOTO seems to preserve RV contractility in patients after TOF-repair and may therefore posses an early protective effect on RV remodelling. RVOTO appeared to result in stronger RV-LV interactions although its potential negative impact on LV strain needs further investigation.

MP2-22

Fetal Tachyarrhythmia – transplacentaltreatment and longterm outcome in 153 subjects

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Introduction: Fetal tachycardia carries a risk for morbidity and mortality and there is no consensus concerning medical treatment. The objective of the study was to evaluate the experience of fetal tachycardia in Stockholm and Gothenburg.
Methods: A retrospective, descriptive study including all consecutive pregnancies that presented with fetal tachycardia between January 1990 and December 2012 were conducted.
Results: A total of 153 pregnancies were identified, 98 had atrioventricular re-entry tachycardia (AVRT), 42 had atrial flutter (AF), 6 had permanent junctional reciprocating tachycardia (PJRT), one had atrial ectopic tachycardia (AET), three had atrial chaotic tachycardia (ACT), two had junctional ectopic tachycardia (JET) and one had ventricular tachycardia (VT). Ninety-seven fetuses received transplacental treatment; in foetuses without hydrops, when digoxin was used as first line treatment; 15 of 30 (50%) cardioverted, with sotalol as first line treatment 13 of 19 (68%) cardioverted, the combination of digoxin and sotalol led to cardioversion in 3 of 5 (60%) fetuses. In 46 of 153 (30%) cases hydrops was present, and with digoxin as first line treatment 5 of 19 (26%) cardioverted, with sotalol as first line treatment 6 of 14 (43%) cardioverted and the combination of digoxin and sotalol led to cardioversion 2 of 7 (28%) fetuses. Eight hydropic fetuses died 2-35 days (median 4) after diagnosis. Two children developed severe neurological symptoms (cerebral infarction) related to the tachycardia during the first week of life. At follow up median 8 years (0.5-22) 11 still have problems with arrhythmia, AVRT in nine and PJRT in two. One has undergone heart transplantation.
Conclusions: In the group of hydrops and AVRT 8 of 36 (22%) fetuses died. First-line treatment with digoxin and/or sotalol led to cardioversion in 26 – 43%. Death occurred at a median time of 4 days after diagnosis. At follow up 11 of 139 (8%) still have problems with arrhythmia.

MP2-23

Clinical presentation of isolated transposition of the great arteries in a complete population based cohort. Implications for pre- and postnatal screening routines

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Objectives: To determine to which degree pulse oximetry screening (POS) contribute to an earlier diagnosis of isolated transposition of the great arteries (TGA) and to estimate the proportion of neonates with TGA who would benefit from a prenatal diagnosis.
Methods: Infants with TGA born in our referral area for pediatric cardiac surgery from 2003-01-01 to 2013-08-01 were identified from our surgical files and from the causes of death registry (National Board of Health and Welfare). Data on clinical presentation, diagnosis, management and outcome were collected from hospital charts.
Results: 91 cases were identified of which 34 were born in hospitals using POS. 3 were diagnosed prenatally. 57 developed early symptoms and were diagnosed before routine newborn physical examination and before POS. 7 were detected by POS, 13 at the routine newborn physical examination, 1 after that examination but before discharge and 10 were discharged undiagnosed. Five of those discharged were detected at a routine follow-up visit (one at 6 weeks of age) and 5 came to hospital because of symptoms. None of 34 born in hospitals with POS were discharged undiagnosed compared to 10 of 57 in the remaining hospitals. The age at suspicion of congenital heart disease was significantly lower in those born in hospitals with POS. 61 underwent balloon atrial septostomy (BAS), in 9 cases before 6 hours of age and in 26 cases before 12 hours. One neonate died at 4 hours of age at the referring hospital after an unsuccessful BAS. There was no further deaths at a median follow-up of 4,4y (30d-10y). 9 children had neurological symptoms at follow-up of which 3 had neurological symptoms (seizures) already before the arterial switch operation. These three neonates had severe hypoxia (20-50%) and high levels of lactate at arrival at our centre. One needed CPR immediately after arrival. BAS was performed at 6, 8 and 14 hours of age respectively.
Conclusions: Although POS prevents discharge of infants with undiagnosed TGA, at least 4 of 91 (all born in hospitals using POS) would have potentially benefited from a prenatal diagnosis to avoid death or neurological sequelae.

MP2-24
Prenatal Diagnosis And Outcome For Fetuses With Congenital Absence of The Pulmonary Valve
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Objective: Absent pulmonary valve syndrome (APVS) is a rare congenital anomaly with its hallmark feature of a rudimentary and dysplastic pulmonary valve. In most instances, it is associated with severe dilatation of the main and branch pulmonary arteries (PA) owing to the combination of severe pulmonary stenosis and regurgitation. The aim of this study is to review the spectrum of the prenatally detected APVS and its outcome after diagnosis.
Methods: Clinical data and echocardiographic findings of 11 cases with a fetal diagnosis of APVS between 2008 and 2013 were analyzed in this retrospective two-center study. Collected parameters included: gestational age at referral; associated cardiac, genetic and non-cardiac fetal abnormalities; maximum diameters of the aortic and pulmonary annuli in addition to the main and branch pulmonary arteries.
Results: Median gestational age at diagnosis was 21 weeks. Four subtypes of APVS were observed: (1) with tetralogy of Fallot (TOF) (n = 6; 54%); (2) isolated (n = 3; 27,7%); (3) with CAVSD (n = 1; 9,9%); and (4) with VSD (n = 1; 9,9%). Ductus arteriosus was restricted in 5, absent in 3, and large in 3 fetuses. Two pregnancies were terminated. Two fetuses are still to be delivered. One fetus was stillborn who had trisomy 18. Of the six live births, one neonate died following birth due to severe hypoxia, 3 died after surgery, one remains well after operation, and the last patient is on medical follow up without operation. The presence of ductus arteriosus is not associated with survival. The genetic survey was abnormal in 27,7% of fetuses (trisomy 18 in one, and 22Q11microdeletion in two).
Conclusion: Prenatal diagnosis of the APVS is rather straightforward because of its typical features of a dilated main pulmonary and branch arteries, and color Doppler detection of severe stenosis and insufficiency of the functionally absent pulmonary valve. Outcome of antenatally APVS is poor and survival is mainly determined by the presence of respiratory symptoms due to bronchio-tracheal obstruction in this study.

MP2-25
Idiopathic dilatation of the right atrium. Report of four fetal cases.
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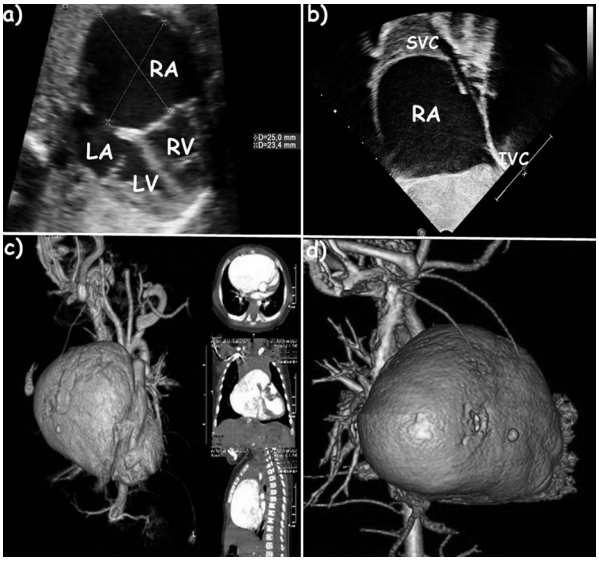
Introduction: Idiopathic dilatation of the right atrium (IDRA) is a rare anomaly defined as isolated enlargement of the right atrium in the absence of other cardiac lesions or predisposing conditions to cause right atrial (RA) dilatation, especially tricuspid valve diseases. The clinical presentation is highly variable from asymptomatic to cardiac failure or even sudden death. It can be associated with atrial arrhythmias, thrombus formation and congestive heart failure.



Methods: We report 4 cases of IDRA of prenatal diagnosis, three children and last one is still a prenatal case. We describe the intrauterine course, the postnatal management and its short-medium term follow-up. Echocardiography of the fetus showed an abnormal 4-chamber view with an increased cardiothoracic circumference ratio at the expense of enlarged RA without other anomalies. During the routine controls the dilatation of RA persisted. None had extracardiac malformations. Postnatal echocardiography demonstrated that the RA was dilated with spontaneous echo contrast (Figure 1a-b). Although none had thrombus formation in the RA we decided initiate treatment due to a potential risk for pulmonary embolism. In all patients thrombophilia study was normal so we started thrombosis prophylaxis with salicylic acid. There has been no need for

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surgical intervention so far because of the lack of arrhythmias or symptoms, although one of our cases we have found an important progression of RA diameters. This patient is 4 months old and the RA is massively dilated (diameter of 65 mmX71 mm detected with angioCT). Cardiac resonance imaging showed global hypokinesia and he is awaiting surgical decision (Fig. 1c-d).

Conclusions: Optimal management of IDRA is controversial and depends on the individual case. This disease probably is under diagnosed, reinforced by the fact we have seen four cases, but all in the last two years.

Long-term follow-up is necessary to monitor progression of RA size and occurrence of arrhythmias. Asymptomatic patients can be managed medically but symptomatic patients may require surgical reduction of the RA. Different imaging techniques including computed tomography and cardiac resonance imaging are useful to diagnose, evaluate and perhaps it may be helpful in treatment decisions. According to Paladini a Holt-Oram syndrome should be ruled out.

MP2-26

Fetal aortic valvuloplasty – how to improve outcome ?

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Introduction: Fetal balloon aortic valvuloplasty F-BAV has already been performed in few institution all over the world. The two biggest series published different outcome: 30% of bivetricular circulation (BV) in Boston and 70% in Linz. There is still not consenus what is the best treatment for neonates and infants after FBAV. Prenatal natural and after FBAV history is still far from understanding. Knowing this we started the program of fetal cardiac interventions in 2011. The objective of this study was evaluation of preliminary results of FBAV.

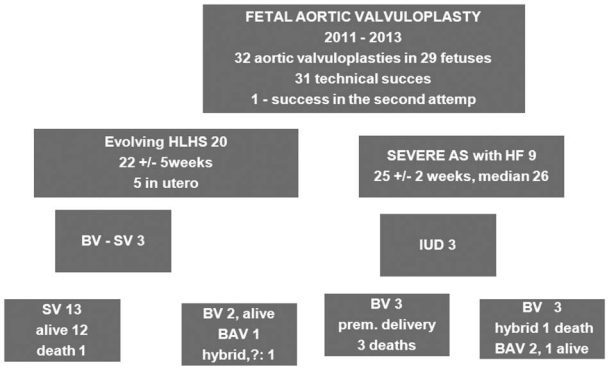
Material: Between 2011–2013 32 FBAV was performed in 29 fetuses. Fetuses were divided into two gropus: evolvingHLHS (eHLHS)-20; severe AS with heart failure-9.

Results: Aortic valve was successfully dilated in all 29 fetuses. 11 procedures were done under general anesthesia of the mother, 13 – intravenous, recent 6 – local. All fetuses had intraumbilical analgesia with fentanyl.

In 20 fetuses with eHLHS there was better flow through the aortic valve and better LV function after the procedure. In spite of this just 2 had biventrivular circulation. One was switched to BV after hybrid procedure in the neonatal period. 15 fetuses from the eHLHS group survived neonatal and early infants period. In 3 the first attempt was BV circulation, but it had to be switched to SV due to very poor LV function. One child from this group died due to severe heart and multiorgan failure.

Fetuses from the second group were in worse condition. 3 were hydropic, 3-severe LVdysfunction with closed Fo and polyhydramnion. In spite of successful procedures, 3 died in utero, 3 were born premature, 2 died without treatment,1 after 2 BAV in the neonatal period. The last after FBAV and stent placement into IAS, died in th 5th week of life after hybrid procedure. There is just one BV survivor from the second group.

Conclusions: FBAV can be successfully performed. The prenatal course after successful dilation of the aortic valve is unpredictable. Fetuses with severe heart failure are in much higher risk than eHLHS. The best postnatal treatment of this difficult patients should be the topic of international discussion.



MP3-1

Long-term fate of children operated for the hypoplastic left heart syndrome in a country with high foetal termination rate and centralized paediatric cardiovascular care

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Objectives: To evaluate long-term results of surgical palliation for the hypoplastic left heart syndrome (HLHS) on a territory (Czech Republic, 10.5 million inhabitants) with a country-wide prenatal detection program (estimated detection rate of HLHS >90%, termination rate of detected HLHS 87%) and postnatal care centralized to one paediatric cardiovascular centre.

Methods: Retrospective analysis of all consecutive newborns with HLHS admitted between 1999-2012. Non-obligatory re-interventions were defined as those additional to the 3 stages of the Norwood pathway.

Results: From a total of 65 consecutive newborns with HLHS 13 pts. (20%) did not receive surgical treatment because of parental decision, associated anomalies or non-fulfilment of the indication criteria. 52 patients (prenatal diagnosis in 33%) were directed to the Norwood pathway (median age/weight 7 days/3.2 kg). Early/total mortality after Norwood stage I was 9.6/19% (10/52 pts.) with a significant risk factor being lower weight at surgery (Cox proportional risk per 1 gram = 0.997, CI 0.995-0.990, $p < 0.001$). Between Norwood stage I and II 15 catheter/surgical re-intervention were carried out in 13 pts. (aortic arch narrowing in 10/13). 42 pts. aged median 6.8 months underwent stage II surgery with a total mortality of 4.8% and 18 subsequent re-interventions in 13 patients. Finally, 26 patients aged median 3.9 years underwent Fontan completion with early/total mortality 0 and 8%, resp. The probability of survival at 1/5/10 years of age was 77/77/71 %. Probability of freedom from non-obligatory surgical/catheter reinterventions was 58/45/41 %. At long-term follow-up (median 7.8 years) 37/38 pts. are in NYHA functional class I or II.

Conclusions: Due to high foetal termination rate the population of live-born HLHS patients is biased towards those without a prenatal diagnosis. Despite a highly centralized care, surgical treatment of HLHS is still associated with significant mortality and morbidity. Long-term survivals, however, have an acceptable

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functional status during childhood corresponding to other groups of patients after surgical palliation for functionally single ventricle.
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MP3-2

Early clinical outcomes for fetuses with severe or greater tricuspid regurgitation in the era of cone reconstruction

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Objectives: Since cone tricuspid valve (TV) reconstruction was introduced, the treatment strategy for fetuses with severe or greater tricuspid regurgitation (TR) has been dramatically changed. However, surgical indications and criteria for biventricular repair (BVR) still remain unclear.
Methods: During 2013, 5 fetuses were diagnosed with severe or greater TR and functional (n = 2) or membranous (n = 3) pulmonary atresia related with Ebstein anomaly (n = 3) or congenital dysplastic TV (n = 2). The median cardiothoracic area ratio was 0.69 (range, 0.64-0.70). Three babies were prematurely born due to fetal hydrops (n = 2) or fetal growth restriction (n = 1) and the median birth weight was 2982 g (1712-3240). One baby was diagnosed with Down syndrome.
Results: Soon after birth, mechanical ventilator support was initiated in 4 patients and peritoneal dialysis was performed to attenuate whole body edema in 2 patients. The median cardiothoracic ratio (CTR) and TR pressure gradient (TRPG) was 91.0% (84.3-91.5) and 27.0 mmHg (17.4-49.0). One patient with fetal hydrops died at 4 days of age without surgical intervention due to low output syndrome. The remaining 4 patients underwent complete BVR consisting of cone reconstruction, the creation of right ventricle to pulmonary artery continuity, and closure of atrial communication at the median age of 10.5 days (8-15). Right atrial cryoablation was concomitantly performed in 3 patients with ectopic atrial tachycardia. TR decreased to a moderate severity level in all patients. There was 1 in-hospital mortality on post-operative day 41 due to persistent pulmonary hypertension related to chromosomal anomaly, and 1 late mortality on day 56 due to septic shock caused by necrotizing enterocolitis The latter patient underwent the Starnes operation conversion as a single ventricular palliation 8 days after BVR because the right ventricle did not compensate. Two patients successfully survived at discharge and the latest follow-up confirmed that the TR grade remained moderate and the CTR was less than 70%.
Conclusions: With cone reconstruction, 2 of 5 critically ill patients successfully obtained complete biventricular circulation. TRPG of more than 40 mmHg without primary pulmonary arterial hypertension seems to be essential for BVR.

MP3-3

Performance and survival of epicardial leads in pediatric patients and congenital heart disease patients. A 15 years single center experience with two unusual cases and a review of the literature

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Objective: Cardiac pacing is needed frequently in patients with congenital heart disease for various reasons, ranging from symptomatic bradycardia due to sinus node dysfunction or postoperative complete heart block to cardiac resynchronization therapy. As the use of epicardial leads has been discussed controversly as far as pacing performance and lead survival are concerned, the actual study adresses this topic.
Design: Retrospective chart review and review of the literature.
Patients: 84 consecutive pediatric or adult patients with congenital heart disease were included. This accounts for 164 epicardial pacemaker leads.
Results: We found (1,2%) pacemaker related early postoperative complications. The incidence of lead dysfunction was 7,3% (12/164) for primary and 3,04% (5/164) for secondary dysfunction. Primary dysfunction occured after a median of 3,44 years (1,77–5,13). Reasons for primary lead dysfunction were lead fracture (n = 10) and macrodislocation (n = 2). There were no infections reported in this study group. Stable median measurements for impedance (RA/RV/LV of 577/482/610 Ohm), sensing threshold (RA/RV/LV of 2,0/11,0/10,6 mV) and pacing threshold (RA/RV/LV of 0,75 V at 0,4 ms/1,0 V at 0,49 ms/1,0 V at 0,4 ms) revealed a good mid- to longterm performance. The only risk factor for primary lead dysfunction identified, was young age at implantation.
Conclusion: The use of epicardial leads in the setting of pediatric and adult patients with congenital heart disease shows comparable long-term outcomes and equal effectiveness as endocardial leads. Therefore the decision on whether to implant endo- or epicardial leads should rather be based on the patients individual characteristics than on technical aspects concerning lead performance or durability.

MP3-4

Early bidirectional cavopulmonary anastomosis: impact on outcome and Fontan completion

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Introduction: The interstage mortality is the highest before bidirectional cavopulmonary anastomosis (BCPA) on the way to Fontan completion. The BCPA improves hemodynamic conditions of systemic-pulmonary shunt physiology. BCPA at younger age would shorten the high risk period characterized by volume load, pathological coronary flow pattern and risk of acute shunt closure. The aim of this study was to evaluate the impact of age at time of BCPA on outcome.
Methods: Retrospective analysis of patient records from patients who underwent BCPA between January 2006 and April 2013. With regard to age at surgery we classified the infants into two groups: group A ⩽90 d and group B 91–183 d and compared early and late outcome parameters.
Results: Group A comprised 32 and group B 112 infants (median age 78 vs. 124 d; $p < 0.01$). Postoperative course: The mean arterial oxygen saturation (maSO2) within 24 h after BCPA was 72% in group A, and 75% in group B ($p = 0.047$). MaSO2 after 24 h, time on respirator, need for oxygen or tcSO2 at discharge

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were not significantly different. Early outcome: there was no significant difference regarding postoperative complications between groups. There was no early death in group A but 6% in group B died within 30 days after BCPA ($p = 0.15$). Late outcome: The invasive hemodynamic data as well as the incidence of major collaterals or pulmonary artery stenosis before total cavopulmonary connection (TCPC) showed no significant difference between groups. At time of admission for TCPC weight (11 vs. 11 kg; $p = 0.5$), tcSO2 (83 vs. 82%; $p = 0.8$) and time after BCPA (18 vs. 17 months; $p = 0.6$) were comparable. At the end of the study 20 patients (63%) in group A and 77 (69%) in group B were completed to TCPC ($p = 0.5$). The late mortality was 3% in group A and 9% in group B ($p = 0.5$).
Conclusion: BCPA below 91 days of age to improve hemodynamic conditions of systemic-pulmonary shunt physiology is feasible without serious complications or deleterious influence on outcome.

MP3-5
Right ventricular outflow tract reconstruction without conduit in neonates with tetralogy of Fallot: comparison of pulmonary stenosis vs. pulmonary atresia
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Introduction: Early primary repair has been applied in tetralogy of Fallot with pulmonary atresia and well-developed central pulmonary arteries supplied by a ductus arteriousus (TOF/PA). A valve conduit for the right ventricular outflow tract (RVOT) reconstruction is extensively used in this group of neonates. Its use is considered as contributing factor to lower freedom from reintervention comparing to neonatal repair of tetralogy of Fallot with pulmonary stenosis (TOF/PS). Since 1997 our institutional approach was to perform primary RVOT reconstruction in TOF/PA with avoidance of conduit placement. We sought to determine early and long-term results of this strategy for TOF/PA and compared them with results of neonatal transannular patch repair in TOF/PS.
Methods: This is a retrospective review of 21 neonates with TOF undergoing RVOT reconstruction without use of conduit at a single centre between 1997 and 2013. Nine TOF/PS and 8 TOF/PA patients underwent transannular patch repair and 4 TOF/PA neonates without continuity between main pulmonary trunk and right ventricle received direct anastomosis of RVOT to main pulmonary artery supported by pericardial patch. During the same period, in 2 neonates with TOF/PA the primary conduit placement was needed.
Results: The mean age at primary repair was 17.5 ± 7 days for TOF/PS and 10.4 ± 6 days for TOF/PA ($p = 0.025$). The hospital survival was 100% for both analyzed groups. Early postoperative course was comparable in both groups according to duration of mechanical ventilation (96 vs. 84 hours; $p = 0.9$), maximum vasoactive-inotropic score (10 vs. 9; $p = 0.78$), intensive care unit stay (6 vs. 7 days; $p = 0.85$) and hospital stay (11 vs.14 days; $p = 0.61$). One patient with TOF/PA died during follow-up. Overall freedom from RVOT reintervention during median follow-up of 9 years (0.3-16.3) was 77.8% in TOF/PS and 72.7% in TOF/PA group ($p = 0.79$). There was no difference in right ventricular end-diastolic dimension, tricuspid regurgitation, QRS duration and gradient across RVOT at 1, 5 and 10 years.
Conclusions: In our limited experience, RVOT reconstruction with avoidance of conduit placement can be safely accomplished in majority of neonates with TOF/PA. RVOT reintervention rate during follow-up is comparable to TOF/PS after neonatal transannular patch.

MP3-6
Safety and Efficacy of Percutaneous closure of ventricular septal defect with Amplatzer duct occluder II in small children and adults
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Background: Nonsurgical closure of congenital ventricular septal defects (VSD) has become increasingly acceptable with the availability of different occlusion system. Percutaneous and transcather device treatment is used for perimembranous and muscular defects. Atrio-ventricular block (AVB) remains the most troublesome complication of device closure. The aim of this study is to describe our experience with closure of ventricular septal defects (perimembranous and muscular) using the Amplatzer Duct Occluder II (ADO II) as an "off-label" approach.
Methods: Between 2004 and 2012 transcatheter closure of 31 VSD (20 perimembranous, 10 muscular VSD and 1 ruptured sinus valsalva) with ADO II was undertaken in patients between 3 months and 55 years of age and with a body weight ranging from 4 to 105 kg in our institution.
Results: In 29 of 31 procedures the defect was successfully closed (93,5%) without any significant complications. No increase of aortic and tricuspid valve regurgitation in any case after procedure. Small residual shunts were observed immediately after the device implantation, but disappeared during a median follow-up period of 38 months (0,4-63) in 27 of 31 patients. There was no incidence of AV-block or other conductance abnormalities during implantation or follow-up.
Conclusion: The ADO II device is safe and effective for transcatheter VSD closure, but still it is a "off-label" use. After long-term follow-up in a large number this device may also admitted for VSD-closure in the future.

MP3-7
Long-term outcomes and complications of transvenous pacemaker implantation in small infants
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Objectives: Evaluation of long-term outcome and complications of transvenous pacemaker implantation in paediatric patients weighing less than 10 kg.
Methods: We retrospectively analysed the outcome and complications of all implantations in small children (<10 kg) of pacemakers (PM) with transvenous leads between September 1997 and October 2001. Indications for PM-implantation, age at implantation and follow-up duration were noted. Furthermore, both cardiac and PM function and complications due to transvenous approach were evaluated.
Results: During the study period 7 patients underwent implantation of a VVI(R) PM system with a transvenous lead. The median age at implantation was 54 days (range, 1 day–1.13 years), the median weight 4.7 kg (range, 2.3–8.7 kg) and the median duration of follow-up 13.7 years (range, 12.1–16 years). Indications for PM-placement were congenital complete heart

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block in 4 patients, long QT-syndrome type II with AV-node dysfunction in 2 and postoperative heart block and sinus node dysfunction in 1. There were no procedural complications. All patients are still alive, with currently no cardiac complaints and a good PM-function. Four patients got an upgrade to a DDD system. Two patients suffered from vascular occlusion of the left subclavian vein, 6 and 8 years after PM-implantation respectively. In 2 patients a small thrombosis on the PM-electrode developed, 7 and 11 years after PM-implantation respectively; both successfully treated with anticoagulants. In 3 patients the PM-system was converted to an epicardial system due to symptomatic vein occlusion, systolic dysfunction and atrial perforation. One patient developed dilated cardiomyopathy. Six patients developed mild to severe tricuspid valve insufficiency after lead placement, necessitating surgical tricuspid valve repair in 1. Two patients underwent relocation of the PM-battery due to skin traction and 2 patients underwent repositioning of their leads to the right atrium after observance of PM-dysfunction, both within 2 years after PM-implantation. Five patients needed removal of their old PM-electrode because of lead replacement (n = 1), atrial perforation (n = 1) and switch to an epicardial system (n = 3).
Conclusion: Transvenous lead implantation in very small children is associated with a high incidence of severe complications and should be avoided in this patients.

MP3-8
Transcatheter closure of congenital Coronary artery fistulae – Medium to long-term Outcome
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Introduction: Coronary artery fistulas are rare congenital malformations and have been seen in 0.3% of patients with congenital heart disease and 0.06% of children undergoing echocardiography. Transcatheter closure of coronary artery fistulae (CAF) has emerged as an alternative to surgery, but there is paucity of literature with regards to long term outcome.
Objectives: This study was aimed to assess mid to long term outcomes in a series of patients who underwent transcatheter closure of coronary artery fistulae (CAF) at a tertiary cardiac centre.
Methods: It was a retrospective review of all patients with CAF who underwent transcatheter closure at the Institut Jantung Negara (IJN) between 1996 and 2008. Patients who had closure after 2008 were not included so that a reasonable medium to long term follow up could be obtained.
Results: A total of 33 patients aged 2 m to 47 years old with congenital CAF underwent percutaneous transcatheter closure using various devices. 19 were males and median age at intervention was 10 years (Range 2 m–47 yrs). Incidental heart murmur was the most frequent presentation present in 50% of cases. Most common site of fistula (50%) was between left coronary artery to right atrium. An angiogram right after device deployment revealed complete occlusion in 28 patients and trivial- to mild residual flow in 4 patients. Long term follow up was available in 22 patients and ranged from 5–15 years. There were no early or late deaths. All patients were asymptomatic. Only three patients needed repeat angiograms and one patient had significant residual shunt and needed reintervention to close the defect. Two complications noted early on. One patient had occlusion of LAD which was noted soon after and needed stenting of LAD. Another patient had embolisation of coil into LV and it was retrieved successfully.
Conclusions: Transcatheter closure of CAF is feasible with excellent outcome and can be considered as first line intervention as compared to surgery. Intermediate and long term outcome is favourable without any evidence of significant recananlization.

MP3-9
The pediatric interventionist "lost" in the upper abdomen – diagnosis and management of congenital portosystemic venous shunts, liver hemangioma and portal vein thrombosis in children
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Objective: Congenital and acquired diseases affecting the portal vein system and liver hemangioma may become subject of invasive diagnosis and treatment using modern catheter techniques.
Methods: In an institutional case series all consecutive pediatric patients with different types of portosystemic shunt malformations (PSM), liver hemangioma (LH) and portal vein thrombosis (PVT) were reviewed for diagnostic and therapeutic interventional management.
Results: Twelve children at a median age of 2.4 years (range 0-21) and weight 12.6 kg (2.7-62) with different types of PSM (n = 6), infantile LH (n = 3), and PVT (n = 3) were treated between May 2005 and December 2013. Invasive hemodynamic diagnostics focussed on evaluation of pulmonary arterial hypertension (testing) and angiographic diagnostic investigation, which was performed by selective angiography of mesenteric artery, retrograde wedge angiography by occlusion of fistula, or direct angiography of fistula. Catheter interventional treatment in PSM – if suitable – included stepwise partial (n = 2) or complete (n = 2) occlusion depending on hypoplasia of intrahepatic portal veins using Amplatzer vascular plug, detachable coils, and diabolo-shaped covered stents. For LH interventional closure of feeding hepatic arteries was performed using detachable coils. Due to the comorbidity of LH with Kasabach Meritt syndrome we had one postinterventional death.
Conclusions: Diagnostic catheterization with angiography and hemodynamic evaluation provides important information regarding pulmonary and portal hypertension in order to plan any staged catheter interventional partial or complete closure of portocaval fistula, if suitable. Special attention has to be given to neonates with LH, which are severely ill, and target-oriented and quick interdisciplinary team work is necessary to achieve optimal clinical outcome.

MP3-10
Use of 3 dimensional rotational angiography in pediatric interventional cardiology.
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Introduction: 3 dimensional rotational angiography (3DRA) has recently become available in the pediatric catheterisation laboratory. It provides 3 dimensional images of the heart and great vessels and may be used as roadmap during interventions. The aim of this study is to investigate the current use of 3DRA in pulmonary artery (PA) and coarctation (CoA) stenting.

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Methods: We retrospectively reviewed our catheterisation database to obtain all percutaneous interventions performed in children <18 years of age during 2012 and 2013. 3DRA guided PA and CoA stent placements were selected. Patient diagnosis and intervention were noted and results of the intervention and additional value of 3DRA were evaluated.
Results: During the study period 492 interventions were performed, 77 concerning 3DRA guided stent placement in PA (n = 60) or CoA (n = 17). Underlying diagnosis in patients with PA stenting were Tetralogy of Fallot (n = 13), Pulmonary Atresia (n = 11), Transposition of the Great Arteries (n = 12), univentricular heart with shunt (n = 1), Partial Cavo Pulmonary Connection (n = 5), Total Cavo Pulmonary Connection (n = 5), Alagille syndrome (n = 4), Truncus Arteriosus/occult pulmonary artery (n = 3), homograft stenosis post Ross-Kono procedure (n = 4), Williams syndrome (n = 1) and supravalvular and PA branch stenosis (n = 1). PA interventions performed were Melody procedure (n = 7), RPA or LPA stenting (n = 35), RPA and LPA stenting (n = 13), PA stenting (n = 2) and MAPCA stenting (n = 3). 3DRA provided excellent 3D information of the great arteries and was considered of great additional value in detecting PA branch stenosis in Glen/Fontan circulation and after arterial switch operation due to its ability to provide cranial angulations. 3DRA provided additional information on the surrounding anatomical structures (e.g. coronary arteries and aorta in case of PA stenting). In 4 additional patients a Melody procedure was aborted due to close proximity of the coronary arteries clearly revealed by 3DRA. Finally 3DRA provided accurate roadmaps for the intervention.
Conclusion: 3DRA guided stent placement has become a routine procedure at our centre.
With 3DRA real time 3D anatomical information of the lesion and its surrounding structures is obtained and a roadmap to guide the intervention is provided. This leads to better understanding of the lesion and should lead to safer and more successful procedures.

MP3-11
Is percutaneous patent ducts arteriosus closure a safe and effective alternative to surgery in small infants?
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Background: Although transcatheter closure is nowadays considered treatment of choice of patent ductus arteriosus (PDA), this approach is still challenging in small infants (weight <6 Kg) in whom most of available devices are considered off-label. Aim of this study is to report on feasibility, safety and follow-up results of percutaneous PDA closure in infants weighing <6 Kg in a high-volume tertiary referral centre.
Methods: From April 2000 to December 2013, 22 of the 586 patients (3.7%) submitted to trans-catheter PDA closure at our Institution were <6 Kg. Their mean age and weight were 4.3 ± 2.4 months (range 0-8) and 4.9 ± 0.8 Kg (range 3.1-6), respectively. Indications for the procedure were clinical signs and symptoms of congestive heart failure, left chamber volume overload at echocardiography and/or need of anti-congestive drug therapy. Ductal occlusion was achieved under general anesthesia from venous (n = 11), arterial (n = 9) or combined approach (n = 2). Ductal morphology was conical in 50% (n = 11), tubular in 41% (n = 9) and window-like type in 9% (n = 2) of the cases. Two patients (9%) had associated defects (pulmonary sequestration) that were treated during the same procedure.
Results: The mean PDA diameter was 2.7 ± 1.2 mm (range 1.5–5.5 mm), resulting in moderate pulmonary artery hypertension (mean pressure 28 ± 9 mmHg, range 14-46 mmHg; RV/LV pressure ratio 0.6 ± 0.2, range 0.3-1) and moderate left-to-right shunt (mean QP/QS 2.4 ± 1, range 1.5–4.5). According to the patient's weight as well as ductal morphology and size, different devices were used [Amplatzer Duct Occluder type I (n = 10), Amplatzer Duct Occluder type II AS (n = 8) and controlled-release Cook coils (n = 4)]. Procedural feasibility was 100%, with a complication rate of 9% (moderate anemia submitted to blood transfusion in one patient and femoral artery thrombosis in one patient). No fatalities were recorded. Immediate occlusion rate was 36%, rising to 86.3% at the last follow-up control. Echocardiographic follow-up failed to show any significant aortic isthmus or pulmonary artery branches flow abnormalities.
Conclusion: Percutaneous closure of large, symptomatic PDA might be considered feasible, effective and safe also in small, low-weight infants, by choosing the device according to the patient's profile and ductal anatomy.

MP3-12
A population based study of survival and lifestyle assessment after staged surgical menagement of hypoplastic left heart syndrome in Croatia – 12 years study: Does the fact that surgery is performed in developed European centers but rest of care in small country effects survival and quality of life?
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Introduction: Since, all children born in Croatia with anatomy of hypoplastic left heart syndrome (HLHS) are operated in centers in Western Europe but the rest of care, including preoperative transport, postoperative and care after hospital discharge is performed in Croatia, we tried to evaluate our results and to test if the fact that surgery is performed abroad but rest of care here, effects interstage and late survival and quality of life.
Methods: All children with HLHS born in Croatia from 2002–2011, who underwent stage surgery were included and followed till July 2013. year. Patient status was determined from review of interinstitutional medical records and direct patient contact. To determine quality of life and functional status of survivers, symptom-limited cardiopulmonary exercise test was performed and the Pediatric Quality Of Life Inventory (PedsQL) Version 4.0 was completed.
Results: 62 patient were treated. The 1,2,5 and 10-year survival for the entire cohort was 61.3, 54.8, 53.2,51.6% respectively. The majority of deaths (50%) occurred at the time of stage I surgery, 43% at stage II, and one late death after Fontan procedure. The median age at stage I surgery was 12 days, 14.5% were older than 14 days, one had less than 3 kg, 9.7% had prenatal diagnoses. The median age at stage II surgery was 124 days (22 to 281). The Fontan procedure was performed in 28 patients, with median age

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of 37.9 months (14.3 to 57.8) and mean follow up 49.2 months (24–127). We determined no significant association between anatomic subtype, prenatal diagnoses, type of stage I anastomosis (Sano vrs Blalock Tausig), older age at stage I operation and survival. 93% of survivals fulfilled PedsQL and showed satisfactory physical, psychosocial, emotional and school functioning. 52% of patients with Fontan procedure older than 4 years, performed cardiopulmonary exercise test with mean peak oxygen uptake 30.1 ml/kg/min (range 24 to 37).
Conclusion: Our results showed that survival and quality of life is satisfactory and isn't effected with the fact that surgery is performed in developed centers but rest of care in small country as Croatia.

MP3-13
N-terminal pro-BNP Correlates with Myocardial Involvement in Patients with Acute Kawasaki Disease
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Introduction: N-terminal pro-BNP (NT-proBNP) has been recognized as a marker of Kawasaki disease (KD). This was based on the hypothesis that cardiac inflammation during acute KD causes myocardial wall stress. Coronary involvement is well established, myocardial injury however has not been well studied. The objective was to examine the correlation between serum values of NT-proBNP, systolic myocardial dysfunction, and electrocardiographic changes in KD patients upon onset of the disease.
Methods: Parameters of myocardial involvement determined by electrocardiogram (PR interval and QT dispersion) and by echocardiogram (shortening and ejection fractions, left ventricle mass) were correlated with serum level of NT-proBNP in the acute phase of Kawasaki disease. KD patients were further subdivided into 2 groups according to the levels of NT-proBNP, KD-1 with normal NT-proBNP (NT-proBNP Z-score <2), KD-2 with elevated NT-proBNP (Z-score >2) and compared to a febrile control group.
Results: There were a total of 56 subjects, 14 controls, 19 KD-1 and 23 KD-2 patients. Age was similar between groups (Control vs KD, 3.77 ± 4.31 vs 3.32 ± 2.27 years-old, $p = 0.609$). There was a significant difference for the NT-proBNP z-score between controls and KD patients (0.87 ± 1.41 vs 2.26 ± 1.24, $p = 0.024$). There was a significantly reduced shortening fraction in KD patients, more intensely in KD-2 as witnessed by a diminished shortening fraction Z-score (-0.43 ± 1.49 in KD-1 vs -1.66 ± 1.46 in KD-2; $p = 0.012$). There was also a lower ejection fraction in KD-2 compared to KD-1 ($59.61 \pm 4.26\%$ KD-1 vs $55.11 \pm 9.05\%$ KD-2; $p = 0.047$). In contrast, there were no significant differences for left ventricular (LV) mass ($p = 0.939$), index LV mass ($p = 0.063$) or LV diameter ($p = 0.805$). Likewise, there were no significant differences for the PR interval ($p = 0.476$) or QT dispersion ($p = 0.580$).
Conclusions: In acute KD systolic dysfunction is present, especially in cases with elevated NT-proBNP. There does not seem to be any ECG changes between groups. KD patients with elevated zNT-proBNP may warrant specific myocardial follow-up and assessment even in the absence of CA involvement.

MP3-14
Value of research of plasma homocysteine-risk factor for cardiovascular diseases, in obese school children
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Background: Elevated plasma homocysteine level is an independent risk factor for cardiovascular disease and a sensitive marker of inadequate vitamin B12 and folate status. Such high values were highlighted and in the child obesity, homocysteine can play a role in the higher risk for cardiovascular disease in these children.
Purpose: to research plasma Hcy values in obese child compared with a control group of non obese children and correlations with blood pressure values and blood levels of total cholesterol and tryglyceride.
Methods: Patients: 41 children aged 8-18 years of which 31 obese and 10 nonobese healthy children, without cardiovascular diseases. All children received clinical examination, determination of plasma levels of homocysteine in the morning after 12-hour fasting, total cholesterol, tryglyceride, bloog glucose. In all children was assessed morphological and functional cardiac aspects by echocardiography.
Results: Plasma levels of homocysteine were elevated in 41,6% of children with obesity, compared to the control values:the mean $8,9 \pm 1,7$ μmol/vs $6,1 \pm 2,4$ μmol/. Increased levels of Hcy in obese children was more important to the in obese children with hypertension (5 cases). Elevated Hcy were often associated with high total cholesterol ± triglycerides in obese children compared with control values. Blood glucose levels were normal. Eccocardiographic changes in obese children: slight increase in LV wall thickness, dilated left atrium, LV diastolic dysfunction in 35% of cases, but with normal LV ejection fraction, mainly in cases the most increased values of Hcy. C onclusions. In obese children and mainly in those with hypertension were noted an increase in plasma homocysteine values often associated with elevated total cholesterol ± triglyceride levels. Research and systematic monitoring of values of plasma homocysteine correlated with cholesterol and triglyceride levels allows an objective estimation of cardiovascular risk in children with obesity and an indication for cardiovascular prevention effective measures.

MP3-15
An Audit of Rheumatic Fever in South Wales Region
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Introduction: We carried out a retrospective case-note based audit of Rheumatic Fever in South Wales region for the past 17 years. The objective of the study was to see if the diagnosis of Rheumatic Fever was made correctly according to Jones criteria, and that the management and prophylactic antibiotic was administered appropriately.
Methods: We performed a search of 'Rheumatic Fever' from our computer database (Cardiobase) between the year of 1996 to 2013.
Results: 31 patients met Jones criteria, with 9 patients labelled as probable Rheumatic Fever. 19 men (47%) and 21 women (53%), with a median age at diagnosis of 9 years and an incidence of 2-3 per year. The commonest Major criteria was carditis (72%), followed by chorea (45%), arthritis (35%), erythema marginatum (10%) and subcutaneous nodules (5%). Commonest Minor criteria was arthralgia (50%), followed by fever (42%), raised CRP (35%) and prolonged PR interval (12%). All 40 patients had evidence of Group A Steptococcal infection (29 with raised ASOT, 9 raised anti-DNAse and 5 with positive throat swab). Of the 29 patients with carditis, 26 had valvulitis, 4 pericarditis and 8 with dilated left heart. Only 17 patients (58%) had a clinical audible murmur. All 26 with valvulitis had Mitral valve involvement (100%), with 13 of them having both Mitral and Aortic Valve (50%) involvement. All patients received antibiotic to eradicate the infection. 31 patients who met Jones criteria had prophylactic antibiotic till at least 21 years of age, 6 of the

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probably Rheumatic Fever group received prophylactic antibiotic till 21 years of age, and the remaining 3 with probable Rheumatic Fever had prophylactic antibiotic for 1 year and discontinued. 14 patients received Aspirin treatment for arthritis, 3 had steroids for carditis, 7 patients needed ACE inhibitor and 4 had diuretics treatment. 1 patient unfortunately died from thrombus following complication of valve repair surgery. 3 patient received Haloperidol and 2 patients had Carbamazepine for chorea.
Conclusion: In conclusion, Rheumatic Fever is an uncommon disease but not rare, and all patients were diagnosed and managed appropriately with good long term outcome.

MP3-16
Health Related Quality of Life in Pediatric Patients with Marfan's syndrome
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Introduction: The "Health Related Quality of Life" (HRQOL) is a term that became increasingly important during the last 15 years. It is not yet known however if children with Marfan's syndrome are particularly impaired in different aspects of their Quality of life (QoL). We hypothesized that the QoL of patients with MFS is age-dependent due to increasing clinical features, and, compared to healthy children, reduced due to skeletal abnormalities, aortic root dilatation with consecutive need for drug therapy and regular medical checkup.
Understanding the relationship between subjective QoL in children with MFS has important implications for preventive care strategies that may impact the outcome of HRQoL.
Methods: 220 children between 4-16 years were included in this study. 46 patients (4-16 years, mean age 10.8 years) of our outpatient clinic completed the KINDL-R survey to assess life quality. Patients were included if MFS was determined genetically or clinically according to Ghent criteria, 39.1% were female, 60.8% male. Additionally data of 174 kindergarten and school children were collected (4-16 years, mean age 10.9 years), 48.8% were female, 51.1% male. Samples were divided into 4 age groups and subjective QoL was compared. Additionally we created subgroups based on the main diagnostic criteria of the revised Ghent nosology.
Results:

QoL total score	4-7 years	MFS 77.6 (95% ci 70.9-84.3)	control group 77 (95% ci 73.1-80.8)	$p>0.05$
QoL total score	8-11 years	MFS 75.3 (95% ci 68.5-82)	control group 74 (95% ci 70.8-77.1)	$p>0.05$
QoL total score	12-16 years	MFS 75 (95% ci 70.9-79.1)	control group 68.4 (95% ci 66.2-70.6)	$p<0.05$
QoL total score	8-16 years	MFS 75.1 (95% ci 71.5 –78.8)	control group 70.4 (95% ci 68.6-72.3)	$p<0.05$
QoL total score	8-16 years	MFS patients with lens luxation 81.6 (95% ci 75.2-88),	control group 70.5 (95% ci 68.6-72.3)	$p<0.05$

Conclusions: We report, that pediatric MFS patients showed partially higher HRQoL than children in the control group. MFS patients aged 12-16 years showed significant higher QoL than children in the control group although clinical features develop gradually. Analysis on different subgroups based on Ghent nosology confirmed these results. The results suggest that children with MFS experience increased QoL compared to children without this genetic predisposition. It can be speculated that parents may facilitate a stable social environment in the face of a children's disease. Furthermore children with MFS may be schooled through their environment to engage in more positive coping skills that increases their subjective experience of life quality.

MP3-17
Surgical treatment of infective endocarditis in children and young adults
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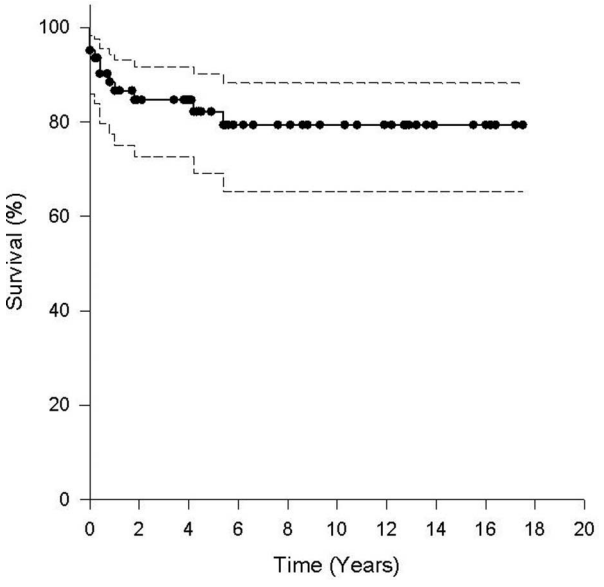
Objectives: To evaluate results of surgical treatment of infective endocarditis (IE) in childhood and adolescence.
Methods: A retrospective study of 63 consecutive pts undergoing surgery for IE between 1978 and 2013 (age 2 weeks–21 years, median 11 years) and followed-up for a median of 8.8 years.
Results: 54 pts (86%) had either surgically repaired (N = 31) or native (N = 23) congenital heart disease (CHD) with ventricular septal defect (N = 15, small perimembraneous native in 12/15) and common arterial trunk (N = 7) being most common. 9 pts (14%) had a structurally normal heart. The predominant pathogens were staphylococci (52%) and streptococci (31%), with negative IE cultures in 8%. The etiology of the initial bacteremia was unknown in 66.6% of pts. The most common indication for surgery was pulmonary or systemic embolism (N = 24), infected foreign material (N = 23, conduit in 10/23) and heart failure (N = 13). During surgery infected material was removed in all pts with 92% of pts undergoing associated procedures. Freedom from recurrence was 89% at 6 months. All pts with recurrence were subjected to repeated surgery. Hospital mortality was 11% (7 pts). Late death possibly related to IE occurred in another 3 (5%) pts (Figure).
Conclusions: Surgical treatment of IE in the young had an acceptable mortality and recurrence rate. Native small perimembranous ventricular septal defects and conduits were the two most commonly infected substrates. Etiology of the initial bacteremia was unknown in the majority.
(Supported by MH CZ-DRO, University Hospital Motol, Prague, Czech Republic 00064203).



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MP3-18

Relationships between renal vasculature abnormalities and arterial hypertension in children and adolescents

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Introduction: This study investigated hypertensive children for non-stenotic renal vascular abnormalities. There is no evidence in the literature of any correlation between these conditions.
Methods: Patients referred for elevated blood pressure were considered. Excluded from the study were those with hypertension secondary to known causes such as aortic coarctation, morbid obesity, hyperinsulinism, acute or chronic kidney disease, neurological diseases, oncological diseases and/or steroid therapy for other immunological diseases, and genetic diseases such as Williams syndrome or neurofibromatosis. Forty-three patients were selected with persistent hypertension (blood pressure $>95^{th}$ centile) of more than one year's duration and confirmed by 24 h ambulatory blood pressure monitoring. They were studied for sodium and caloric intake, blood chemistry studies for renal function (glomerular filtration rate GFR, renin and aldosterone), thyroid function, adrenal function (catecholamines, cortisol and ACTH), and abdominal ultrasound with Doppler study of the renal arteries. All patients underwent CT-angiography or MRI scanning to rule out possible renal artery stenosis and Echocardiograghy to evaluate left ventricular diastolic dysfunction/hypertrophy (LVDD/H).
Results: 37 patients (86%) had abnormal renal vasculature, either arterial or venous, in terms of the number of arteries, size and course. 26 pts (60%) had a polar accessory artery which was unilateral in 23 (89%) patients and in 3 (11%) was bilateral. Seven patients (16%) showed "nut-cracker syndrome" an abnormality of the left renal vein which coursed between the aorta and the mesenteric artery which formed an acute angle constricting the left renal vein as it went towards the inferior vena cava. Two patients (5%) had triple left renal veins and two (5%) showed a congenital gothic aortic arch. Only six patients (14%) showed a normal renal vasculature pattern. Eleven patients (25%) showed LVDD/H.
Conclusion: The incidence of hypertension in children is increasing in recent decades for several reasons, particularly obesity. Young patients with persistent elevated blood pressure also should be investigated for renal vasculature abnormalities and possible LVDD/H which may be treatable.

MP3-19

Establishment of a German Research Network for Congenital Heart Defects

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CNCHD Central Office Berlin, Germany (1); CNCHD Steering Committee (2); National Register for Congenital Heart Defects Management Board (3)

Objectives: The Competence Network for Congenital Heart Defects (CNCHD) was established to facilitate multicentre medical und socio-medical research in the field of congenital heart defects (CHD), which is intended to lead to an improvement of health care and out-come for this relatively new and continuously rising patient group. Due to the characteristics of this disease (high variability of morphological heart defects, chronic illness), the network infrastructure has to overcome specific challenges such as research in underage patients or the implementation of a legal framework for long-term storage of data and biomaterial.
Methods and Results: The CNCHD succeeded in implementing a sustainable research infrastructure involving all important stake-holders throughout Germany. The infrastructure is built around the non-profit registered association National Register for Congenital Heart Defects (NRCHD e. V.) that provides a dynamic and flexible IT-platform for different types of database-systems for register studies and a multicentre biorepository that collects blood-derived DNA and cardiac tissue from heart surgery. Thanks to central patient and ID management, data of different formats and recorded at different times can be clearly assigned to respective patients, thus allowing multicentre and longitudinal investigations. Electronic case report forms and remote data entry are used to centrally collect and store the data. Specific role based access rights management can be implemented for decentralised data entry by different users (physicians, researchers, documentation staff, monitors, sample laboratory etc.), e.g. within the scope of multicentre studies. This enables the integration of national and international research units, which is of particular importance with regard to recruiting new cooperation partners.
In January 2014 the National Register comprises 46.333 participants. The majority (64%) is underage, sex is evenly distributed. Simple heart lesions represent 38%, moderate 52%, and severe/complex 10%. The DNA collection currently comprises samples from aproximately 3000 participants covering a wide range of CHD phenotypes. The collection includes also trios (patient + unaffected parents) and families with more than one affected member.
Conclusions: The CNCHD provides a comprehensive basis for high-level research in the field of CHD with high standards of ethics, data privacy, IT management and sample logistics.

MP3-20

Predictors of Left Ventricular Hypertrophy after Correction of Aortic Coarctation

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Introduction: Left ventricular hypertrophy (LVH) is a risk factor for adverse cardiovascular events. The association between untreated aortic coarctation (Co) and LVH is known, but LVH prevalence in treated patients is not well established.
Objective: Identify predictors of LVH in patients submitted to Co correction.
Methods: Retrospective study of 100 randomly selected patients submitted to surgical or percutaneous treatment of Co, with a minimum follow-up of 6 months. Patients with LV outflow obstruction were excluded. For patients younger than 21 years, LVH was defined as a LV mass above 2 Z scores. For older patients, LVH was defined as LV mass index >95 g/m2 in

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women and 115 g/m2 in men, according to American Society of Echocardiography guidelines.
Results: 68% of patients are male. Median age at time of treatment was 4.4 years old (10 days to 75 years). 56 patients were submitted to surgical correction and 44 to percutaneous treatment (24 balloon dilation and 20 stent implantation). Mean follow up time was 11.0 ± 10.3 years (6 months to 44.8 years). 42 patients have hypertension (HT) and 26 LVH. Univariate analysis revealed a higher incidence of LVH in male gender (33.8% vs. 9.4% in females, $p = 0.013$), age at follow-up >21 years (47.8% vs. 7.4%, $p < 0.001$), age at treatment above 1 year (34.8% vs. 8.8% under 1 year, $p < 0.01$) and presence of HT (40.5% vs. 14.3% in non HT patients, $p < 0.01$). LVH was not influenced by type of treatment (26.8% surgical vs. 25.0% percutaneous, p = NS) nor presence of bicuspid aortic valve (15.8% vs. 32.3% for tricuspid, p = NS), nor residual gradient (p = NS). Multivariate analysis revealed that male gender ($p < 0{,}001$) and age >21 years at follow up ($p < 0.001$) were independent determinants of LVH.
Conclusion: There is a high incidence of patients with HT even after effectively treated Co. The incidence of LVH in our study (26%) was higher in males and older patients, and was not influenced by type of treatment nor presence of bicuspid aortic valve. These findings reinforce the need for long term follow up of effectively treated Co patients as HT and LVH are frequent late complications.

MP3-21
Myocardial infarction in Kawasaki disease patients
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Objectives: Kawasaki Disease (KD) has become the main reason of children's acquired heart diseases in the world. It is a real challenge to diagnose some patients due to the lack of specific disease markers Myocardial Infarction (MI) is the most serious complication, it's course require further investigation.
Methods: The data of 163 early stage KD patients' aged 0-17 yo examined in children's cardiology department in 1994-2009 have been analyzed.
Results: Acute MI attacked 10.4% of patients (17 out of 163) during a year from beginning of KD, including 7.4% (12) in acute stage. MI emerged in the complete and incomplete form of KD (10 and 7, $p \geq 0.2$). 29.4% (5 of 17 pts) had infarction on the 3rd week, 41.2% (7) on the 3-6 weeks, in all 70.6% (12). Acute heart failure appeared in 66.7% (5 of 12), and 33.3% (4) had cardiac arrest. There were 29.4% (5) more MI cases over the period of 6 weeks up to 1 year from the acute attack of KD. MI clinical presentation included neurological dysfunction in 88.2% (15 of 17), dyspnea and tachycardia in 68.7%, 11, bubbling rales (58.8%, 10), gallop rhythm (56.2%, 9). Left coronary artery aneurysms/ectasia took place in 70.6% (12 of 17). ECG revealed Q-Infarction in 58.8% (10), subendocardial MI in 17.6% (3), in addition 23.5% (4) had not typical MI ECG-signs but there were segmental hypokinetic wall motion and severe mitral valve regurgitation due to infarction of papillary muscles. 17.6% (3 of 17) died at an early KD stage; 64.3% (8) of 14 survivors had LV dilation, including 50% (7 of 14) with low LV ejection fraction. All MI patients had delayed KD diagnosis and immunoglobulin treatment. Mammary-coronary bypass on the beating heart was successfully performed on the 5 mo infant with an acute coronary thrombosis and refractory cardiogenic shock.
Conclusions: 7.4% of KD patients had MI at the 3-6 week, cardiac arrest occurred in 33,3% of them; 3% more had MI during the 1st year. Q-Infarction emerged in 58.8%, papillary muscles infarction in 23.5%. Mammary-coronary bypass surgery on the beating heart was performed on 5 mo infant.

MP3-22
Cardiac rehabilitation in postoperated Tetralogy of Fallot children: an useful tool?
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Introduction: exercise capacity of children with cardiac heart disease is often depressed, partly because physical inactivity.
The object of this study was to describe the effect of exercise by cardiac rehabilitation program (CRP) in postoperated Tetralogy of Fallot patients using echocardiography and ergoespirometry tests.
Methods: twelve children with total correction of Tetralogy of Fallot were chosen under precise criteria. All patients underwent ergospirometry plus rest and post-stress echocardiography.
Systolic (LVSF and RVSF) and diastolic (LVDF and RVDF) biventricular function were determined by echocardiography at rest and after exercise (during 3 first minutes of recuperation period). We measured: LVEF (Simpson's method), LVSF (M mode), TAPSE, RVFAC, E/E′, E′ and Tei index. VO2, VE/VCO2 slope and VAT were collected from ergospirometry.
After 3 months of CRP with column table and respiratory physiotherapy included, we repeated the same tests. This results were compared with those obteined previously.
We performed quality of life tests to check the psicological effects.
Results: After CRP, ergospirometry demonstrated an improvement in tolerance under maximal stress in 9:12 patients. VO2 also showed amelioration in 8:12 patients. We found a discrete increase in average values of LVSF (EF 62.5% to 65.9% and SF 32.67% to 34.67%) and RVSF (TAPSE 13.44% to 14.71% and FAC 46.28% to 51.65%) in rest conditions when pre and post CRP results where compared. After stress test biventricular systolic function improved in 3:12 patients, 1 also showed LVDF improvement, 1 presented isolated LVSF amelioration and 2 RVSF improvement. Biventricular diastolic function was reduced in 2 patients, however, 1 of them showed RV systolic amelioration. Only 1 patient had RVDF improvement and another RVSF isolated. Only 1 had no changes in cardiac function despite CRP. 3 patients exhibit worsening in diastolic function. Remaining patients presented no significant functional parameter changes.
At the end of the CRP all children showed increased selfconfidence in establishing social relations
Conclusions: cardiac rehabilitation could improve the exercise performance and quality of life in POTF. Routine use of cardiac rehabilitation may reduce the morbidity and safely improves physical conditions in POTF without mayor adverse effects.

MP3-23
Total Anomalous Pulmonary Venous Connection: Clinical Presentation and Long Term Outcome of 60 Patients at a Single Institution in 32 Years
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Background: Total anomalous pulmonary venous connection (TAPVC) is notoriously difficult to diagnose antenatally as well as posing diagnostic challenges postnatally in very sick neonates.

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We aimed to report our 32 year experience on the clinical spectrum and outcome of TAPVD in Wales.
Patients and Methods: We reviewed clinical records of 60 patients underwent TAPVC repair between 1980 and 2013. Fifty patients had isolated TAPVC (group I), and 10 had associated complex congenital heart disease (group II).
Results: The anomalous drainage was supra cardiac in 28 (46.6%), cardiac in 14 (23.3%), infracardiac in 14 (23.3%), and mixed in 4 patients (6.6%). Major associated cardiac anomalies were present in 10 patients (6 right atrial isomerism + CAVSD + DORV, 2 RAI + CAVSD, 1 DILV, and 1 HLHS). The antenatal detection rate was 11%. The median age at diagnosis was 11 days (1-270 days) in group I, and one day (1-11 days) in group II. Cyanosis (51%) and respiratory distress (35%) were the main symptoms. 19 patients with isolated TAPVC had severe pulmonary hypertension (38%) and the initial diagnosis was persistent pulmonary hypertension in 5 neonates. Severe PV obstruction was found in 42% of infracardiac type, and in 14% of supracardiac type. The median age at operation was 28.5 days (1 day-7 years). The median follow up was 11.3 years (11 months-35 years). PV obstruction developed in six patients, baffle obstruction and narrowing of SVC occurred in 4. 18 patients (36%, 18/50) with total correction developed rhythm problems including right bundle block, low atrial rhythm, junctional rhythm (one requiring pacemaker) and atrial fibrillation. Six patients in group II (60%) and 2 in group I (4%) died in the follow up. The 1-year and 5-year survival rates were 96% in group I.
Conclusion: The antenatal detection of TAPVD is unacceptably low. Early recognition of TAPVD requires careful echocardiographic examination of newborns presenting with PPHN or severe respiratory symptoms. The outcome of isolated TAPVC is favorable, however when it is associated with major cardiac anomalies it has higher mortality and morbidity. The PV drainage site *per se* was not associated with adverse outcome.

MP4-1

Catheter ablation of supraventricular tachycardias in children and adolescents – safety and effectiveness

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Accessory AV- pathways (AP) or dual AV node physiology are the most common substrates for paroxysmal supraventricular tachycardias (SVT) in children and adolescents. Definitive treatment can be achieved by catheter ablation with either radiofrequency (RF) or cryoenergy. Aim of this study was to assess effectiveness and safety of RF- and cryoenergy ablation in a pediatric population.
A total of 562 EP studies (EPS) have been performed in 479 children and adolescents with SVT during the last ten years at our institution. Indications for catheter ablation were patients' preference (67.3%), drug refractory SVT (31.5%) and malignant arrhythmias (1.2%). RF was used in 75%, cryoenergy in 16%. Both energy sources were used in 8%, no ablation was performed despite identification of a substrate in 1%. Congenital heart disease was present in 39 cases (6.9%).
APs were the substrate in 54.6% of the cases whereas AVNRT was found in 42.6%. APs and AVNRT were present in 2.8%. Overall procedural success rate was 93% independent from the substrate. APs were successfully treated in 90.5% (RF)/90% (cryoenergy). Slow pathway ablation/modulation was achieved in 98.5% (RF)/100% (cryoenergy). In 70 patients, repeated EPS due to SVT relapse or recurrent preexitation pattern was necessary. In 3 cases (0.5%) pacemaker implantation due to AV-block after RF ablation was necessary (AVNRT n = 2; AP n = 1). On routinely performed coronary angiography after ablation insignificant coronary artery narrowing was detected in 2 patients (0.3%) after RF ablation of right posteroseptal AP. No cryoenergy related coronary artery abnormalities were evident. In 1 case (0.2%), thrombosis of the right coronary artery occurred after intercoronary mapping with a 2F EP catheter and was treated by local application of tissue plasminogen activator. In 2 cases (0.2%), pericardial effusion was evident and required puncture after cryo- and RF- ablation, respectively. Vessel injuries at the puncture site with need for surgery occurred in 6 cases (1%).
Catheter ablation of SVT in children and adolescents was safe and effective. Major complications were noted only after RF-application. Therefore cryoenergy may be considered as energy source of choice for ablation in the rightseptal area.

MP4-2

The use of electroanatomic mapping for diagnosis of arrhythmogenic right ventricular cardiomyopathy

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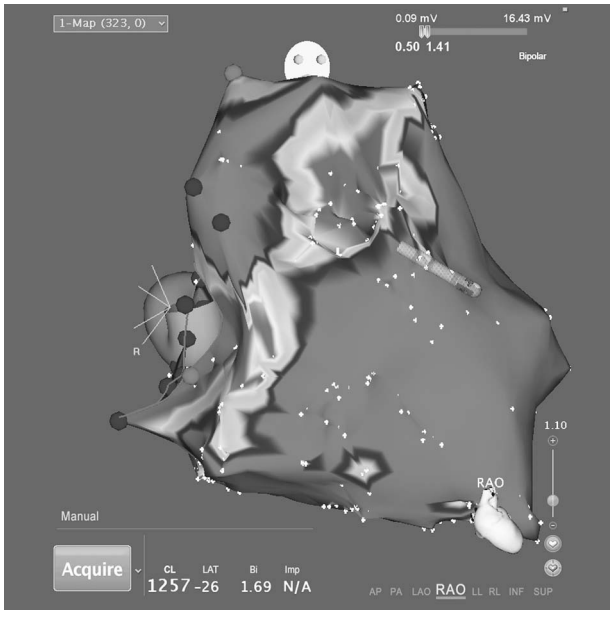
Objectives: To evaluate the results of electroanatomic voltage mapping (EAM) and targeted endomyocardial biopsy (EMB) from the low voltage areas in young pts with suspected arrhythmogenic right ventricular cardiomyopathy (ARVC).
Methods: Intracardiac electrophysiologic study with EAM (CARTO) of the right ventricle (RV) was performed in 9 consecutive pts (period 1/2012–11/2013) with possible/borderline ARVC using current non-invasive task force criteria at median age of 16.5 (range 11.5-38.0) yrs. One pat had positive family history, 7/9 pts had arrhythmia symptoms and 5/9 had documented ventricular arrhythmias. Surface ECG changes suspicious of ARVC were present in 4/9 and cardiac MRI was positive in 3/9 pts. Late ventricular potentials were present in all pts.



Results: EAM showed low voltage areas in RV inlet and/or RV outflow tract in all pts (Fig.). EMB was positive in 5/7 pts and

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programmed RV stimulation in 2/9 pts. Definite diagnosis of ARVC was thus established in 4/9 pts. An ICD was implanted in 3 pts for either primary (2 pts, first adequate therapy 2 months later in 1) or secondary (1 pat) prevention.
Conclusions: EAM with targeted EMB is highly useful for establishing final diagnosis of ARVC and may facilitate the decision on primary preventive ICD implantation in selected pts.
(Supported by MH CZ—DRO, University Hospital Motol, Prague, Czech Republic 00064203).

MP4-3
QT correction methods in children
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Introduction: accurate determination of the QTc interval in children is important especially when considering a drug's ability to prolong cardiac repolarization. Previous work suggests the most appropriate correction formula is QTc = QT/RR0.38 We set out to compute the best population derived and age related formula correction factor in our normal childhood population.
Methods: we enrolled a cohort of 1200 healthy children. In a quiet state a digital 12 lead electrocardiogram (50 mm/second) was recorded and stored. The QT and RR intervals were measured digitally in lead 2. Subjects were divided into 4 age groups; 0-1 years (n = 379); 1-5 (n = 280); 5–10 (n = 268) and >10 years (n = 273). For each age group the QT/RR curve was fitted with 2 regression analysis, a linear regression for constant α, whereby QTc = QT + α x (1-RR), and natural log-linear regression analysis for constant β whereby QTc = QT/RRβ. Furthermore, linear regression analysis of QTc/RR for the two formulae were performed (least squares method), obtaining slope and R2.
Results: Mean age: (0-1 years age group) 0.3 years, SD + 0.27; (1-5) 2.8 SD + 2.8 years; (5-10 years age group) 7.3 SD + 1.4 years; (>10 years age group) 13.3 SD + 1.2 years. From linear regression analysis correction factor was α = 0.275, β = 0.43 for 0-1 years, α = 0.26, β = 0.46 for 1-5; α = 0.19, β = 0.41 for 5–10; α = 0.18, β = 0.39 for >10 years. Linear Regression plots of QTc against RR intervals: QTc linear: slope < 0.005, R2 < 0.01 for the 4 formulae; QTc log-linear slope: < 0.001, R2 5*10-2 for the 4 formulae.
Conclusion: For the full range of pediatric subjects studied the optimum population-based correction factor α and β, decreased with increasing age. It appears that more specific correction factors, based on age and gender, are required. These are being further evaluated.

MP4-4
Clinical spectrum of frequent premature ventricular contractions in children
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Frequent premature ventricular contraction (FPVC), has been generally considered as benign, although some recent studies suggest that long-term FPVC may cause progressive ventricular dysfunction and tachycardia induced cardiomyopathy(TIC). PVC burden of 24% of total QRS/24 h had a sensitivity in separating the adult patients (pts) with impaired versus preserved LV function. TIC is a rare entity, only few studies investigated the effect of ventricular arrhythmias (VA) on the "seemingly structurally normal heart" in children.
The aim of the study was to evaluate clinical spectrum of FPVC in children with initial diagnosis of idiopathic VA (normal echocardiogram and QTc).
Material and methods: Retrospective review of complex cardiac examination from 260 pts with VA: 20pts- FPVC (50-85%QRS/24 h, mean 61,6%), 240pts-non-FPVC (0,01-49%QRS/24 h, mean 18,89%). Evaluated data pertain to clinical findings, ECG, Holter, echo, exercise test (203pts), radionuclide ejection fraction (EF)(280pts), magnetic resonance imaging MR (130pts)(focuses of fatty infiltration–FFI-31pts), endomyocardial biopsy (EMB-52pts; ARVD -12, Myocarditis-31pts), AA drugs (156pts), outcomes. Statistical analysis: $p < 0.05$ statistically significant, NS = non significant difference FPVC vs non-FPVC pts.
Results: EMB was normal in 2, suggestive of ARVC in 4 and MYO in 5 pts with FPVC(NS). FFI had 4/10 pts with FPVC who underwent MR (NS). No significant differences were found between FPVC and non-FPVC patients in age of VE diagnosis (8,5 vs 8,1 yrs;NS), follow up (6,6 vs, 6,2 yrs; NS), radionuclide left (55,45 vs 56,3%; NS) and right ventricular EF (52 vs 53,87%; NS). During follow up in pts with FPVC arrhythmia completely disappeared in 40% (NS), augmented in 15% (NS), no deaths were observed. Pts with FPVC had more frequently but NS symptoms (15 vs 9,6%), PVC-RBBB morphology (45 vs 28,1%), FFI in EMB (36,4 vs 19,5%) or MR (40 vs 22,5%). Pts with FPVC had significantly more frequently diagnosed sVT (52,6 vs 13,7%; $p < 0,0001$), more episodes of VT/24 h (6541 vs 360VT/24 h; $p < 0,0001$), longer QTd (45,7 vs 27,4 ms; $p = 0,001$), longer PVC-QRS (146 vs 129,7 ms; $p = 0,002$), higher number of AAD (4 vs 2,1; $p < 0,0001$) and, longer time treatment (4,6 vs 2,2 yrs; $p < 0,0001$).
Conclusions: 1. In analyzed group of children with FPVC, normal echocardiogram and different etiology there was no evidence of decreased radionuclide LVEF for diagnosis of "latent" asymptomatic TIC, VA seems to be benign and in 40% of patients completely disappeared during mean 6 yrs of follow-up. 2. Children with FPVC had longer QT dispersion and wider PVC-QRS duration.

MP4-5
Comparison of Cryo- vs. Radio-Frequency-Current-ablation in patients with av-nodal-reentry-tachycardia [AVNRT]
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Introduction: Cryo-ablation is a safe treatment for av-nodal-reentry-tachycardia [AVNRT] with a low risk for catheter-induced heart block, but its efficacy seems to be lower compared to radiofrequency-current [RF] ablation.
Methods: We retrospectively analyzed the efficacy and safety of cryo- vs. RF-ablation in patients with AVNRT. Targeted end point were non-inducibility of tachycardia, absence of dual av-node physiology [DAVNP], defined as an ah-jump>50 ms during atrial extra stimulus pacing, or at least no echo-beats and/or absence of sustained slow pathway conduction [SSPC].
Results: Between 2003 and 2013 n = 98 patients with AVNRT were treated either with RF- (n = 62) or cryo-ablation (n = 36). Mean age was 12.6 ± 3.4 years in the cryo- and 12.5 ± 3.6 years in the RF group, mean follow up 4.9 ± 1.5 and 5.5 ± 3.5 years, respectively. Cryo-ablation was effective in 32/36 patients (89%) and in all patients with RF-energy. The four patients which could not be

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treated by cryo-ablation were effectively treated by RF-energy during the same procedure, accordingly n = 66 patients were treated with RF-energy.
Two of 62 patients (3.2%) in the RF group and one out of four patients in whom we switched to RF got complete heart block. Thus the risk for heart block is 3/66 (4.5%) for all RF-ablations whilst none of the patients treated with cryo-energy developed heart-block.
Seven of the 32 patients (22%) after cryo-ablation and 2/66 patients (3%) in the RF-group had recurrence of tachycardia, despite the targeted endpoint was reached after the first ablation. In the cryo-group were two successfully treated by cryo- and three by RF-ablation. Two got reappearance after another cryo-ablation and were finally treated by RF-energy. The two recurrences in the RF-group were finally treated by a second RF-ablation.
All together 9/32 patients (28%) could not be treated by cryo-ablation and finally had to be treated with RF-energy.
Conclusions: For the ablation of AVNRT cryo-energy is very safe, but has a lower efficacy and a high recurrence rate. In contrast RF ablation is highly effective but associated with a risk for permanent heart block. Switching from cryo- to RF-energy is probably associated with a higher risk for completed heart block.

MP4-6
Development of a Whole Heart Model in Patients with Tetralogy of Fallot
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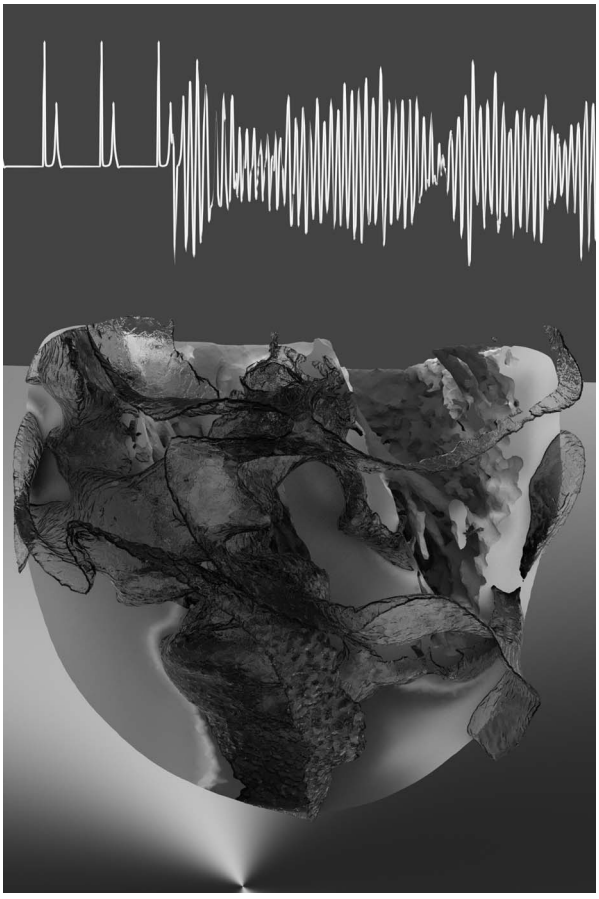
Introduction: The development of heart models so far has been limited significantly by the complexity of the electrical and mechanical mechanisms in the heart. Computing times were inacceptable leading to oversimplification of the used models. Using a "blue gene/Q IO" environment we are in the process of developing an computational whole-heart model for patients with Tetralogy of Fallot (ToF). Those patients have scars or patches within the ventricular tissue, able to create abnormalities in electrical conduction and mechanical contraction. This can cause ventricular arrhythmia and/or unexpected sudden death. Creating an individual model based on patient imaging data might help to understand arrhythmogenisis and risk factors for the individual patient.
Methods: Patients who had Magnetic Resonance Imaging (MRI) of their heart performed for clinical diagnostic questions were selected and de-identified. Using "Cardioid" a highly scalable electrophysiology code developed at IBM and Lawrence Livermore National Laboratory individual whole heart models are created after segmenting using black blood sequences. These models are tested for their behaviour under normal sinus rhythm and compared with the expected behaviour in a feasibility study.
Results: We were able to create whole heart models of the selected patients reflecting their individual electric and mechanical properties. So far the models come very close to the expected behaviour although the information about tissue qualities derived by MRI so far are somehow limited.
Conclusions: The preliminary results are encouraging that with improved MRI resolution, additional information from late enhancement and further increase in computational power we might be able to create a risk matrix for quantification of arrhythmia risk and sudden death.
This research was supported by a Victorian Life Sciences Computation Initiative (VLSCI) grant number VR1475 on its Peak Computing Facility at the University of Melbourne, an initiative of the Victorian Government, Australia.



MP4-7
Do sick children with severe acute malnutrition in Kenya die of arrhythmia?
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Background: Annually 2.1 million children die as a consequence of severe acute malnutrition (SAM). Fatal arrhythmias due to electrolyte perturbations and fluid shifts during refeeding have been postulated as a possible cause, although the data supporting this is limited. We investigated the role of arrhythmias among children with SAM in Kenya.
Methods: As part of the CArdiac Physiology in MALnutrition (CAPMAL) Study, children with SAM (marasmus or kwashiorkor) were matched by age and gender with equally sick non-malnourished controls. All presented to a rural Kenyan hospital between March and November 2011. Serial 12-lead ECGs and electrolytes at days 0, 7 and 28, and also 7 day continuous Holter monitoring were recorded.
Results: 88 SAM cases and 22 controls were well matched for age, gender and clinical presentation, with similar baseline characteristics apart from anthropometry (weight for height z-score -3.2 in

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cases vs -1.1 in controls) and HIV status (22.7% vs 0.0%) (p all <0.05).
Mean heart rate was lower among SAM cases than controls at admission (132 vs. 142/min; $p = 0.0412$), this difference disappeared by day 7. Corrected QT interval (QTc) was short (<2nd centile for age) in 27.3% cases and 4.6% controls ($p = 0.023$). This difference persistent until day 7 but was not associated with death (OR 0.52 $p = 0.426$). Holter data showed 31 episodes of ventricular tachycardia (VT) in 7 cases and 1 control and 13 episodes of bradycardia in 3 cases. Fifteen episodes of VT (48.4%) occurred in a child who died 9 days after admission with disseminated tuberculosis, 2 occurred in a patient who died of sepsis. All bradycardias and other VTs remained asymptomatic and were not associated with poor outcome. A difference in median potassium levels at admission (3.2 vs 4.2 mmol/L; $p = 0.0018$) and median magnesium levels at day 7 (0.91 vs. 0.97 mmol/L, $p = 0.0029$) in SAM cases vs controls was observed. No difference in electrolyte levels at any other stage was apparent and these were not affected by the presence of HIV.
Conclusions: We found many episodes of VT in children with SAM, but these did not appear to lead to unexpected death during the re-feeding period.

MP4-8
Prolonged QTc in atrial septal defect – an example of mechano-electrical feedback due to right ventricular volume overload?
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Objectives: The aim of our study was to assess the effect of long-term right ventricular (RV) volume overload on repolarization. The reason was a frequent observation of prolonged QTc intervals in children prior to atrial septal defect (ASD) closure.
Methods: QRS, QTc and JTc intervals were measured manually from leads II and V5 of standard ECGs in 45 children with an isolated ASD a day before and at least 6 months after the ASD closure. Each QTc was compared to gender and age matched normal values. RV dimensions were obtained from echocardiographic M-mode images.
Results: The prevalence of prolonged QTc decreased from 22.2 to 2.2% after surgery ($p = 0{,}007$). Individual QTc intervals shortened significantly (Table) whereas QRS duration did not change. There was a highly significant correlation between the change in QTc and JTc intervals ($R = 0.741$, $p < 0.001$). RV dimensions did not correlate with QTc values. Inter- and intra-observer coefficient of variation for QTc measurements was 2.53 and 1.45%, resp.

QTc ms	Prior to surgery	After surgery	P value
Lead II mean (SD)	418 (25)	380 (25)	<0.001
Lead V5 mean (SD)	412 (26)	378 (25)	<0.001

Conclusions: Prolonged QTc intervals occur frequently in children with hemodynamically relevant ASD and may not reflect an inborn channelopathy but rather a reversible effect of myocyte stretch on ion channel function. Such mechano-electrical feedback might participate in arrhythmogenesis in patients with more complex heart defects and significant residual RV volume overload (Supported by MHCZ-DRO, University Hospital Motol, Prague, Czech Republic 00064203).

MP4-10
Myocardial Perfusion Single Photon Emission Computed Tomography (SPECT) in children with ventricular arrhythmias
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The aim of the study was to evaluate left ventricle (LV) myocardial perfusion defects (PD) in pediatric patients (pts) with ventricular arrhythmias (VA).
Material and methods: Retrospective review of data from 96 pts with: VA (VE- ventricular ectopic beats >2000/24 hour, 50pts with ventricular tachycardia –VT, 57% VA with right ventricular outflow tract morphology), normal QTc, normal echocardiogram (initial diagnosis "idiopathic VA"). Radionuclide LVEF (ejection fraction at rest) and qualitative 99mTcMIBI SPECT (rest, exercise-Bruce protocol) were performed in all pts (100%). Mean age at VA diagnosis 9,6 yrs, at SPECT 14,2 yrs. PD had 69pts (72%): transient (TDP during exercise)-55pts, fixed (FPD during rest and exercise)-14pts. Diffuse PD (DPD in whole area of the analyzed LV wall)-30/69pts. Magnetic resonance imaging (MR) of the heart had 94pts (lipogenesis –20, PD-16/20pts), endomyocardial biopsy (EMB) 40pts: myocarditis-25 (PD 19/25), ARVC-8(PD 7/8)pts. Statistical analysis: $p < 0.05$ considered to be statistically significant.
Results: PD had not significant correlation in pts with/without abnormalities in EMB or MR. PD had more often pts with older age at VA diagnosis (10 vs 8 yrs, $p = 0{,}03$). Pts with normal EF/SPECT/MRI had lowest mean age during SPECT (11 yrs, $p = 0{,}001$). Symptomatic VA had pts with PD (67 vs 41%, $p = 0.21$), negative LV-T waves in ECG pts with DPD (60 vs 29%, $p = 0.06$), polymorphic VA (50 vs 16%, $p = 0{,}07$) or higher VT rates (205 vs 158/min, $p < 0{,}05$) pts with FPD. PD in anterior (67 vs 31%, $p < 0.05$) or posterior (63 vs 16%, $p < 0{,}05$) LV segment had pts with DPD. Group with both normal EF/SPECT was younger (7,2 vs 12,8 yrs, $p = 0{,}01$), had shortest mean duration of VE-QRS (127 ms, $p = 0.03$). Pts with PD had lower mean LVEF (53 vs 56%, $p = 0{,}05$), especially with FPD (48 vs 54%, $p = 0{,}04$) and children with both abnormal EF/SPECT had highest daily frequency VE/24h ($x = 18\%$QRS/24 h, $p = 0{,}08$)—suspected subclinical "VA induced cardiomyopathy".
Conclusions: 1. Perfusion defects in 99 mTc MIBI SPECT were found in LV walls in 72% of children with ventricular arrhythmia and normal echocardiogram but presence of PD did not correlate with abnormalities in EMB or MR. 2. PD were more often found in children with lower mean rest radionuclide LVEF, polymorphic morphology of VA, frequent VE in Holter-ECG, negative LV-T waves in ECG and older age at VA diagnosis.

MP4-11
Comparison of a simplified, minimally invasive, non-fluoroscopic approach for catheter ablation of supraventricular arrhythmias in children and adults
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Introduction: Although the "near zero-X-ray" or "no-X-ray" catheter ablation (CA) approach has been reported for treatment of various arrhythmias, few prospective studies have strictly used "no-X-ray" simplified 2-catheter approaches for CA in patients with supraventricular tachycardia (SVT). We compared the feasibility of a minimally invasive, non-fluoroscopic (MINI) CA approach in adult and pediatric such patients.
Methods: Data were obtained from a prospective multicenter CA registry of patients with regular SVTs. After femoral access, 2 catheters were used to create simple, three-dimensional electroanatomic maps (Ensite Velocity NavX, St Jude Medical, St. Paul, MN, USA) and to perform electrophysiologic studies. The medical staff did not use lead aprons after the first 10 MINI CA cases.
Results: The first 188 patients (age, 45 +/- 21 years; 35 pts <19 years; 55% women) referred for the no-X-ray approach were included. They were compared to 714 consecutive patients referred for a simplified approach using X-rays (age, 52 +/- 18 years; 50 pts <19 years; 55% women). In pediatric population (n = 35, age, 14 +/- 4) the procedure time (64 +/- 20 vs. 63 +/- 29 min, p = NS), incidence of major complications (0% vs 0%, p = NS) and complete non-fluoroscopic imaging in MINI CA approach (94% vs 95%, p = NS) were similar as compared to adult population. In patients with age <19 an acute (98% vs. 98%, P = NS) success rate were similar in MINI CA approach and X-ray approach (n = 50, age 15 +/- 3), however significant increase in recurrences were reported in pediatric population (15% vs 5%, p < 0.01, in both MINI CA and X-ray approach, within a mean 10-month follow-up). The implementation of MINI CA approach in pediatric population with SVT resulted in very significant reduction of mean X-ray exposure time from 11.1 +/- 9.4 to 0.1 +/- 0.7 min (p < 0.001).
Conclusions: The implementation of a strict "no-X-ray, simplified 2-catheter" CA approach is safe and effective in the majority of children and adults with SVT. No-fluoroscopic approach should become "the golden standard" in training of next generation of electrophysiologists, especially in pediatric population that is prone to higher incidence of recurrences and redo procedures.

MP4-12

Transseptal Perforation for Radiofrequency Catheter Ablation in Patients with Fontan Procedure

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Introduction: Atrial tachyarrhythmia is a common late complication and a major cause of late morbidity after Fontan surgery. The reentrant tachycardia may be localized to the pulmonary venous atrium, transseptal perforation should be needed for mapping and radiofrequency catheter ablation (RFCA).
Methods: A total of 12 patients had attempted transseptal perforation. Median age was 22 years. One patient had undergone atrio-pulmpnary conection procedure, eleven had undergone total-cavo pulmonary connection. We chose intracardiac echocardiography in 11 cases, and trans-esophargeal echocardiography in one case, using Brockenbrough needle for transseptal puncture. The procedure of balloon dilatation was needed in two cases.
Results: The indication for pulmonary venous access was RFCA of atrial flutter and atrial tachycardia, and atrio-ventricular reentrant tachycardia. There were two clinically significant complications. The needle tip passed into the ascending aorta in one case following L-R shunt from aorta to right atrium. We performed coil embolization for the shunt. Transmuscular perforation of left ventricle in another case had no L-R shunt. Both had no hemodynamically insignificant event. In ten patients after successful puncture, we had successful RFCA in seven cases (70%).
Conclusion: Transseptal perforation can be a challenging method of obtaining pulmonary venous atrial access for electrophysiologic procedures in patients with Fontan procedure. Acute success rate of RFCA was acceptable, and then RFCA can be an effective option to treat refractory tachycardia following Fontan surgery.

MP4-13

Genetic diagnosis of early-onset cardiomyopathies using next-generation sequencing technologies

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Introduction: Cardiomyopathies (CMPs) are a heterogeneous group of diseases with several etiologies ranging from heart specific muscle diseases to multiorgan syndromes. Early-onset CMPs, with symptoms emerging during prenatal life, infancy or early childhood, are typically severe and progressive, with poor prognosis. The majority of these patients still have had no molecular diagnosis. Developing the understanding of genetic causes and clinical features of these early-onset CMPs has become possible through novel next-generation sequencing (NGS) techniques. We undertook a detailed molecular study of 57 Finnish infantile CMPs, from the sole national centre in Finland in charge of cardiac transplantation. The aim of our study is to investigate the success of NGS in identification of disease causing mutations in a naive material, and to characterize the CMP molecular background in Finland.
Materials and methods: Our patient material is composed of 19 cases of hypertrophic cardiomyopathy, 28 cases of dilated cardiomyopathy, 8 cases of left ventricular noncompaction and 2 patients with restrictive cardiomyopathy. Nine patient DNA samples were analyzed by whole-exome sequencing (WES) and 48 with targeted sequencing of a custom-made panel of 117 cardiac genes using a HaloPlex custom kit. We developed a bioinformatic pipeline to analyze the NGS data. SNVs, indels, splice sites and copy number variants were evaluated for their potential pathogenicity. The candidate DNA variants were validated by analyzing the segregation pattern in the family and confirming their absence in Finnish controls.
Results: From the samples investigated with WES, mutations were confirmed in four patients, and from targeted sequencing, strong candidates for disease-causing mutations were identified in 14 cases. In depth analysis to identify further mutations, and stringent verification of pathogenic role of the identified variants is ongoing.
Conclusions: In the current stage of the study, WES has led to a success rate of approximately 45% in the identification of early-onset CMP-causing mutations, while for targeted sequencing a success rate of 30% is anticipated. While WES offers the potential to identify mutations in novel CMP genes, targeted sequencing offers a better coverage in known cardiomyopathy genes, as required for routine diagnosis.

MP4-14

Influence of aortic valvuloplasty by tricuspidalization and leaflet extensions on proximal aorta morphology in bicuspid aortic valve

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Introduction: Types of bicuspid aortic valve (BAV) morphology influences the development of aortic valve stenosis/regurgitation as well as aortic root/ascending aorta dimensions. The aim of the study was to evaluate changes on proximal aorta morphology (annulus, root and ascending aorta) in patients with congenital valve disease after surgical valvuloplasty by tricuspidalization and leaflet extensions.
Methods: In 2005-2013, 66 patients with congenital valvular disease underwent aortic valve repair at our institution. For our study we selected 33 patients (M/F 27/6 patients) with BAV and severe aortic stenosis (with maximum mild-to-moderate aortic regurgitation); in 17 patients (51.5%) was the surgery performed due to combined aortic disease, 16 patients (48.5%) had isolated aortic stenosis.
Mean age at operation was 14.4 ± 6.3 years. Nine patients (27.3%) had previous intervention (6.66 ± 5.6 years prior to valvuloplasty).
In a prospective mid-term FU, with median 4.6 years (1.1 months–7.3 years), repeated echocardiographic measurements were performed. Differences in proximal aorta diameters (annulus, root, ascending aorta) converted to Z- scores were statistically evaluated.
Results: Morphology of BAV was in 28 patients (84.8%) with right-noncoronary cusp (R-N) raphe and in 5 (15.2%) with left-right coronary cusp (L-R) raphe configuration. No noncoronary-left cusp (N-L) raphe configuration was present.
Preoperative measurements: Aortic valve annulus was dilated (Z-score>2) in 9 patients (27.3%), aortic root was not dilated in all (but below Z-score<2 in 5 patients (15.2%)), ascending aorta was dilated (Z-score>2) in 24 patients (72.7%).
Postoperative measurements: Aortic valve annulus Z-score grew proportionally in all patients after surgery (mean 1.26 ± 1.03 to 0.91 ± 1.99, $p = 0.21$); ascending aorta diameter Z-score decreased significantly (mean 2.46 ± 1.92 to 1.16 ± 2.27, $p = 0.0054$); and aortic root diameter Z-score increased slightly, reaching normal values (mean -1.12 ± 0.92 to -0.13 ± 1.59, $p = 0.0050$).
Conclusions: BAV analysis in our patients with aortic stenosis showed dominant occurrence of R-N valve configuration (with minor L-R and no L-N configuration); and with this setting predominantly ascending aorta dilatation. In patients after aortic valvuloplasty by tricuspidalization, who remain with stable valve function, we found normalization of ascending aorta dimensions as well as normal growth of aortic root. This study may contribute to the discussion of the impact of hemodynamics on aortic morphology.

MP4-15

Recovery of long-term volume overloaded right ventricle after pulmonary valve implantation in patients with ToF repair

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Introduction: Right ventricular (RV) volume overload due to pulmonary regurgitation after tetralogy of Fallot (ToF) repair is the most frequent and expected finding. Although well tolerated for years, should be treated before RV compensatory mechanisms are exhausted. Magnetic resonance imaging (MRI) is gold standard for establishing exact RV end-diastolic/end-systolic volumes (EDV/ESV). RV > 170 ml/m^2 is considered of not capable to recover but the cut-off point for re-operation is still not definitively established.
Methods: Analyzed were 26 patients (17M/9F) after previous ToF correction with isolated severe RV volume overload who underwent pulmonary homograft (or bioprosthesis) implantation at our institution (median age 21.5 y., median period after ToF repair 17.9 y.).
RV was evaluated pre-operatively and during follow-up (median 2.2 y.) by echocardiography (ECHO) and MRI. Compared were patients with MRI RV EDV > 170 ml/m^2 (group A) and RV EDV < 170 ml/m^2 (group B).
Results: Early conduit dysfunction with severe pulmonary regurgitation was present in 2 patients; in all others RV improvement was experienced.
MRI findings: 1.Post-surgical decrease of RV volumes (median, in ml/m^2): EDV 172 → 119 ($P < 0.0001$), ESV 100.5 → 61.5 ($P = 0.0003$); with significant RV EDV difference in groups A/B in absolute values (123.5/104 ml/m^2, $P = 0.04$) but not in % RV size decrease (37.5/34.5%). No significant ESV difference between groups A/B was found (62.5/52 ml/m^2; decrease 37.5/28.5%). 2.No significant overall EF change after surgery (median RVEF 40 → 47%).
ECHO findings: All measured diameters with significant improvement after surgery, though w/o differences between groups A/B. After surgery: 1.most considerable improvement in RV long-axis diameter (median 38 → 27.5 mm, $P < 0.0001$), RV dilatation (>33 mm) decreasing from 90 → 15.4% of patients ($P < 0.0001$); 2.significant change of RV dilatation according to 4-chamber ratio RV/LV>1 from 80.8 → 4.2% of patients ($P < 0.0001$); 3.no change in RV systolic function after surgery.
Conclusions: Our study showed that RV > 170 ml/m^2 must not represent the upper limit for re-do. After successful elimination of volume overload RV was capable of significant improvement regardless its pre-operative size. Though, the degree of dilatation was important, as RV proved ability to decrease only by 30-40%; and this was similar in more/less dilated RVs. Very severely dilated RV is probably not able to regress to normal values. ECHO was as useful as MRI for serial pre-/post-surgical RV assessment.

MP4-16

Clinical, neurohormonal and psychological predictors of survival in patients with congenital heart disease

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Introduction: The growing cohort of patients with congenital heart disease (CHD) has to face problems regarding its medical condition per se, but also psychosocial challenges and concerns.

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Aim of this study is to assess the value of established depression scores Beck Depression Inventory (BDI) and Zung self-rating depression scale (Zung SDS), along with B-type natriuretic peptide and exercise parameters in predicting adverse clinical events.
Methods: Sixty ambulatory patients with various forms of CHD, mean age 28.911.4 years old, 53% male, were recruited from a tertiary center. Patients' neurohormonal and psychological status and exercise capacity were assessed through plasma B-type brain natriuretic peptide (BNP) measurement, Beck depression inventory and Zung depression scale questionnaires and cardiopulmonary exercise test. Patients were followed for 5.11.1 years for major cardiovascular events (MACE), including death or hospitalization.
Results: Scores on Zung scale ranged from 20 to 63 and on BDI from 0 to 36. Seventeen patients (28.3%) had positive both scores and were therefore characterized as having depression. Patients with depressive symptoms had diminished exercise capacity, as expressed with peak VO2 ($p = 0.019$) and VE/VCO2 ($p = 0.028$), and higher levels of BNP ($p = 0.03$), compared to non-depressed patients (Table). During the follow-up period 22 patients (36.6%) experienced a MACE. Among them, eleven (50%) patients were depressed. The univariate Cox proportional hazard ratio analysis revealed that all parameters examined (VO2 peak, VE/VCO2, BNP, depression) were significant predictors of MACE. Depressed patients had 2.428 times higher ratio of MACE or death, compared to non-depressed ones (95% CI: 1.630 to 3.616, $p < 0.05$).
Conclusions: Patients with CHD with depressive symptoms have impaired physical activity, associated with excessive neurohormonal activation. BNP levels, cardiopulmonary exercise parameters and the presence of depression strongly predicted MACE. The identification of prognostic parameters maybe helpful in managing patients with CHD. Table: Differences in clinical and functional variables between patients with depressive and those without depressive symptoms.

Variables	Depressed (n = 17)	Non-depressed (n = 43)	p-value
Age (years)	22.710.2	28.311.6	0.298
Gender: Male/Female(%)	8/9 (13/15)	24/19 (40/32)	n.a.
Peak VO2 (ml/Kg/min)	13.83.2	24.29.3	0.019*
VE/VCO2	58.511.7	3514.4	0.028*
BNP (pg/ml)	332.22157.5	207.9225.6	0.030*

*significant at level $p < 0.05$

MP4-17
Percentiles for central blood pressure in childhood
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Objectives: Aim of this study was to generate percentiles for central systolic blood pressure noninvasively in children and young adults living in Germany with an oscillometric device.
Methods: 2206 children, adolescents and young adults living in Germany (mean: 13.2 years; range 8.0-21.9 years; 49.1% female) were recruited from different school and community types. Peripheral (pSBP) and central systolic blood pressure (cSBP) were obtained noninvasively by an automatic oscillometric device (Mobil-O-Graph, I.E.M., Germany) after 5 minutes rest in supine position. Subjects with hypertensive BP values (systolic/diastolic blood pressure >95th percentile) and/or obesity (BMI > 97th percentile) were not included into the reference population (n = 761). Percentiles are calculated using LMS-chartmaker-pro to fit smooth centile curves. With this method the changing distribution according to some covariate is presented as median (M), coefficient of variation (S) and skewness (L). Reference centiles for cSBP with regard to age and body height are presented for girls (f) and boys (m).
Results: cSBP was positively correlated with age ($r = .457$), height ($r = .483$) and pSBP ($r = .798$); (all $p < .001$). For the reference percentiles 1445 (49% female) subjects fulfilled the inclusion criteria. Only cSBP values of normotensive, non-obese participants were used to calculate the reference percentiles. cSBP was higher in boys from 14 years onwards and ranges were wide for both sexes (boys:74-136 [mmHg]; girls: 75-123 [mmHg]). The reference percentiles for cSBP differ slightly according to sex. The curves for boys show a steeper increase until the age of 14 compared to girls. The reference percentiles for girls are smoother all over the age range.
Conclusions: Centiles for cSBP can be helpful in the difficult decision-making process for treating (pre)hypertension in youth. Practical implications for clinical use of these reference percentiles have to be clarified in following studies.

MP4-18
Tipping the Scales – Distribution of Body Mass Index in Adults with Congenital Heart Disease
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Introduction: Obesity is one of the world's biggest public health concerns. Weight control is important to preserve healthy living and avoiding secondary co-morbid conditions due to excess weight gain. In patients with adult congenital heart disease (ACHD) excess weight may be associated with worsening cardiac function.
We assessed the distribution of body mass index (BMI) of patients with ACHD attending the Yorkshire ACHD clinic and their associated cardiac pathology.
Methods: Retrospective database analysis of patients who underwent transthoracic echocardiography and who attended our ACHD clinic in 2008. All patients ⩾16 years with structural heart disease were included. Severity of ACHD disease was graded according to the 2008 AHA/ACC guideline criteria. BMI ranges were categorised as ⩽19 = underweight; 20-24 = normal; 25-29 = overweight; ⩾30 = obese. As a further analysis we will show the temporal changes in BMI between 2008 and 2013.
Results: 475 patients (men = 237) with ACHD were analysed. A similar number of men and women comprised the group. The distribution of severity of ACHD was similar between sexes.
Approximately half the cohort had a BMI ⩾ 25 with 16% of men & 22% of women being obese. Six percent (n = 29) of the cohort were morbidly obese (BMI ⩾ 35). Almost 60% of patients (n = 171) aged 31-50 years had a BMI which exceeded 25.
Over three quarters of patients had moderate or severe ACHD. Of the patients with severe ACHD a smaller proportion were overweight or obese but more were underweight compared to patients with mild/moderate severity ACHD. In a binary logistic

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model which included age, sex and ACHD severity, each 5 year increase in age was associated with a 20% relative increase in risk of reaching a BMI ⩾ 25.
Conclusions: There are epidemic numbers of patients in our ACHD population who are overweight or obese. Increasing age is associated with higher BMI. New strategies are required to help patients maintain a healthy weight.

MP4-19
Is there a link between ferritin and the cardiac function in anorexic adolescents ?
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Introduction: Very little is carried out on iron and hematological status in AN patients. Normal serum iron and elevated serum ferritin concentration were the most revelant results seen in few studies based on small cohorts of AN patients. In cardiac literature an overload of iron stores raises the risk of ischemic heart disease. Because concentrations of serum ferritin are directly proportional to intracellular ferritin concentrations, it is considered to be the best clinical measure of body iron stores and we used this parameter in our study. Iron overload has been found to increase vascular oxidative stress and accelerate arterial thrombosis by promotion of oxidized low-density lipoprotein and direct endothelial injury.
Aim: The aim of this study was to compare in the acute state of undernutrition, biochemical, endocrine and cardiac parameters between female anorexic adolescents with a high ferritin and a normal ferritin.
Method: The study has a prospective design as all patients with anorexia nervosa (AN) admitted to our Eating Disorder Unit from 2002 till 2012 were scheduled to undergo a full clinical examination, biochemical and endocrine bilan together with a cardiac examination (Echocardiographic evaluation + 12 lead ECG). In total 311 adolescent girls (9.8-17.87 years) were referred during the current study and all fulfilled the DMS-IV (Diagnostic and Statistical Manual of Mental disorders, 4th edition DSM-IV) criteria ascertained by a child psychiatrist and a paediatric nutritionist. None of the patients had a family history of cardiac diseases or of any systemic disease involving the cardiovascular system. Laboratory and clinical parameters: Blood samples were drawn for: BUN, creatinine, electrolytes, cholesterol, thyroid hormones, insulin growth factor-1, iron, ferritin, zinc, hemoglobin, hematocrite, leukocytes, fibrinogen and liver tests. Body mass index (kg/m^2) was calculated and curves and centiles were used as reported by Rolland-Cachera et al. (1991).
Results: In total we identified a high ferritin level >137 ng/ml in 82 of the 311 in the anorexic girls (26,4%). Risk factors for developing a high ferritin are: BMI: $14.41 \pm 1.59\,kg/m^2$; HR: 56.01 ± 16.36 bpm; LVM/height $^{2.7}$: $23.99 \pm 5.25\,g/m^{2.7}$; cholesterol: 192.65 ± 50.70 mg/dl; AST: 31.58 ± 12.11 U/L; ALT: 43.93 ± 47.67 U/L, IGF-1: 113.16 ± 74.97 ng/ml and FT3: 3.08 ± 1.80 pmol/L. The euthyroid sick syndrome was found in 33% of the patients with high ferritin versus 18.3% in the normal ferritin group.
Conclusion: A higher ferritin level is correlated with a higher cholesterol level and lower FT3 level. Both parameters are cardiovascular risk factors for atherosclerosis. Further research is needed.

MP4-20
Screening for cardiac risk in children and young adolescent athletes-data from outpatient sports clinic
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Introduction: In an attempt to improve the identification of young athletes at potential risk of sudden cardiac death (SCD), screening protocols are established. There are two main approaches to pre-participation cardiovascular screening of the athletes: those of American Heart Association (AHA), and the other one of European Society of Cardiology (ESC) and the International Olympic Committee (IOC). The AHA recommendations consist of 12-key elements of personal and family medical history, and physical examination. The ESC/IOC recommends using a standard 12-lead ECG in addition to a focused medical history and physical examination.
In this study, implementation and results of a standardized pre-participation cardiovascular screening in young athletes, according to relevant guidelines, are described.
Methods: In 2011, 1240 children and young adolescent athletes aged 5-17 years underwent cardiac evaluation as part of a pre-participation screening program. The majority were male and participated in football, team handball and martial arts. Level of training was different. They were investigated in outpatient sports clinic in Samobor, Zagreb County, Croatia, by physician, licensed specialist in sport medicine. Cardiac evaluation consisted of a health questionnaire (relevant personal and family medical history), physical examination, and 12-lead ECG-interpreted in accordance with the 2010 ESC recommendations.
Results: Of examined athletes, 49 (4%) were reffered for further cardiovascular evaluation due to symptoms (6 athletes; 12.2%), detection of a cardiac murmur or other abnormalities in physical examination (16 athletes; 32.7%), an abnormal ECG (25 athletes; 51%), and a presence of a family history of inherited cardiac disorder or premature (⩽50 years old) SCD (2 athletes; 4.1%). The most frequent ECG changes included first degree AV block, complete RBBB and left-axis deviation. After attending specialist cardiac clinic, 27 (55%) athletes showed normal findings, 15 (31%) need further follow-up, and for 7 (14%) there were no feedback data.
Conclusions: In a study population of young athletes undergoing pre-participation screening, the prevalence of abnormal ECG findings, suggestive for underlying cardiac disease and mandating additional testing, is low (2% of the overall population). Therefore, under existing financial circumstances, a correct interpretation of an ECG should not represent additional burden in prevention and saving lives.

P-1
Effects of methylphenidate treatment on the heart rate variability in patients with attention deficit and hyperactivity disorder
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Introduction: Methylphenidate (MPH) is a psychostimulant drug and commonly used for attention deficit and hyperactivity disorder (ADHD). It has been reported that the sympathomimetic effects of methylphenidate may cause disturbances in cardiac rhythm.

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Methods: Heart rate variability (HRV) analyses were performed to all patients before and at the end of first month of MPH treatment by 24-hr rhythm Holter. Control group consisted of age and gender matched healthy children. Heart rates, maximal QT and QTc durations were analyzed. The time-domain analysis included average heart rate, standard deviation of all normal sinus RR intervals over 24 h (SDNN), standard deviation of average NN intervals (SDANN), SDNN-index, percentage of successive normal sinus RR intervals >50 ms (pNN50) and root-mean-square of the successive normal sinus RR interval difference (rMSSD). The following frequency domain analysis indices were obtained: total power (TP), low frequency (LF) power, high frequency (HF) power, and LF/HF ratio.
Results: A total of 33 patients (24 boys, 9 girls) and 36 healthy control subjects (21 boys, 15 girls) were enrolled in this study. The mean age of the patients and controls were 9.7 ± 2.6 years and 9.5 ± 2.8 years, respectively ($p > 0.05$). The mean average heart rate before and after MPH treatment was 93.9 ± 8.5 and 95.9 ± 9.3, respectively. While the minimum, maximum and average heart rate were similar between two groups before MPH treatment ($p > 0.05$) only maximal QTc duration was significantly increased under MPH treatment in the ADHD patients (mean 452.8 ± 19.1 ms and 442.9 ± 14 ms, respectively; $p = 0.02$) (Table). The comparison of pre- and post-treatment 24-h HRV analyses with the values of control group demonstrated no significant differences both in all time and frequency domain parameters ($p > 0.05$). Also, no significant arrhythmia was detected in the Holter recordings.
Conclusions: ADHD seems not to have a significant effect on HRV in children. Also, in contrast to majority of the previous studies, MPH treatment did not affect the HRV significantly in our study. However, increase in maximal QTc duration was statistically significant after MPH treatment.

	Controls (n = 36)	Pre-treatment (n = 33)	P value	After treatment (n = 33)	P value
Average HR (beats/min)	90 ± 11.7	89.3 ± 8.6	>0.05	91.2 ± 9.4	>0.05
Max. QT (ms)	428.5 ± 13.2	428.2 ± 15	>0.05	430.5 ± 16.2	>0.05
Max. QTc (ms)	442.9 ± 14	449.6 ± 13.8	>0.05	452.8 ± 19.1	0.02
SDNN	131.5 ± 43.1	137.1 ± 33.5	>0.05	133.7 ± 36.5	>0.05
SDANN	112.4 ± 41.4	121.7 ± 32.7	>0.05	120.2 ± 38.6	>0.05
SDNNi	64.5 ± 19.7	65.8 ± 18.1	>0.05	63.2 ± 18	>0.05
RMSSD	44 ± 16.1	46.3 ± 18.2	>0.05	45.3 ± 18.5	>0.05
PNN50	18.4 ± 10.7	19.1 ± 10.3	>0.05	19.2 ± 12	>0.05
TP	4296 ± 2601	4401 ± 2178.8	>0.05	4014 ± 1983	>0.05
LF	892.8 ± 464.4	993.2 ± 509.8	>0.05	875.1 ± 433.6	>0.05
HF	568.1 ± 293.3	657.1 ± 384.1	>0.05	619.6 ± 367.1	>0.05
LF/HF	1.7 ± 0.5	1.9 ± 1.1	>0.05	1.8 ± 1.1	>0.05

P-2
Sotalol and sinus arrest; an unreported side effect in children resulting in presyncope

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Introduction: Sotalol, a class III antiarrhythmic drug with nonselective beta- blocker properties, is widely used in treatment of supraventricular and ventricular tachycardias in children. Its side effects are mostly related to prolongation in QT interval. Sinus arrest during sotalol treatment in standard doses had not previously been reported. In this paper we report an adolescent who developed presyncope due to sinus arrest during sotalol treatment.

Case report: A 14 years old girl was admitted with the complaints of chest pain and palpitation. She was taking sotatol (2×80 mg) for the frequent ventricular premature contraction (VPC) for the last one month. Due to the increase in frequency of palpitations she was admitted to our clinic. Medical history was normal; nevertheless family history revealed that his 17- year-old brother was taking antiepileptic drug for seizures. Physical examination was normal, blood pressure was 110/70 mmHg, heart rate was 61 bpm and respiratory rate was 16/minute. Serum electrolyte levels, complete blood count, renal and thyroid functions and transaminase levels were normal. Echocardiographic evaluation was normal. Surface ECG was normal and QTc was measured as 0.40 ms. Holter monitorization was performed. During monitorization, she developed presyncope with sweating while sitting. Electrocardiographic recordings during presyncope showed 5.4-minute sinus arrest (Fig. 1). Additional 3-minute pauses were present. Also frequent VPCs with two triplets were detected. Sotalol was stopped, and daily 24-hour ECG monitorizations were repeated. No new event and pause were detected. During exercise test, VPCs disappeared after the heart rate of 110/bpm, and reappeared during recovery. During the following 5 months no new event occurred and repeated Holter monitorizations revealed only presence of uniform frequent VPCs.

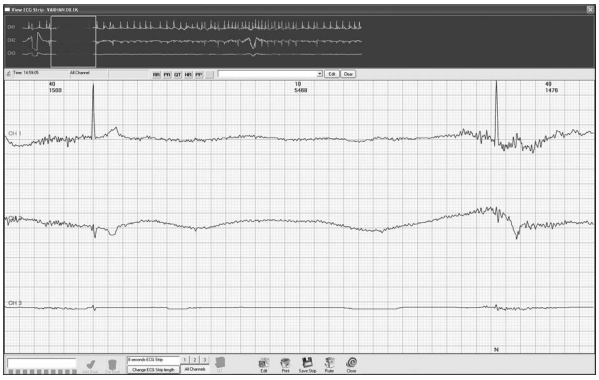

Fig. 1 Electrocardiographic recordings during presyncope showed 5.4-minute sinus arrest

Conclusion: Children who take sotalol, that is mostly known with its effects on cardiac repolarization, should also be carefully followed for sinus arrest that can result in presyncope/syncope. This side effect had not been reported in children previously.

P-3
Successful endocardial catheter ablation of epicardial premature ventricular complexes by contact force-guided lesion size formation

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Introduction: Transthoracic epicardial catheter ablation in the vicinity of the right aortic cusp may be problematic due to the risk of right coronary injury and epicardial fat. Furthermore, it may be associated with epicardial adhesions precluding epicardial re-access in later life, which should be considered in young patients. Since contact force is a key determinant of lesion size in irrigated radiofrequency catheter ablation, certain epicardial ventricular arrhythmias may be ablated from the endocardial space.

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Case report: A 16-year old male patient was referred for frequent premature ventricular contractions (PVC) leading to exercise intolerance. Antiarrhythmic medication failed to reduce symptoms and the frequency of PVCs. Activation mapping employing a 3D electroanatomical mapping system suggested an epicardial origin beneath the right coronary artery (local ventricular electrograms preceding the QRS onset by -23 ms and -26 ms in the right aortic sinus and in the inferior right ventricular outflow tract, respectively). Initial success could be achieved by targeting the RVOT exit site with an open irrigated ablation catheter (Celsius ThermoCool, Biosense Webster, Diamond Bar, CA/USA), however with recurrence after several days. PVCs also recurred after a second initially successful ablation procedure retargeting the RVOT exit site with a catheter design providing enhanced local irrigation (Therapy CoolFlex, St. Jude Medical, St. Paul, MN/USA). During the third and final procedure, the suspected epicardial origin could be reconfirmed by thorough activation mapping. While PVCs could not be abolished by conventional RF ablation from the right aortic cusp, contact force-guided RF energy delivery from the RVOT exit site yielding an estimated lesion depth of 10 mm (40 Watts, RF energy delivery duration 85 s, average force 25 g; TactiCath Quartz, Endosense SA, Geneva, Switzerland/St. Jude Medical) led to a definite ablation success. An injury of the right coronary artery was ruled out by coronary angiography. At 3 months of follow-up, the patient was free of palpitations and there was no recurrence of PVCs on 24h-holter monitoring.
Conclusions: Control of lesion size formation by contact-force guided catheter ablation may allow successful endocardial catheter ablation of certain epicardial ventricular arrhythmias. This novel approach may avoid epicardial access and its associated possible sequelae.

P-4

Efficacy of Implantable Loop Recorders in Establishing Symptom-Rhythm Correlation in Children with Unexplained Syncope; The first experience from Turkey

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Background: Syncope is a frequent complaint in children and adolescents. While the etiology is benign in the vast majority of cases, syncope may be an ominous sign for serious pathologies. Although the patient history, family history, and a physical examination are sufficient to make a diagnosis in most cases of syncope, are occasionally unable to establish a diagnosis. Despite a wide array of diagnostic tools (electrocardiogram, echocardiography, Holter monitoring, exercise testing, cardiac event monitoring, electroencephalographic recordings, tilt testing, and invasive electrophysiological study (EPS)) the cause of syncope is not determined yet after initial investigations in from one-third to half of all patients. In this study, our aim was to evaluate the diagnostic yield of the implantable loop recorder (ILR) in children with unexplained syncope.
Patients and Methods: Retrospective review of clinical data, indications, findings, and final management strategy in patients who underwent ILR implantation in two different centers.
Results: Between March 2010 and December 2013 a total of 12 patients (mean age of 9.3 ± 4.5 years) underwent ILR (Reveal Plus, Medtronic, USA) implantation. Indication was the syncope in all of the patients. Family history was normal in all of the patients. Routine cardiac assessment, including resting 12 lead electrocardiograms, transthoracic echocardiography, 24 hour Holter recordings and event recorder were normal except one patient with operated tetralogy of Fallot. ILR was implanted subsequently to the EPS in 6 patients because EPS did not reveal the etiology. After median 20 months (1–36 months), 6 patients presented with symptoms. ILR memory was showing torsades de pointes-ventricular fibrillation (3), CPVT (1), asystole and ventricular tachycardia (1), and normal sinus rhythm (1). ILR was explanted from prior 5 patients and transvenous ICD implantation was performed in prior four patients. Six patients are still in follow up with no symptoms for 25,2 months (20-35 months). Complications that needed reintervention were occurred in 2 patients.
Conclusion: Implantable loop recorders plays an important role in the diagnosis of life-threatening arrhythmias whose syncope is otherwise unexplained. ILRs should be remembered in children whose symptoms are strong correlated with rhythm disturbances.

P-5

Can the use of flecainide for incessant refractory supraventricular tachycardia reduce ablation requirements in neonates and infants?

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Objectives: The aim of this study was to evaluate and compare the results of medical and ablation therapy in neonates and infants with incessant refractory SVT.
Patients and Methods: The study included a sample of 11 neonates and infants receiving medical and/or ablation therapy in our center between January 2010-December 2013.
Results: The mean age and weight were 101.6 ± 96 days and 5.3 ± 1.9 kg. In the echocardiographic assessment, the mean fractional shortening of the cases treated with ablation and medical therapy were 24% ± 4 and 28% ± 7, respectively. Two patients had VSD, 1 had PDA, 1 had HCMP, 1 had a complex cardiac anomaly (c-TGA-VSD, right ventricular hypoplasia-pulmonary hypertension), and 6 patients had PFO. In the first two years of the four-year period, 5 patients had to receive ablation therapy, whereas the remaining 6 patients who presented over the last 2 years were treated with medical therapy to control the refractory SVT. The most noteworthy point here was the addition of flecainide into the medical therapy (which consisted of adenosine, amiodarone, esmolol, and propafenone cardioversion) administered before the ablation procedure (Flecainide as a class Ic agent has only been available in our country over the past two years). Interestingly, we managed to control the refractory SVT by administering a triple therapy regimen with esmolol-propranolol, amiodarone and flecainide in all of the 6 patients admitted in the last two years. The mean follow-up time was 18 months (range, 3 month-4 year). While one patient died during the follow-up, another patient had recurrent tachycardia. The patient diagnosed with myocarditis developed an atrial flutter complicated with a concealed accessory pathway. That patient was given ECMO support due to cardiopulmonary failure. Although the patient's SVT was controlled, we lost the patient on the 15th day of ECMO support. The patient with recurrent tachycardia, who had previously received ablation therapy for a complex cardiac anomaly and WPW, was treated with ablation again. No recurrence of tachycardia was observed in any of the other 9 patients.
Conclusion: It seems that the use of propranolol-esmolol and amiodarone combined with flecainide in the medical treatment of drug-resistant SVT may reduce the ablation requirement in neonates and infants.

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P-6

Swallowing Syncope in a 14 year-old girl with an atypical course of the oesophagus

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Introduction: Reports about swallow-induced syncopes are rare, mostly diagnosed after several years and multiple syncopes and seen in patients with cardiac diseases or oesophageal disorders such as achalasia, spasm, herniation, diverticulum and cancer.
Patient: We present a 14 year-old female who was admitted to hospital due to a single syncope. In subsequent Holter monitorings intermittent episodes of complete AV-Block with pauses up to 5.4 seconds could be documented.
The Holter monitoring showed a coincidence of the AV-block with meal times. Retrospectively the patient indicated to have dizziness always when eating.
An ECG recorded while the patient was eating and swallowing confirmed an intermittent complete AV-block.
A mediastinal MRI scan revealed an abnormal course of the oesophagus, which crosses caudal the thyroid gland to the left and runs for 2–3 cm in close proximity to the left common carotic artery and the vagus nerve.
According to the current guidelines a pacemaker was implanted to prevent bradycardia induced syncopes.
After implantation swallowing-induced pacemaker activity could be seen in ECG, the Holter monitoring showed also multiple pacemaker stimulation during meal times.
During follow up no further syncopes or dizzines have been noted, repeated Holter ECG and pacemaker interrogations confirmed that the pacemaker is in frequent use.
Conclusion: To the best of our knowledge this is the first report of an atypical course of the oesophagus in connection with a swallow-induced syncope. By this close proximity of the oesophagus and vagus nerve it is conceivable that swallowing can cause an abnormal vagal reflex leading to a high-degree AV-block.

P-7

Life threatening QT prolongation associated with congenital hydrocephalus in a preterm infant

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Introduction: Temporary QT-interval prolongation following intracranial haemorrhage and hydrocephalus has been repeatedly reported in adults.
Case: We present the first case of a preterm infant of 31 weeks gestational age suffering from intrauterine aqueductal stenosis. Postnatal cardiopulmonary adaption was unremarkable. Serum electrolytes were within normal limits and a cranial ultrasound showed hydrocephalus without signs of elevated intracranial pressure. On day two of life he developed sudden bradycardia with poor peripheral perfusion and mild arterial hypotension, which resolved spontaneously after 10 minutes. ECG showed a sinus rhythm with 70 bpm and an excessive QT prolongation (QTc 560 ms) resulting in 2:1 atrioventricular conduction due to ventricular refractoriness as reported in LQT patients. Noticeably, the AV nodal tissue is unaffected and AV conduction is normal in this patient. Several episodes of bradycardia occurred until day 5 of life when they finally stopped. Family history of LQT syndrome was negative and the identical twin brother had repetitive normal ECG's.
Discussion: This case illustrates a potentially life threatening QT prolongation associated with congenital hydrocephalus. The cause of the temporary QT prolongation remains unclear. Sympathetic-parasympathetic imbalance or electrolyte fluctuations are reported theories in adults. As this association has never been reported in a neonate before, it remains notional if the QT-interval was influenced rather by an intermittent intracranial pressure elevation, or if there was a hydrocephalus caused impairment of the sympathetic-parasympathetic balance during the process of perinatal adaption. The authors encourage hemodynamic monitoring of these patients as the reported phenomenon occurred suddenly, no data exists to identify affected individuals and serious hemodynamic compromise has to be expected in case of a longer-lasting episode.
Conclusion: This is the first report of excessive, life threatening QT prolongation associated with congenital hydrocephalus in a preterm infant.

P-8

Increased Microvolt T-wave Alternans in Pediatric Patients with Renal Failure Before Developing Severe Hypertrophy and Arrhythmia

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Background: The most important cause of morbidity and mortality in pediatric patients with chronic renal failure is cardiovascular involvement. There are no studies about non-invasive and low negative predictive tests used to determine the risk of ventricular arrhytmias and sudden cardiac death in children. Microvolt T-wave alternans (TWA) test is a non-invasive diagnostic method used in risk stratification for sudden cardiac death. The purpose of this study is to compare the measurements in microvolt TWA between pediatric patients with chronic renal failure and the control group.
Methods: Forty patients with chronic renal failure and 42 healthy controls were included in the study. The history, echocardigraphy and microvolt TWA values based on 24-hour ECG recordings of the patients were evaluated. Analysis of microvolt TWA was considered on the basis of three leads (V5, V1 and aVF).
Results: Interventricular septum thickening of the heart in patients with chronic renal failure was significantly higher compared to the control group ($p = 0.001$) but no increase observed in the wall of the left ventricle. However, end-systolic and end-diastolic volumes were significantly increased ($p = 0.04$ and $p = 0.01$ respectively). While there was no systolic disfunction, Mitral-E values decreased ($p = 0.015$). On the other hand, no severe arrhythmia except "kouplet ventricular extrasystole" in 2 patients observed. When TWA values compared between patients and controls, all of the three leads TWA values increased in chronic renal failure group, but the only statistically significant increase was in lead V5 ($p = 0.03$).
As a result, microvolt TWA values were increased before developing severe left ventricular hypertrophy and arrhythmia in pediatric patients with chronic renal failure. To determine the cut-off levels of microvolt TWA and to determine its possible relations with mortality, long-term follow-up of patients required and numerous, long-term studies are needed.

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P-9

Sudden arrhythmic death syndrome (SADS): Diagnostic yield of comprehensive clinical evaluation of paediatric first-degree relatives

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Background: Sudden arrhythmic death syndrome (SADS) is most often caused by heritable cardiac diseases. Studies in adults have identified evidence of inherited cardiovascular diseases in up to 53% of families, but data on the prevalence of familial disease in children are scarce. The aim of this study was to evaluate the yield of clinical screening in paediatric first-degree relatives of victims of SADS or aborted cardiac arrest (ACA) using a systematic and comprehensive protocol.
Methods: All consecutive paediatric patients referred for family screening between 2003 and 2013 after a SCD or ACA of a family member were retrospectively enrolled into the study. Systematic evaluation of the children included clinical examination, family history, ECG, echocardiogram, 24-hour tape and signal averaged ECG. Older patients also underwent exercise testing, cardiac MRI and ajmaline provocation testing.
Results: A total of 110 children from 63 consecutive families were included in the study. An inherited cardiac disease was identified in 12 1st-degree children from 11 (17.5%) families (7 children were diagnosed with Brugada syndrome, 2 with long QT syndrome, 1 with CPVT, and 2 had late potentials on signal averaged ECGs).
Conclusions: These data show a high prevalence of inherited heart disease in paediatric first-degree relative in families with a history of SCD or ACA. The results highlight the importance of a systematic, comprehensive approach and ongoing screening of paediatric family members.

P-10

Heterogeneity of ventricular repolarization in newborns with intrauterine growth restriction

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Introduction: Intrauterine growth restriction (IUGR) is associated with structural and functional cardiac alterations, but the possible electrophysiological consequences of these disturbances remain unknown. The aim of this study was to explore the distribution of ventricular repolarization and its relation to myocardial mechanics in newborns with IUGR.
Methods: Conventional and tissue Doppler echocardiographic data, and electrocardiographic parameters used to describe the distribution of ventricular repolarization (dispersion of QT [QTd] and JT [JTd]), were obtained on the second (D2) and fifth (D5) postnatal day and compared between 25 IUGR newborns and 25 matched-for-gestational age controls.
Results: IUGR was associated with relative intrerventricular septum hypertrophy, lower relative wall thickness (left ventricular posterior wall thickness to left ventricular end-diastolic diameter), increased left ventricular (LV) E/E' ratio and higher LV myocardial performance index (MPI). On both study days, the IUGR infants presented higher QTd and JTd compared to controls (QTd-D2: 66 $\pm$ 20 ms vs. 36 $\pm$ 12 ms, $P < 0.001$; JTd-D2: 54 $\pm$ 13 ms vs. 34 $\pm$ 9 ms, $P < 0.001$; QTd-D5: 61 $\pm$ 14 ms vs. 27 $\pm$ 12 ms, $P < 0.001$; JTd-D5: 54 $\pm$ 13 ms vs. 27 $\pm$ 9 ms, $P < 0.001$). The association between QTd and LV E/E' (D2: regression coefficient beta 0.747, R2 0.585; D5: beta 0.843, R2 0.646) and QTd and MPI (D2: beta 0.680, R2 0.576; D5: beta 0.698, R2 0.650) was also significant ($P < 0.001$ for all analyses).
Conclusion: Our findings suggest that IUGR is also associated with changes in the electrical activity of the neonatal heart, a process which is closely related to the underlying alterations in ventricular mechanics and might predispose to adverse electrophysiological events.

P-11

Monitoring Flecainide levels in Paediatric Arrhythmias

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Introduction: Flecainide is a class 1c antiarrhytmic agent which acts on sodium channels to prolong the action potential. It has been shown to be effective and safe in management of childhood arrhythmias. Optimum dose ranges are between 1-8 mg/kg per day. There is no consensus regarding at what intervals flecainide levels should be taken or whether they should be taken at all.
Methods: We interrogated our hospital pharmacy system between 2010 to 2013 to identify all patients less then 16 year old who have been prescribed Flecainide long term. A retrospective study was carried out to identify dose regimen and serum Flecainide levels.
Results: A total of 23 patients received Flecainide therapy between 2010 and 2013, 15 male and 8 female. All children had normal renal function. 20 patients were on treatment for supraventricular tachycardia and 3 for Ventricular Tachycardia.
10 patients were treated with multiple antiarrythmic agents including Atenolol, Propanolol, Digoxin, Sotalol and Amiodarone. The mean dose at commencement of therapy was 4.6 mg/kg/day (range 2.6 mg/kg/day to 6 mg/kg/day). 12/23 patients were on twice daily regimens and 11/23 on three times a day. The mean maintenance dose was 6.2 mg/kg/day (range 2.6 mg/kg/day to 12 mg/kg/day).
A total of 42 flecainide levels were taken over the 3 year study period. The average Flecainide level was 380 μg/L. The lowest level was 5 μg/L on 4 mg/kg/day. The highest was 994 μg/L on 10.5 mg/kg/day.
A total of 3 levels were recorded greater than 800 μg/L. 835 μg/L on 9 mg/kg/day, 922 μg/L on 6 mg/kg/day and 994 μg/L on 10.5 mg/kg/day. 28 levels were taken below 8 mg/kg/day with only 1 level above 800 μg/L (3.6%). 14 levels were taken above 8 mg/kg/day with 2 levels above 800 μg/L (14%).
Discussion: 3/42 patients had recorded levels above our cut off of 800 μg/L with only 1/28 being on less than 8 mg/kg/day and 2/14 above. At doses less than 8 mg/kg/day Flecainide is relatively safe and within the therapeutic range. Even at doses above this only 2/14 patients recorded a high level. We feel the efficacy of treatment can be monitored using ECG's and levels need only to be considered in dosage above 8 mg/kg/day or where there are clinical concerns about toxicity.

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P-12

Rhythm status in contemporary Fontan patients

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Introduction: Rhythm disturbances are an important cause of comorbidity in Fontan-patients. Currently, the total cavopulmonary connection (TCPC) is performed by using the intra-atrial lateral tunnel (ILT) or the extracardiac conduit (ECC). Aim of the study was to evaluate rhythm abnormalities and compare the surgical techniques in a contemporary cohort.
Methods: In a cross-sectional multicenter study 116 patients (mean age 12.5 ± 3.1 years) underwent rhythm evaluation including ECG, Holter, including heart rate variability (HRV) and exercise testing. Medical history was reviewed for documented episodes of arrhythmia.
Results: Sinus node dysfunction (SND) was found in 29% of patients, 3 of whom required pacemaker therapy. No difference was found in the incidence of SND between ILT and ECC patients. SND was associated with larger end-diastolic volumes as assessed by MRI ($p = 0.026$).
Sinus pauses occurred only in the ILT group. Exercise testing showed no difference in peak heart rate between the groups, however heart rate reserve ($p = 0.042$) and heart rate recovery ($p < 0.001$) were lower in ILT patients.
Heart rate variability was reduced compared to healthy controls, but was not different between ILT and ECC patients.
Atrial arrhythmias were more common in ILT patients (17% vs 2%, $p = 0.004$). Only one patient had symptomatic ventricular tachycardia (VT). However Holter recordings showed subclinical ventricular tachycardia in 6% of all patients, which was associated with larger end-diastolic ($p = 0.035$) and end-systolic volumes ($p = 0.029$).
Conclusion: Overall incidence of arrhythmia was relatively low in this cohort of modern Fontan patients. There was no difference in SND between ILT and ECC patients, but ILT patients had more atrial arrhythmias, sinus pauses and slower heart rate recovery. The significance of asymptomatic ventricular arrhythmias in this young Fontan population remains to be determined.

P-13

PocketECG-a new noninvasive, continuous and real-time ambulatory ECG monitoring in children with non-diagnosed tachycardia episodes – the first experiences

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Introduction: Episodes of tachycardia are common in children and adolescents, but usually it is a big problem with their documentation, especially when the incidents are short lasting. PocketECG system is a new technological solution enabling long term, noninvasive, real-time ECG monitoring with automatic diagnosis of arrhythmias, in which an ECG signal is transmitted over a mobile network.
Methods: We multicenter studied 262 children with non-diagnosed episodes of tachycardia at age from 4.2 to 18.5 years (mean 14.4 ± 3.2 years). In group I – 125 pts the standard 24-hour ECG recording was performed at the beginning of the study and the second one was done one month later. In group II – 137 pts one month lasting ECG monitoring was performed with the PocketECG system.
Results: In all 262 examined children we acquired Holter ECG and telemetric PocketECG recordings of good quality and final reports were prepared. In group I two standard 24-hour Holter monitoring was diagnostic only for 4.8% of the patients, the first examination for 3.2% (4 pts) of the group and, the second one for other 1.6% (2 pts), in 4 pts it was sinus tachycardia (ST), in 2 pts supraventricular tachycardia (SVT) was registered. In group II the new noninvasive, long term real-time ECG telemetric monitoring was effective for 26.3% of them, 10 children informed about the tachycardia and at that time SVT was diagnosed in 9 and ventricular tachycardia (VT) in 1, 19 pts felt tachycardia but it was ST only, in 6 children short episodes of SVT and in 1 VT was registered but they did not feel the incidents.
Conclusions: PocketECG system, as high-quality long term ECG recording with automatic analysis for continuous, noninvasive, real-time ECG monitoring is a reliable method for assessment of heart rhythm and arrhythmic events in children and adolescents and its use should be promoted.
In children and adolescents with non-diagnosed episodes of tachycardia 24-hour ECG monitoring is effective in about 5% of pts, prolongation of ECG monitoring helps to diagnose this disorders. In most of the patients who have felt tachycardia episodes during ECG monitoring sinus tachycardia was diagnosed.

P-14

The First Documentation Of Direct Contact Force Measurement In Catheter- induced Trauma During Ablation Of Atrial Tachycardia In Young Boy-Case Report

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Introduction: Catheter induced trauma (CIT) phenomenon is one of the causes of disappearing of arrhythmia due to mechanical block that may be used as mapping technique or may result in procedure failure. The new method of contact force measurement has been used to evaluate direct mechanical force of catheter on the cardiac tissue during radiofrequency ablation and mapping. There is no data of the direct measurement of contact force required for occurrence of mechanical trauma.
We describe the case of 16-year boy with atrial tachycardia who was referred for RF ablation. Clinical, persistent arrhythmia was reproducibly induced several times during EP study and after infusion of isoproterenol with earliest activation located in right inferior right atrium and tricuspid annulus. Immediately after positioning of ablation catheter on inferior aspect of tricuspid annulus below coronary ostium the "bump" phenomenon

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occurred. Contact force measurement confirmed consistent 20 g pattern of contact without inducibility of arrhythmia for next 3 minutes while catheter was in stable position. Then, the catheter was repositioned into right atrium and within 30 seconds arrhythmia became re-inducible. Than catheter was placed again in the same location and "bump" phenomenon appeared again with a very similar contact force pattern and no inducibility of arrythmia for the next 5 minutes. Afterwards, CIT was recognized and RF application was delivered with total time of 104 seconds. Then ablation catheter was repositioned into right ventricle and during 15 minutes observation period arrhythmia was no longer inducible with repeated infusions of isoproterenol and aggressive pacing protocols. C-MRI performed immediately and 6 weeks after procedure showed intermittent 8 mm tissue oedema in the target site of tricuspid annulus and no complications. The long-term (24 months) follow-up of the patient showed complete remission of arrhythmia.

The case presents interesting phenomenon associated with CIT and the first documentation of direct CF measurement required for CIT occurrence. Further studies are required to evaluate CIT phenomenon in various arrhythmias and location in various group of children and adults.

P-15

Neonatal myocardium is more sensitive to ischemia than the adult one

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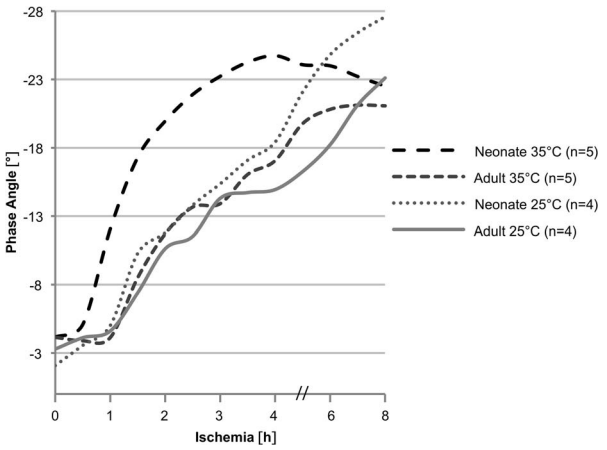
Objectives: Bioelectrical impedance measurement is a non-invasive method to determine alterations in tissue structures. Thus, neonatal and adult hearts were examined at different temperatures (35°C and 25°C) during ischemia to analyze distinctive organ specific changes in appropriate impedance spectra.

Material & Methods: Ischemic isolated hearts of piglets (NEO, n = 9) and adult pigs (ADULT, n = 9) were examined at 35°C or 25°C. Mean age of NEO was 7d (35°C) and 4d (25°C) and of ADULT 2.8 and 2.6 months, respectively. After harvesting of every heart an epicardial probe was placed on the left ventricle parallel to the LAD and continuous measurements of bioelectrical impedance were performed within a frequency range from 100 Hz to 1 MHz. All hearts were incubated at constant temperature (35°C, 25°C) for 24 hours. Furthermore, morphology was evaluated by ultrastructural analyses.

Results: In all groups the phase angle changed in an equivalent way demonstrating a sigmoid curve over time (see Fig.). However, significant differences between NEO and ADULT were evident, especially at 35°C (e.g. p = 0.009 after 4hrs). The maximal changes of the phase angle as indicator for the measured impedance were much higher in both neonatal groups (35°C: -24,74°; 25°C: -28.22°) compared to the adults (35°C: -21.12°; 25°C: -24,34°). Furthermore, at 25°C the onset of the increase in impedance occurred later in NEO and ADULT.

The calculated extracellular space index (ESI) as a marker for the ischemia-induced edema decreased from 54.9% (NEO) and 51.3% (ADULT), respectively, to 10.4% (NEO) and 13.5% (ADULT) at 35°C after 5 and 7 hours. At 25°C ESI decreased from 60.1% (NEO) and 52.5% (ADULT) to 8.4% (NEO) and 8.8% (ADULT) after 9 hours.

Ultrastructural analyses showed first reversible and later irreversible injuries.



Conclusions: Atraumatic measurements of bioimpedance provide an insight in intra-ischemic changes in myocardial membranes and in extracellular space. Ischemic stress-dependent alterations of myocardial tissue can be proven. Our results showed that neonatal myocardium is much more sensitive to ischemia-induced alterations, since irreversible myocardial injury occurs faster than in adult tissue. Thus, it's important to further expand our knowledge of neonatal myocardium during ischemia and thereby of intraoperative myocardial protection.

P-16

A missense mutation in the actin cardiac gene ACTC1 results in a varieties of congenital heart malformations and midline defects

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Family description: A Lebanese family had 15 members with congenital heart defect (CHD): 8 isolated ASD, 2 ASD and PS, 2 ASD and AS or AR, 1 ASD and Ebstein anomaly, 1 VSD and the founder died from cardiac failure although he had neither coronary disease nor left ventricle obstruction. In addition, WPW (2), sinus bradycardia (1), and atrial fibrillation (1) were observed. Beside cardiac anomalies, 5 had a midline defect: pectus excavatum (4), kyphoscoliosis (1), hypertelorism (3), cleft lip and diastema between superior incisors (1). All other family members had neither cardiac nor midline anomaly.

Positional mapping: The genotype of 365 poly (AC) were obtained from 20 family members. Parametric (disease allele frequency 0.001, penetrance 0.95, phenocopy rate 0.06) and non-parametric analyses were performed. There was a major peak reaching 1.99 in the parametric analysis and a single peak in the non parametric analysis (15.13) in the same chromosome 15 region. The haplotypes showed that all affecteds but one had the same allele for marker (D15S1007). A recombination on the centromeric side between D15S1007 and D15S165 and on the telomeric side between D15S1007 and D15S1012 in

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affecteds gave the limit of the mutation interval which encompassed 7,750,000 nucleotides.
Mutation identification: In this interval of 143 genes is located the ACTC1 gene which encodes cardiac actin. A heterozygous missense variant was observed in the ACTC1 gene which changed a methionine to a threonine at position 84 (c.251T> C). The Met84 residue is highly conserved in mammals, physico-chemical properties of both residues are different and all prediction software anticipated a disease causing change. This mutation was found in all affected family members but one who is presumably a phenocopy.
Cell biology: the cDNA of the Actc1 gene was cloned and the mutation was introduced by side-directed mutagenesis to transfect a cell line derived from atrial mouse cardiomyocytes (HL-1). We plan to observe whether the mutant protein is incorporated in actin filaments and whether the kinetics of this contracting cell line is altered due to the mutant protein.
Conclusion: A novel ACTC1 dominant mutation was identified which gave CHD, arrhythmic anomalies and midline defects.

P-17
Possible involvement of IL-23/IL-17 axis in pathogenesis of Kawasaki disease like vasculitis
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Introduction: Although mice of vasculitis induced with Candida albicans water-soluble fraction (CAWS) is widely used as an established model of Kawasaki disease (KD), the pathogenic mechanism of vasculitis has yet to be determined. Some studies have recently reported that IL-23/IL-17 axis is closely related to inflammation and autoimmune response. β-glucan and mannnan of a major component of CAWS enhance the production of IL-23 by dendritic cells through dectins which are receptors for β-glucan and mannnan. Therefore, to test the hypothesis that IL-17-producing cells (IL-23 receptor+) activated by IL-23 from dendritic cells initiate KD vasculitis, we investigated the distribution of IL-23 receptor and IL-17 in predilection sites of CAWS-induced vasculitis.
Methods: CAWS was intraperitoneally injected to mice for five consecutive days. At 1, 7, 14 days after CAWS injection, mice were sacrificed. We studied the expression of IL-23 receptor and IL-17 in the aortic root including coronary bifurcation, which is one of the most vulnerable sites in the experimental vasculitis, with immunohistochemistry.
Results: IL-23 receptor+ and IL-17+ cells presented in the aortic valves, the proximal region of it and adventitia of the aortic root in both CAWS-administrated mice and control. Furthermore, these cells apparently increased by administration of CAWS.
Conclusions: These findings suggest that IL-23/IL-17 axis activated by β-glucan and mannan is involved in the pathogenesis of CAWS-induced vasculitis, and can also explain a part of the mechanism to define the site specificity of vasculitis in this model.

P-18
Possible implication of Proline-rich tyrosine kinase 2 (Pyk2) in the pathogenesis od Kawasaki disease
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Introduction: Kawasaki disease (KD) is a pediatric vasculitis. Although its etiology remains elusive, a line of recent experimental studies implies that some kind(s) of infectious stimuli are implicate in the vasculitis through uncontrolled innate immune systems such as pattern recognition receptor (PPR)-mediated inflammatory signaling. Among macromolecules regarding in the PRRs-dependent signaling pathways, it has recently emerged that proline-rich protein tyrosine kinase (PYK2) is involved in the processes through NF-kB activation. Employing an established animal model for KD, thus, we investigated a possible relevance of Pyk2 in the pathogenesis of KD.
Methods: Pyk2-knock out (Pyk2-KO) and wild-type C57BL/6 mice (WT) were administered Candida albicans water-soluble fraction (CAWS) to induce KD-like vasculitis. Extension of the experimental vasculitis was immunohistochemically determined with anti-MPO antibody. CAWS-stimulated NF-kB activation was evaluated by quantifying nuclear translocation of NF-kB p65 subunit. In peritoneal macrophages isolated from PYK2-KO and wild-type mice in vitro.
Results: Pyk2-KO mice didn't show any apparent defective phenotype. While marked inflammation was observed in the aortic root of CAWS-treated WT mice, such vasculitis was barely detected in CAWS-treated Pyk2-KO mice. CAWS-induced NF-kB activation was also less observed in macrophages from Pyk2-KO mice.
Conclusions: These results indicate that Pyk2 play indispensable rules in the pathogenesis of KD. Pyk2 may be a potential therapeutic target for KD.

P-19
Influence of S. aureus coagulases on bacteria-vessel wall interactions in infective endocarditis
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Introduction: Staphylococcus aureus (S. aureus) is one of the major pathogens causing infective endocarditis (IE). Its high propensity to adhere to endothelial cells (ECs) and to spread via the bloodstream is associated with a high mortality. Von Willebrand factor (VWF) mediates shear-dependent platelet-vessel wall interaction. S. aureus binds VWF under shear stress, however, the mechanisms are incompletely understood. Furthermore, S. aureus activates the coagulation by staphylothrombin, a complex of S. aureus coagulases and prothrombin.
Aim: Given the key role of coagulase activity and VWF binding in the initiation and pathogenesis of intravascular infections, we aim to investigate if S. aureus von Willebrand factor binding protein (vWbp) is a key factor in mediating these two functions.

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Methods: Experiments were performed in a parallel flow chamber under shear stress of 10 dynes/cm2. By using either pharmacological coagulase inhibition, targeting both coagulases (staphylocoagulase and vWbp), or by using mutant strains lacking either staphylocoagulase, vWbp or both, we studied the contribution of VWF binding and coagulase activity in shear-dependent S. aureus adhesion. The effect of S. aureus mediated thrombin activity on EC activation, EC fibrin deposition and bacterial EC adhesion was studied in vitro. Furthermore, we evaluated the interaction of S. aureus Newman and staphylocoagulase- and vWbp-deficient mutants with activated ECs in vivo by using real-time fluorescence videomicroscopy in mesenteric vessels.
Results: We found that in contrast to thrombin, staphylothrombin does not directly activate ECs. However, S. aureus-mediated fibrin deposition increased bacterial retention to ECs in vitro. Furthermore, both coagulases induced the formation of circulating microthrombi in vivo. Staphylothrombin-mediated microthrombus formation and VWF binding both contributed to S. aureus adhesion to activated ECs suggesting a unique dual role for vWbp in bacteria-vessel wall interaction. Pharmacological inhibition of coagulases reduced bacterial adhesion to ECs in vivo. In the mouse IE model, genetic absence of coagulases increased survival.
Conclusion: S. aureus vWbp is a key factor in mediating coagulase activity and VWF binding and might serve as a target for alternative treatment strategies in S. aureus IE.

P-20

Homozygous loss-of-function mutation causes the lethal disorder mitogenic cardiomyopathy in two siblings

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Introduction: Two siblings from consanguineous parents of Turkish descent presented with isolated dilated cardiomyopathy, leading to early death in infancy. The diagnosis of an extremely rare and lethal disorder, mitogenic cardiomyopathy, was made histologically.
Methods: Genomewide parametric linkage analysis was performed, SNP typing platform was used in a recessive model. Genotyping was done in parents and both the unaffected and affected siblings.
Exome sequencing analysis was performed. Data analysis was done using commercial and in-house developed software. Only variants in genes from the linkage regions were retained. All homozygous calls were excluded in the parents and the unaffected sibling, reference calls were excluded in the affected sibling. Only exonic and splicing variants were included, synonymous variants were excluded. Variants occurring with a frequency of $<1\%$ in the 1000 genomes project or with an unknown frequency were included.
Results: Linkage analysis identified 8 regions. After variant filtering of the exome sequences, 6 candidate genes were identified in the linkage regions with homozygous mutations in the patient, inherited from both parents, and for which the unaffected sibling is heterozygous or reference. This gene list was manually curated using functional data and genotype-phenotype correlations. We identified a deleterious mutation in the ALMS1 gene as the most likely cause. Results were confirmed by Sanger sequencing. The two affected siblings are homozygous for a frameshift deletion of one basepair in the ALMS1 gene. This is predicted to cause a premature stop at position 5 downstream. The unaffected sister and parents are heterozygotes.
Conclusions: Linkage analysis combined with exome sequencing identified a homozygous deleterious mutation in the ALMS1 gene as the cause of this phenotype. Alström syndrome is characterized by a typically transient dilating cardiomyopathy in infancy, suggesting that mitogenic cardiomyopathy represents the extreme phenotype, resulting in demise before the other clinical symptoms become evident. This observation further illustrates the role of ALMS1 and cell cycle regulation. Reaching a genetic diagnosis in rare disorders remains a challenge. We illustrate that even in a single family with only two affected individuals, the identification of the underlying defect is feasible, using a combination of the sophisticated genetic tools.

P-21

Trio analysis using Next generation sequencing technology to identify de novo mutations in individuals with syndromic cardiopathy

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Introduction: Advances in genetic sequencing technology have the potential to enhance testing for genes associated with genetically heterogeneous diseases, such as congenital heart defects (CHD). Until recently, a major limitation in genetic research was genetic testing for mutations in a large number of genes simultaneously. Novel technologies such as next generation sequencing (NGS) using high-throughput massively parallel sequencing methods may circumvent this limitation. With current clinical and genetic investigation, an etiological diagnosis can be reached in 55% of cases. Nevertheless, there is a need for better diagnosis, since it is in this group that genetic counseling is frequently requested with regard to recurrence risks as well as prognosis with regard to intellectual disability. An important proportion of cases have a de novo monogenic cause, i.e. a newly occurring mutation, not present in one of the parents, and altering the function of a gene essential in embryonic development, including the heart.
Methods: Patients were included with a syndromic cardiopathy of unknown cause after extensive evaluation by an experienced clinical geneticist and with high resolution array-CGH in the patient and both parents showing no abnormal, unclassified variants. Whole exome sequencing analysis was performed. Data analysis was done using commercial and in-house developed software. Patients were filtered against 72 in-house exomes for detection of de novo mutations. Only exonic and splicing variants were included, synonymous variants were excluded. Variants occurring with a frequency of $<1\%$ in the 1000 genomes project or with an unknown frequency were included.
Results: Eight pairs of Trios were analysed. After variant filtering of the exome sequences, the gene list was manually curated using functional data and genotype-phenotype correlations. We identified a deleterious mutations in 4 cases (50%) as the most likely cause. Results were confirmed by Sanger sequencing.
Conclusions: Reaching a genetic diagnosis in individuals with a syndromic cardiopathy remains a challenge due to different factors including atypical or milder manifestations. We illustrate that using NGS technology in Trio-analysis, the identification of the underlying genetic mutation is feasible in most cases, thus also identifying new genes not previously associated with syndromic CHD.

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P-22

Sympathetic arousal in response to acute and chronic stress

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Electrodermal activity (EDA) is considered as a noinvasive index of sympathetic nervous system depending on the sweat glands activity and blood vessels. The aim of this study was to assess the effect of acute and chronic mental stress on the EDA and peripheral temperature in healthy young people.
Methods: Forty young people (mean age 23.1 ± 0.2 yr., 25 women) were examined in two periods: at the beginning of semester (P1, rest period) and one day before the last exam (P2, stress period) in the following order: rest (T1) – Stroop test (T2) – recovery (T3) – mental arithmetic test (T4) – recovery (T5) – negative emotional stress (T6) – recovery phase (T7). EDA evaluated as a skin conductance (μS) and peripheral skin temperature (°C) were continuously recorded and evaluated from each period lasting six minutes.
Results: EDA was significantly higher during acute mental stress (T2, T4, T6) and recovery phases (T3, T5, T7) compared with baseline values (T1, $p < 0.001$) in both periods (P1, P2). Peripheral temperature showed inverse pattern: significantly lower during stress (T2, T4, T6) and recovery phases (T3, T5, T7) compared with baseline values (T1, $p < 0.001$) in both periods (P1, P2). In chronic stress, EDA was significantly lower in the stress period (P2) compared to the rest period (P1, $p < 0.001$). In contrast, peripheral temperature was without significant changes in the P2 compared to the P1.
Conclusion: EDA and peripheral temperature could represent the sensitive markers of sympathetic arousal in response to acute mental stress. Both parameters did not return to baseline values indicating a potential sympathetic excitation after acute stress. Interestingly, reduced sympathetic activity indexed by lower EDA in the stress period might indicate an allostatic overload evoked by a long-term daily intensive study in healthy students. We suggest that the altered sympathetic response can be associated with a higher risk of the cardiovascular complications in chronic stress. Support: VEGA 1/0087/14, APVV 1/0235/12.

P-23

Prognostic value of BNP in newborns with congenital heart defects

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Introduction: B-type natriuretic peptide (BNP) is elevated during ventricular strain. Data are lacking on whether the use of BNP improves the prognosis in newborns with congenital heart defects (CHD).
Methods: A prospective study of a cohort of newborns with CHD admitted at a NICU in a tertiary hospital was performed between August 2012 and December 2013. Participants had BNP evaluated in the cord blood or at the first biochemical sample collected for clinical purposes within the first 48 hours of life.
Results: From the 26 patients with CHD amenable to surgery and with BNP determined within the first 48 hours of life, 10 had cardiac surgery in the newborn period: systemic to pulmonary shunts (n = 3); arterial switch (n = 2); Norwood (n = 1); aortic valvuloplasty (n = 1); pulmonary banding (n = 1); correction of coartation of aorta (n = 1); correction of interruption of aortic arch (n = 1). Patients that needed cardiac surgery in the newborn period had statistically significant higher BNP levels (73.7 pg/mL) in the first 48 h of life than patients that were able to go home without surgery (27.6 pg/mL) ($p = 0.011$).
Conclusions: BNP levels in first hours of life seem to have a prognostic value for the need of neonatal cardiac surgery. However, BNP is not a stand-alone test, it should be a complement of history, physical examination, echocardiography and clinical judgment.

P-24

The Effect of modafinil in pulmonary hypertension rat models

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Introduction: Pulmonary arterial hypertension (PAH) is difficult to treat and is characterized by increased pulmonary arterial pressure. PAH causes right ventricular failure and possibly even death due to progressive increase in pulmonary vascular resistance. PAH has been shown to be refractory to most of the conventional pharmacological therapies. So we want to verify the effect of modafinil in pulmonary hypertension rat models. Modafinil increases cyclic AMP concentrations in aortic smooth muscle cell (SMC)s and phosphorylated KCa3.1 channels. KCa3.1 channels are related to vessel relaxations and proliferation of SMCs. There was no any report about modafinil in pulmonary hypertension.
Methods: Six-week-old male Sprague Dawley rats were used. The rats were grouped as follows: The control (C) group (subcutaneous injection of saline), monocrotaline (MCT) group (subcutaneous injection of MCT 60 mg/kg), modafinil group (MD group) (gavage feeding of modafinil 50 mg/kg/day). One day after MCT injection, rats were sacrificed in weeks 1, 2 and 4. Pulmonary arterial pressure (PAP) was measured by a catheter introduced into the internal jugular vein.
Results: The mean RV pressure significantly increased in the MCT group compared to the C group and significantly decreased in the MD group compared with the MCT group in weeks 1, 2 and 4. Systemic pressure showed no significant changes in the three groups. The ratio of RV/LV+septum significantly increased in the MCT group compared to the C group in weeks 2 and 4 and significantly decreased in the MD group compared to the MCT group in week 4.
Conclusion: After modafinil treatment, there were improvements of RVH and mean RV pressure. Additional research on the dosage and frequency of modafinil is needed to determine the optimal parameters for PAH treatment.

P-25

Changes of pulmonary pathology and genes according to dose of umbilical cord blood derived mesenchymal stem cells in monocrotaline-induced pulmonary artery hypertension rat

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Introduction: Pulmonary arterial hypertension (PAH) causes right ventricular failure and possibly even death due to a progressive increase in pulmonary vascular resistance. Human umbilical cord blood derived mesenchymal stem cells (hUCB-MSCs) transfusion have recently been studied to evaluate their potential as a source of cell therapy. The purposes of this study are to investigate changes of hemodynamics, pulmonary pathology and ten gene expressions such as ET (endothelin)-1, ET receptor A (ERA), endothelial nitric oxide synthase (eNOS), matrix metalloproteinase (MMP)-2, tissue inhibitor of MMP (TIMP)-1, interleukin (IL)-6, tumor necrosis factor (TNF)-α, Bcl (B cell leukemia/lymphoma)-2, caspase-3, vascular endothelial growth factor (VEGF) according to dose in monocrotaline (MCT)-induced PAH rat models after hUCB-MSCs transfusion.
Methods: The rats were divided into four groups as follows: the control (C) group (subcutaneous injection of saline 0.1 mL/kg), M group (subcutaneous injection of MCT 60 mg/kg), the UA group (hUCB-MSCs transfusion 1.5 × 106/mL/cm2), the UB group (hUCB-MSCs transfusion 3 × 105/mL/cm2). They received transfusion through the internal jugular vein 1 week after MCT injection.
Results: The mean right ventricular pressure (RVP) significantly decreased in the UA and the UB group compared with the M group in weeks 2 and 4. Right ventricle (RV) weight and the ratio of RV/left ventricle (LV)+septum significantly decreased in the UA and the UB group compared with the M group in week 2. Medial wall thickness of the pulmonary arteriols was significantly decreased in the UA group compared with the UB group in week 4. The number of intra-acinar arteriols was significantly decreased in the UB group compared with the UA group in week 4. Gene expressions of ET-1, ERA, eNOS, MMP-2, TIMP-1, IL-6, TNF-α, Bcl-2, caspase-3 and VEGF significantly decreased in the UA and the UB group compared with the M group.
Conclusion: After a transfusion of the UB dosage of hUCB-MSCs, there was also a similar improvement of RVH and mean RVP compared with UA dosage. Decreases in several gene expressions were also observed. Additional research on determing the optimal dose of hUCB-MSCs infusion is needed in PAH treatment.

P-26
How to Correctly Evaluate Blood Pressure Values in Children? Strengths, Limitations and Gaps of Current Pediatric Blood Pressure Nomograms: a Critical Review

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Background: Despite blood pressure (BP) is an essential parameter at all the age and in any clinical setting, various limitations still persists in current pediatric nomograms for BP.
Aim: To review the strength/limitation of current pediatric BP nomograms.
Methods: A literature search was performed in June 2013 within the National Library of Medicine using the keywords *BP, pediatric, reference values/nomograms.* The search was further refined by adding the keywords *neonates/infants/adolescents, range/intervals*. Analysis of sample size and its characteristic (age distribution, morphometric details, racial differences) was previously performed. Secondly we evaluated methodological/statistical issues: how BP was measured, how data have been normalized, how they have been expressed, whether a correct analysis was applied and whether potential counfounders/source of errors have been investigated.

Table-1: Major pediatric BP nomograms divided according to geographic area. G=gender, M=males

Author	Sample Size	Age Interval	W/H/BMI	G (M)
America				
Minter USA 2004	5582	5-17 yrs	nr	51,1%
Nichols 2006 Trinidad and Tobago	3749	12-18 yrs	M/F BMI 20.6 (4.2),21.8 (5)	1610
Paradis 2004 Canada	3589	9-16 yrs	nr	nr
Park Texas USA1989	1554	2 weeks-5 yrs		747
Park 2006 Texas USA	7208	5-17 yrs	nr	nr
Rosner 2007 USA	49,967*	1-17 yrs	nr	25651
Fourth report 2004 USA	63227	1-17 yrs	nr	51%
Australia				
Blake 2000 Perth Australia	2876	1-6 yrs	BMI 17.0 (1.4) 16.2 (1.3) 15.8 (1.8)	546
Roy 1984 Australia	9851	5-13 yrs	18-53 kg	4884
Europe				
Andre France 1980	17067	4-18 yrs	nr	8647
De Min 2004 France	28043	4-19 yrs	nr	nr
De Swiet 1992 London	1895	4 days-10 yrs	nr	nr
Menghetti 1999 Italy	11519	5-17 years	nr	6258
Sanchez Spain 1992	34986	1-18 years	nr	nr
Tuner 1999 Turkish	5599	0-18 y.o.	nr	2835
Asian				
Mehri 2009 Beirut	5710	5-15 yrs	nr	2918
Chadha 1999 India	8293	5-14 yrs	Every year	4623
Jafar Pakistan 2005	5641	5-14 yrs	BMI 152. (4.3) M 15.3 (4.3)	2974
Kelishadi 2006 Iran	21111	6-18 yrs	BMI 18.53±3.84	10253
Krishna 2006 India	2278	3-18 yrs	nr	2500
Okumura 1981 Japan	17422	7-18 yrs	nr	nr
Sharma 1991 India	2453	7-16 yrs	For every year	159
Ayatollahi 2012 Iran	2270	6.5-11.5 yrs	nr	1174

Results: 66 studies were selected for final analysis. The analysis highlighted the accuracy of latest studies but also underscored that some limitations still remain. In many studies the number of healthy subjects was limited, particularly for some age groups (i.e. neonates/infants), some geographic area and ethnic groups. The methodologies utilized for the performance and normalization of measurements were heterogeneous as well as the expression of the normalized data (percentiles/mean values). Although most studies adjusted measurements for age and/or height, classification of age and/or height subgroups varied, and the relationships of the measurements to other morphometric parameters (i.e. body size and heart rate) were not always addressed. Gender differences were generally but not universally accounted while other confounders such as ethnicity, hereditary, seasonal and socio-economical variations were seldom evaluated.
Hence, while for older children reference intervals were substantially reproducible, in neonates and infants greatly varied. Data on ambulatory BP and response to stress test furthermore are extremely limited and criteria of stress test interruption even absent.
Conclusions: Valid BP pediatric nomograms have been recently become available. However a comprehensive nomogram, which involves a large population of healthy children, uses standardized methodology, and fully evaluate the influence of various confounders is still lacking thus affecting the accuracy in the evaluation of BP in children, especially in neonates.

P-27
Molecular lesions in right ventricular infundibulum in Tetralogy of Fallot

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Introduction: Narrowing of the sub pulmonic infundibulum of right ventricle is characteristics of Tetralogy of Fallot (TOF). Transcription factors, signaling molecules, and structural proteins involved in embryonic development of right ventricular outflow

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tract (RVOT) could be key candidates in the pathogenesis of the infundibular stenosis from increased muscle mass in the region.
Objective: To elucidate the causal factors for regional muscle growth the gene expression pattern of cardiac transcription factors such as Mef2c, HAND2, GATA4 and ISL1 and proteomic profile of RVOT myocardium of patients with TOF were studied.
Methods: Sub pulmonic infundibular muscle tissue were obtained from six patients with TOF who underwent corrective surgery (age: 1 to 19 years) and four donor healthy human hearts harvested for transplantation (age: 11 to 40 years). The expression of candidate genes were analyzed and validated by semi quantitative RT-PCR (Applied Biosystems). Tissue proteins were subjected to proteomics protein expression analysis by 2D nano- LC ms /MS (Waters). Immunoblot experiments were performed to substantiate differential expression of selected proteins.
Results: An increased expression of Islet-1 (ISL1) was observed in all patients. ISL1 is a member of LIM-homeodomain transcription factor family and a marker of resident cardiac progenitor cells that are derived from second heart field (SHF) region. Mef2c, GATA4 and HAND2 had a decreased expression pattern in patients with TOF. In LC ms/MS analysis, 1500 proteins were identified in the infundibular muscle tissue among which 113 proteins were differentially expressed in patients and normal individuals. Among them, Retinaldehyde dehydrogenase 2 (ALDH1A2), an upstream element required for the down-regulation of ISL1 was decreased while there was increased expression of proteins such as MAP4, required for cellular growth and division.
Conclusions: Decreased expression of transcription factors such as Mef2c, HAND2, GATA4 in the myocardium of patients with TOF suggest the underdevelopment of RVOT. Increased expression of ISL1 together with proteins for cell proliferation in RVOT indicate the presence of progenitor cells in the region, their inadequate differentiation and proliferative potential, all of which could contribute to the infundibular stenosis.

P-28
Tissue engineered heart valve laeflet substitute in orthotopic aortic position in a sheep model – preliminary results

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Objectives: Currently used patch materials are associated with limited longevity of aortic valve repair. To overcome this limitation, we try to create a living replacement material with regeneration and growth capacity made of homologous cells.
Methods: Myofibroblasts harvested from umbilical cord of a lamb are isolated, cultivated and expanded at 37°C for 14 days. Then the cells are seeded in two steps at 24 hour interval on the scaffold. Seven days static cultures are followed by cultivation in a bioreactor for another 14 days. Then the tissue engineered patch (TEP) is implanted in the adult swiss white mountain sheep (59-70 kg). After median sternotomy or lateral thoracotomy extra-corporeal circulation (ECC) is established. The aortic valve is exposed. The acoronary leaflet is explanted and the TEP tailored to implant it as leaflet substitute. 6 hours later the animal is sacrificed. Post mortem analyses including histology are done.
Results: So far, we operated on seven sheep (median sternotomy [n:6], right lateral thoracotomy [n:1]). One sheep developed ventricular fibrillation (VF) due to unknown reason right after sternotomy; after ending of ECC VF recurred and the sheep died. Nevertheless, the operation was finished in all seven attempts. One sheep developed VF refractory to medical or electrical treatment after weaning from ECC and died also before planned scarification. Mean follow-up time after chest closure was 3.2 hours. Postoperative transthoracic, epicardial or transesophageal 2 D colour Doppler echocardiography revealed excellent function of leaflet substitute with good coadaptation of the aortic valve leaflets without signs of relevant aortic valve stenosis (none) or regurgition (none or trivial). In one case an angiogram was done demonstrating a sufficient valve. CT scan was done in 3 cases (2 to 3 hours post.op) demonstrating none to trivial aortic valve regurgitation. Obtained histologic samples at sacrification showed cell migration (red blood cells as well as lymphocytes) into the scaffold.
Conclusions: Tissue engineered heart valve laeflet substitute in orthotopic aortic position in a sheep model shows excellent acute hemodynamic results regarding aortic valve function. Chronic animal model testing is needed to test longevity of this new approach.

P-29
Left Ventricular Assist Device Support for 2 Months Followed by V-A ECMO for 68 Days as a Bridge to Cardiac Transplantation in an Infant with Dilative Cardiomyopathy

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Introduction: Infants and young children are considered the most difficult group to bridge to orthotopic heart transplantation. Due to donor shortage, many patients die while on waiting list. Although pumps designed for small children have been introduced and are used as a bridge to transplantation or recovery, mid-term or long-term mechanical circulatory support (MCS) for small children with heart failure is still difficult.
Case report: We report on a 7-month-old male infant (weight 8 kg) with advanced heart failure due to idiopathic dilative cardiomiopathy, who was successfully supported for 130 days with left ventricular assist device (LVAD) with centrifugal pump (62 days) and V-A ECMO (68 days) before orthotopic heart transplantation. Infant developed pulmonary oedema and oliguric renal failure due to progressive left ventricular failure while on Eurotransplant high-urgent heart transplant list. The decision was made to implant the Levitronix↓ centrifugal pump. Attempts at weaning were unsuccessful. Long-term pediatric MCS system (BerlinHeart Excor) was not available at our institution at the time. Infant developed Serratia marcescens mediastinitis with septic shock. Support was switched to V-A ECMO because of ARDS and worsening right ventricular function. Renal function recovered after initiation of MCS. There were no thrombembolic events. No graft failure, acute rejection or infection were noted in the early posttransplantation period. 18 months after transplantatation the child is doing well.

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Intensive developmental therapy resulted in significant improvement in functional status.
Conclusion: Infants with end stage heart failure, are the most challenging group to bridge to heart transplantation with currently available MCS support systems. To our knowledge, LVAD support with the centrifugal pump followed by V-A ECMO in an infant of such duration as a bridge to successful heart transplantation has previously not been reported.

P-30
Effect of cardiac resynchronization therapy in children with dilated cardiomyopathy and refractory heart failure with a narrow QRS complex
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Introduction and objective: Dilated cardiomyopathy (DCM) is the most common cardiomyopathy in children. In México, half of the cases die on the waiting list for a heart transplant. Cardiac resynchronization therapy (CRT) has demonstrated efficacy with an improvement in functional class, ventricular function and survival in the adult population with a QRS complex >150 mseg. Currently there are no guidelines for the application of CRT in children with heart failure (HF). The objective of the study was to analyze the effect of CRT on ventricular function and functional class in children with refractory HF secondary to DCM with or without a QRS complex >150 mseg.
Methods: Open clinical trial, uncontrolled, non-randomized (intervention study of a single group, with assessment before and after). Cardiac dyssynchrony was assessed with real-time three-dimensional echocardiography obtaining an index of asynchrony (IA).
Results: Seven children with DCM received CRT from october 2013 to november 2014. Four girls. Age 3.02–14.6 years, weight 11.3–61 kg, QRS: 93 ± 31.5, ejection fraction (LVEF) 8-30%, AI) 5–11.9. Four children were in functional class III, Two in II and one in IV. Two implants were performed via epicardial and endocardial via 5. Time of hospitalization was 2-12 days. A girl with DCM secondary to anthracycline in functional class IV with dysfunction in more than 2 organs, died at 11 days after implantation. In the six living children, the functional class improved by at least 1 category, the LVEF improved at least 40% from baseline and the IA was less than 5 in all. No complications were associated with the implant.
Conclusions: CRT showed improvement in functional class and in ventricular function in a small group of children with refractory HF secondary to DCM with a narrow QRS complex.

P-31
Diagnostics and clinical patterns of idiopathic pulmonary hypertension in Russia. A two-center study
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Objective: To identify clinical, functional, haemodynamic, and biochemical characteristics of idiopathic pulmonary arterial hypertension (iPAH) in children.
Methods: 90 pts with PAH (52% female) aged 4 months—17 years studied in Jan 2011 – Dec 2013. Tests included routine cardiological examination; CT angiography & scintigraphy; HIV, thyroid, liver function tests; ruling out lupus, scleroderma, rheumatoid arthritis; endothelin & NT-proBNP measurements; six-minute walk test (6MWT – age 7+ only); right heart catheterization (RHC-in 76%).
Results: iPAH confirmed in 21 (23%, thereof 71% female). At first examination 24% had functional class (FC) II, 47.5%-FC III; 28.5%-FC IV. 76% had dyspnea aggravated by physical effort. Syncope registered in 4 pts; recurrent hemoptysis in 3 pts. Median duration of symptoms was 23 (2-48) months. Dilatation of the right heart chambers, elevated RV wall thickness, dilated main PA, short acceleration time of RV ejection, and abnormal end-diastolic septal curve found in 86%. RV ejection fraction decreased in 57% of pts (29.5 ± 3.2%). Tricuspid annular plane systolic excursion (TAPSE) constituted 14.2 ± 4.7 mm. arked variability of mean pulmonary artery pressure (32 to 98 mmHg) and pulmonary vascular resistance (8.5 to 29.3 UW) were detected. Cardiac index was 1.9 ± 0.5 L/min/m^2 and median arterial oxygen saturation was 91.5 ± 2.7%. Elevated values of endothelin (0.43 ± 0.18 pmol/ml) and NT-proBNP (64.8 ± 18.6 pg/ml) were found. Two pts were treated with sildenafil, 13 pts-with bosentan and 6 pts-with a combination of the two drugs. In 5 pts the atrioseptostomy was performed. Median duration of follow-up was 36 months (2 to 42 months). Reduction of dyspnea and improvement of functional characteristics observed in 11 (52%) pts. 6MWT distance increased by 156.2 ± 12.7 m. RV ejection fraction reached 32.1 ± 4.5%, pulmonary vascular resistance decreased by 8.9 ± 3.4 UW. Five IV class pts and two III class pts have died.
Conclusions: Late diagnostics of iPAH in children is common and was observed in 71% of pts. FC and hemodynamic parameters are the strongest predictors of survival in children with iPAH. Reduction of dyspnea and increased distance on the 6MWT are the main markers of a positive response to therapy.

P-32
Abnormal ductus venosus peak velocity index measured during fetal echocardiographic examination can predict failure of functional closure of the foramen ovale in the postnatal period
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Objectives: To investigate the applicability of ductus venosus (DV) wave velocities and/or DV peak velocity index (DV-PVIV) for veins in the prediction of failure of the foramen ovale to close functionally in the early postnatal period.
Methods: For this prospective study, we performed fetal echocardiography on 400 healthy women with uneventful pregnancies between the 20-24th gestational weeks. Ductus venosus blood flow and DV-PVIV data was collected using 2D and PW Doppler echocardiography. All subjects were called for a repeat echocardiography on the 30th postnatal day. Newborns with clearly visible foraminal flaps, interatrial septal defects smaller than 5 mm and right-to-left shunting through the defect were accepted as patent foramen ovale (PFO). Prenatal ductus

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venosus blood flow velocities and DV-PVIV data of the fetuses with and without PFO on the postnatal 30th day were compared.
Results: Data concerning the BMI of the pregnant women, birth weight and gender of the newborns were similar ($p > 0.05$). A statistically significant difference was detected between the groups with and without PFO in terms of mean DV-PVIV values (0.61 ± 0.12 and 0.73 ± 0.12, respectively; $p < 0.001$), DV-D wave velocity (52.14 ± 10.70 and 48.23 ± 11.40, respectively; $p = 0.003$) and DV-a wave velocity (30.55 ± 6.72 and 25.09 ± 6.73, $p < 0.001$). ROC (receiver operating characteristic) analysis showed that increased DV-PVIV values are related to and predictive of PFO (AUC = 0.75; $p < 0.001$) and that a threshold value of 0.62, which is associated with the highest Youden index, has a sensitivity of 86.8% (95% CI %: 78.1-93.0%) and a specificity of 51.7% (95% CI: 46.1-57.5%).
Conclusions: Ductus venosus peak velocity index for veins detected above 0.62 at the time of fetal echocardiographic examination may be used as an early predictor for failure of functional closure of the foramen ovale.

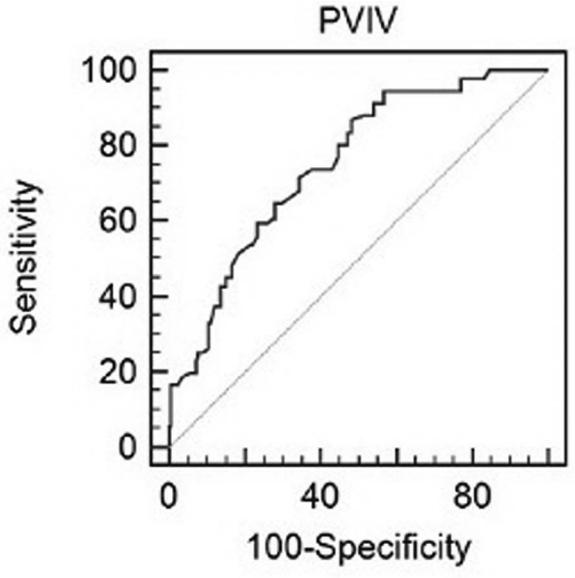

Figure 1. ROC curve of DV-PVIV for discrimination between the groups with and without PFO.

P-33
Impact of gestational diabetes mellitus on fetal cardiac morphology and performance
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Purpose: Fetal echocardiogram is indicated in several reasons like family history of congenital heart disease (CHD), maternal illness, abnormalities determined during fetal screening, etc. Gestational diabetes (GD) is an important maternal cause of fetal cardiac morphological and functional deterioration. In this respect, our aim was to evaluate the impact of GD on fetal cardiovascular system.
Methods: 146 women (56 of GD and 90 healthy subjects) at gestational ages between 17 and 32, underwent fetal echocardiographic evaluation in our institution from 2008 to 2011 were recruited a prospective cross sectional case control study. Diagnosis of GD was based on fasting serum glucose and oral glucose tolerance test. Fetal cardiovascular morphology, systolic and diastolic functions were assessed by 2D, M – Mode and Doppler echocardiography.
Results: Gestational diabetic pregnants (GDPs) were significantly obese (71,4%, BMI > 25, $p = 0{,}003$) and their HbA1c levels were higher than control group ($p < 0.001$). The frequency of fetuses with CHD was 9,5% where 10% in patient group and 8,9% in healthy subjects. The major cardiac malformation was VSD (2%). Systolic left ventricular free wall and septal thickness, aortic and pulmonary peak systolic velocities were significantly higher in fetuses of GDPs ($43{,}49 \pm 11{,}21$ mm, $p = 0{,}001$; $34{,}66 \pm 7{,}12$ mm, $p = 0{,}002$; $88{,}05 \pm 15{,}38$ cm/sec, $p = 0{,}006$; $64{,}17 \pm 8{,}34$ cm/sec, $p = 0{,}007$ respectively). Mean ejection fraction (EF) of GDPs fetuses were $77{,}88 \pm 10{,}87\%$ and $78{,}43 \pm 8{,}08$ ($p = 0{,}744$), however fractional shortening (FS) was found increased in GDPs' fetuses ($44{,}47 \pm 10{,}05$; $p = 0{,}048$). GDPs fetuses tricuspid mean early/atrial velocity ratio (E/A) was $0{,}72 \pm 0{,}06$ and mitral E/A was $0{,}79 \pm 0{,}09$, which wasn't different from healthy pregnants' fetuses ($p > 0.05$). Mean myocardial performance indexes of left and right ventricles were 0.28 and 0.210, where they were 0.30 and 0.31 in the control group ($p = 0{,}088$ and $0{,}069$ respectively).
Conclusions: High frequencies of structural abnormalities was thought of our institution being a tertiary center. Higher velocities in great vessels, increase in left ventricular wall thickness and diastolic dysfunction of both ventricle were supporting previous publications. Increased FS while the EF remaining similar when compared with healthy pregnant fetuses were different from the previous reports. These findings suggested that there may be a global geometric distortion in ventricular contraction of the affected fetal myocardium

P-34
Congenital Heart Disease and the Placenta
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Introduction: The cardiac-placental axis is associated with parallel development of the placenta and heart that utilizes many common molecules and reflects synergistic growth of both organs. Little information exists on placental pathology in the presence of fetal congenital heart disease (CHD). Therefore; our objective was to describe and characterize the placenta in neonates with CHD and to investigate the association between CHD and placental abnormalities in fetus.
Methods: This study is a retrospective case series. Cases include all infants who were born at Johns Hopkins Bayview Medical Center (JHBMC) and underwent cardiothoracic surgery at Johns Hopkins Medical Institution (JHMI) within 6 months of life from 2000-2013. Cases with the following characteristics were excluded: maternal diabetes, maternal hypertension, maternal coagulopathy, maternal renal failure, alcohol, smoking, and illicit drugs due to established confounding effect on the placenta. The following infant characteristics were identified and excluded as well: isolated PDA, mild CHD, chromosomal abnormalities and multiple births. Eight out of the 59 patients at JHBMC had met the inclusion criteria. These eight placentas were examined by a pathologist with expertise in study of the placenta.
Results: The mean gestational age for this CHD sample was 35 weeks. Infants in this sample had average placental BW

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ratio = 0.20, and median placental BW ratio = 0.19, which is a normal ratio for this gestational age. We found that 50% of CHD cases had eccentric cord insertion on the placenta. One out of the eight placentas had a small infarct, 2 showed signs of acute chorioamnionitis, two had meconium staining, and three were characterized as normal.
Conclusion: Eccentric cord insertion was present in placenta of half of CHD cases, which suggests placental implantation that has been described in other congenital defects. No other obvious abnormalities were found in this small sample. Larger studies are needed to characterize the placenta in fetal congenital heart disease.

P-35
Prenatal diagnosis of cardiac tumours and postnatal outcomes
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Introduction: Primary cardiac tumours are rare, with a reported prevalence of 0.001 to 0.03% at autopsy series. They are usually benign and may induce life threatening symptoms both, in the pre and postnatal periods. The cardiac manifestations are dependent on location and extension of the masses.
Objectives: The aim of this study was to evaluate the morphologic/hemodynamic/functional findings by fetal echocardiography as well as the postnatal outcomes.
Methods: The study group consisted of 20 fetuses with cardiac tumours followed from August 1998 to December 2013. The tumours were classified in small ($<30\,mm^2$), medium (>30 and $<100\,mm^2$) and large ($>100\,mm^2$).
Results; Maternal age ranged from 17 to 33 years (mean = 22.5 ± 3.6y) and the gestational age at diagnosis ranged from 22 to 34 weeks (mean = 31.1 ± 4.8w). The majority (72.7%) of tumours were small (mean = $11{,}0\,mm^2$); 21.2% were large (mean = $272.7\,mm^2$) and 6.1% were classified as medium sized (mean = $60.5\,mm^2$). Rhabdomyoma, the most common tumour, accounted for 60.0% (12/20) of all cases, and tuberous sclerosis were diagnosed in 8(66.6%) of these patients. There were no histological diagnosis in four patients; mixoma was confirmed in three and teratoma in one. Nineteen fetuses had intracardiac tumours and only one had a pericardial tumour. According to the location, 65.0% (13/20) were seen in the right chambers, 30.0% (5/20) in the left chambers and 5.0% (1/20) in the pericardium. The large tumours were associated with blood flow obstruction ($p = 0.01$), ventricular dysfunction ($p = 0.00$), arrhythmia ($p = 0.03$), congestive heart failure with pericardial effusion, cardiomegaly, hydrops ($p = 0.01$) and death ($p = 0.00$). There were four deaths, three intrauterine and one in the early neonatal period. Of these cases, two patients developed arrhythmia and three had large tumours.
Conclusions: All the identified tumours were benign, and rhabdomyoma was the most common. Tuberous sclerosis was observed in almost half the fetuses. Hemodynamic/functional disturbances and death were correlated with large tumours and location of the masses at the ventricular outlets or inlets. The widespread use of fetal echocardiography has significantly contributed to both, earlier diagnosis and treatment, and thus improved postnatal survival in this population.

P-36
Prenatal diagnosis of Tricuspid Atresia in a family with hereditability positive for NKx2.5 mutation, atrial septal defect and atrioventricular block
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Introduction: Mutations in NKx2-5 have been described in families with autosomal dominant heritability for secundum atrial septal defect (II-ASD) and progressive atrioventricular (AV) block. We report a case of prenatal diagnosis of Tricuspid Atresia (TA) in a family with hereditability positive for NKx2-5 mutation, II-ASD and AV block.
Case report: A 36-years-old woman was referred to our attention for fetal echocardiography at the time of her first pregnancy with family history positive for II-ASD, AV block (proband and her mother) and sudden cardiac death (two proband's mother sisters). A NKx2-5 mutation (c 458 del T in exon 2) was found both in the female and in her mother while the fetal echocardiography and genetic analysis (array CGH) were found to be normal. This child is currently 5 years old and without cardiac problem. Recently this woman came back to our attention during her second pregnancy at 17 weeks of gestational age, at fetal echocardiography we found a type Ib tricuspid valve atresia (TA). The array-CGH revealed the same NKx2-5 mutation identified in the family. After extensive counseling the couple opted for termination of pregnancy. The fetus's cardiac abnormality was confirmed by autopsy.
Discussion: Heterozygous NKx2-5 mutations are increasingly recognized having key role as molecular determinant for nonsyndromic CHD with or without progressive AV block. More than 40 heterozygous NKx2-5 germline mutations have been observed in familial, and more rarely, sporadic CHD cases. The most common phenotypes, are II ASD associated with progressive AV block, but other cardiac anomalies have been reported. However, to our knowledge, a correlation between TA and NKx2-5 mutation in a family with ASD, progressive AV block and NKx2-5 mutation has not been documented in literature.
Conclusion: considerable progress has been made towards the understanding of mechanisms leading to CHD. However, the mechanism by which the same cardiac malformations have been exhibited in different gene mutations or the same mutation produce diverse cardiac malformations remains largely to be determined. Identify a new possible genotype phenotype correlation it will be important to better understanding of NKx2.5 mutation role in cardiac development.

P-37
Characteristics of the Atrial Septum in Foetuses with d-Transposition of the Great Arteries
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Objectives: to review the prenatal features of foramen ovale in foetuses with Transposition of the Great Arteries (TGA), focusing on those who needed an urgent balloon atrial septostomy (BAS) for a restrictive interatrial communication at birth.

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Background: also neonates with prenatally diagnosed TGA may die immediately after birth because of the inadequate mixing at the level of the atrial septum.
Methods: we included patients with a prenatal diagnosis of TGA between January 2000 and December 2013. Foetuses with ventricular septal defect were excluded. Clinical data comprised gestational age (GA) at last echocardiogram and at birth, delivery conditions, weight and oxygen saturation at birth. Assessed prenatal echocardiographic data were foramen ovale and septum primum (SP) appearance, ductal and pulmonary veins flow pattern. SP was considered hypermobile if oscillated between both atria, redundant if it herniated more than 50% toward the left atrium, restrictive if was hypo-mobile and thick with a small orifice. For those foetuses with a restrictive foramen ovale, delivery was scheduled in the cardiac unit. Neonates with a restrictive foramen ovale associated to a severe hypoxemia (oxygen saturation ⩽60%) underwent an urgent BAS (within 30 minutes after delivery).
Results: 41 foetuses had a diagnosis of TGA. Last fetal echocardiogram was performed at a median gestational age of 37 weeks. 37/41patients were delivered via caesarean section. Median GA and weight at birth were respectively 39 weeks and 3 Kg. Thirteen patients (32%) required an urgent BAS. Prenatally 6/13 had a restrictive SP appearance and were delivered in the cardiac unit. Among the other 7 fetuses, 6 presented a redundant SP and one a hypermobile SP at last fetal echocardiogram. In these 13 foetuses we did not find any significant abnormal flow pattern in the pulmonary veins or in the ductus arteriosus compared to the rest of the study population.
Conclusions: the antenatal evaluation of the atrial septum for restriction is still challenging. Based on our experience, not only foetuses with a restrictive atrial septum before birth but also those who showed a redundant SP bulging towards the left atrium, are to be considered at risk of an urgent BAS.

P-38
Left ventricular systolic function in preterm and term neonates: Calculation of reference values and of z-score values of the M-mode derived mitral annular plane systolic excursion

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Background: The mitral annular plane systolic excursion (MAPSE) is a quick and reliable echocardiographic tool to assess longitudinal left ventricular (LV) systolic function in children and adults. As this parameter is affected by the LV longitudinal dimension, pediatric and adult normal values are not suitable for preterm and term neonates.
Objective: We determined growth and birth weight related changes of MAPSE to establish normal Z-score values in preterm and term neonates.
Methods: A prospective study was conducted in a group of 261 preterm and term neonates (gestational age: 26/0-6to 40/0-6, birth weight: 670 g to 4140 g).
Results: The MAPSE ranged from a mean of 0.36 cm (Range: 0.26–0.46 cm) in preterm neonates with gestational age of 26/0-6 to 0.56 cm (Range: 0.40–0.73 cm) in term neonates with a gestational age of 40/0-6. MAPSE, gestational age and birth weight are strongly correlated: Pearson's correlation coefficient was 0.56 for gestational age – MAPSE ($p < .001$), and 0.58 for birth weight – MAPSE ($p < .001$). There was no statistically significant difference of normal MAPSE values between females and males ($p = 0.946$).
Conclusions: Absolute values and Z-scores of normal MAPSE values were calculated and percentile charts were established to serve as reference data in preterm and term neonates with congenital heart disease and acquired LV dysfunction (e.g. in neonates with asphyxia). Determination of LV function using MAPSE might be useful in vulnerable infants where a prolonged examination is inappropriate and in neonates with suboptimal visualization of the endocardium.

P-39
Fetal left ventricular non compaction cardiomyopathy and fatal outcome due to deficiency of mitochondrial trifunctional protein

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Introduction: Mitochondrial trifunctional protein (TFP), an enzyme of fatty acid beta-oxidation, is a multienzyme complex composed of four molecules of the alpha-subunit (HADHA) and four molecules of the beta-subunit (HADHB). The most common TFP disorder is LCHAD deficiency caused by the c.1528G > C (p.Glu510Gln) mutation in the HADHA gene. HADHB mutations are relatively rare. We report a fetal case with fatal outcome having a novel HADHB mutation.
Methods: The parents were of Turkish origin, with history of a pregnancy loss due to fetal heart failure and hydrops. In the next pregnancy the fetus developed left ventricular non compaction and increasing pleural effu-sions after 31 gestational weeks. The fetus was small for gestational age and the long bones were short. The baby was born by caesarean section severely asphyxiated at 32 gestational weeks. Medical decision was taken to withdraw intensive care due to failure to thrive and a suspicion of severe mitochondrial disorder. The autopsy was withheld. Consanguinity was present in the parents and paternal grandparents.
Results: Post mortem brain MRI suggested microcephaly with a simplified gyral pattern. The lateral cerebral ventricles were normal. Chromosome analysis was normal (46,XX). Fibroblasts cultured from the skin biopsy of the baby revealed the large homozygous deletion c.1109 + 243_1438-703del in the HADHB gene, and heterozygous mutations were detected in both parents. The deletion has not been reported earlier.
Conclusion: Trifunctional protein deficiency is a relatively rare disorder. Mutations in the HADHB gene cause a systemic disorder with cardiomyopathy originating in fetal life. It is important to differentiate systemic meta-bolic diseases from disorders that affect only the cardiac muscle. Understanding the molecular genetic defect of the patient allows counseling of the family. In this case, the finding has significance for future pregnancies because of a 25% risk of recurrence. The specific diagnosis was made even though autopsy was withheld.

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P-40

Neonatal arterial morphology in fetal growth abnormality

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Background: Fetal growth abnormalities are related with cardiovascular disease later during adulthood. Very little is known about the effect of fetal growth abnormalities on the cardiovascular phenotype during the newborn stage. The objective was to study the regional arterial morphology with respect to gestational age and newborn body morphometrics.
Methods and Results: We studied the arterial morphology of 156 newborns born between 31 and 42 weeks of gestation, including neonates small, large and appropriate for age birth weights, with very-high resolution vascular ultrasound (35-55 MHz). Statistically significant associations were observed between carotid, brachial and femoral arterial lumen dimension (LD), wall thickness (intima-media-adventitia thickness (IMAT), intima-media thickness (IMT)) and end-organ circumference, male gender, gestational age, body weight, and body surface area. In linear multiple regression models these explained a large proportion of the arterial variance (R2 range 0.38 to 0.50 for LD; R2 range 0.25 to 0.41 for IMAT; and R2 range 0.15 to 0.25 for IMT; all models $p < 0.001$). After adjustments, gestational age and male gender remained significant for all arterial LDs ($p < 0.01$).
Conclusion: These preliminary results suggest that fetal arterial growth is primarily related to gender and gestational age.

P-41

Feasibility and precision of transcutaneous very-high resolution ultrasound for the quantification of arterial structures in neonates

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Background: Non-invasive transcutanous very-high resolution vascular ultrasound (VHRU, 25-55 MHz) has recently been developed to quantify vascular structures in adults and children. The performance of the method has yet not been evaluated in neonates. The aim of the study was to assess the feasibility and precision of VHRU in the analysis of superficial arteries in neonates.
Methods: 120 images from central elastic (common carotid, CCA) and peripheral muscular (brachial, BA; femoral, FA) arteries were obtained in 20 neonates of different ages (range 33+0 to 41+5 gestational weeks) and weights (range 1825 to 4950 g). Manual electronical calipers were used to measure lumen dimension (LD) and intima-media thickness (IMT) for all arteries, and intima-media-adventitia thickness (IMAT) for muscular arteries. The intra- and inter-tester variabilities were assessed.
Results: Adequate images were obtained from all CAAs with 35 and all FAs/BAs using 35 and 55 MHz frequencies. IMTs of the smallest BAs and FAs were beyond the axial resolution (<0,05 mm) and, thus, unmeasurable. The intra- and inter-coefficients of variation (CV) were inversely related to the size of the measured dimensions for LDs (range 1.44-2.62 mm, CVs between 1.6–3,2%), IMATs (range 0.141-0.161 mm, CVs between 8.8-19.9%), and IMTs (range 0.062-0.165 mm, CVs between 12.8-24.8%) of the different arteries. No bias between readers was found.
Conclusion: VHRU is feasible and precise in the assessment of superficial arteries in the neonatal population. Neonatal arterial wall layers thicker than axial resolution may be quantified with VHRU.

P-42

Is amiodarone a safe and effective alternative drug in persistent fetal tachycardia?

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Background: Persistant fetal tachycardias, especially when complicated with hydrops, are associated with a poor prognosis. If digoxin and flecaine are the usual first–line treatment but with inconstant results, amiodarone remains a last choice treatment because of its known complications.
Aims: In this retrospective study, we reviewed the use of amiodarone in patients with resistant fetal tachycardia, to determine the safety of this drug and its efficiency.
Methods: Between 1986 and 2012, 16 pregnancies admitted for fetal tachycardia were treated with amiodarone. 4 had atrial flutter and 12 had supra-ventricular tachycardia. The presentation was severe: 10 fetuses (63%) had hydrops and 2 (12.5%) had an isolated pericardial effusion. Amiodarone was never used as a first line therapy, but as a second line therapy in 6 fetuses and as a third line therapy in 6 fetuses.
Results: Amiodarone was given during 28.2 ± 15.3 days and was effective in 10 of the 16 (63%) patients (defined as resolution to sinus rhythm or slowing the tachycardia under 190 bpm), and despite the presence of hydrops (efficiency was obtained in 4/6 fetuses of the non hydropic group versus 6/10 of the hydrops group, p = NS). Among mothers, 2 complications were noticed: mild hypothyroidism and hepatic cytolysis. Hypothyroidism was present in 3 patients who did not require any substitutive treatment. Hepatic cytolysis was also present in 3 patients but never above 3 times normal level: therefore, none of them required amiodarone interruption. Fetuses were born at 35.8 ± 3.2 WA, weighed 2805 ± 579g, and 5 of them required oral intubation at birth. One fetal death occurred (sinusal rhythm was obtained but hydrops with ventricular dysfunction persisted and a ventricular thrombus appeared). Hypothyroidism was present in 6 patients (including one goiter): 3 had transient hypothyroidism that resolved in 2 weeks, 2 were treated for 6 months and one is still treated. All children had normal neurological development.
Conclusion: Persistant tachycardias complicated with hydrops remain a medical challenge. Amiodarone seems to be a safe and efficient alternative drug in this indication.

P-43

Gorlin syndrome: a familial diagnosis

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Gorlin syndrome (or nevoid basal cell carcinoma syndrome) is characterized by a wide range of developmental abnormalities with prevalence from 1/57.000 to 1/256.000, a male-to-female ratio of 1:1. It is caused by mutations in the PTCH1 gene and is transmitted as an autosomal dominant trait with complete

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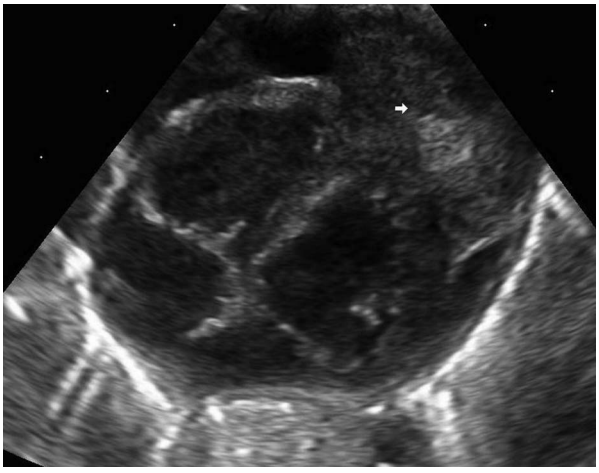
penetrance and variable expressivity. This case report describes a familial Gorlin syndrome discovered in utero by a cardiac tumour. A cardiac tumour localized at the apex, septum and lateral wall of the left ventricular was discovered on antenatal echocardiography in a girl, associated with ventricular septal defect and a cerebral ventricular dilatation. A Gorlin syndrome was suspected but no genetic study was realized. In the familial history, she had a sister who presented a syndrome involving atrial septal defect, microcephaly, psychomotor retardation and epilepsy. She died accidentally at 4 years of age but no genetic study for Gorlin syndrome was realized. Our patient was born at 37 weeks without hemodynamic or respiratory disorders. The postnatal echocardiography confirmed the cardiac tumour at apex of the left ventricular and the muscular ventricular septal defect, associated with other anomaly (facial dysmorphism, talus valgus and moderate cerebral ventricular dilatation). A thoracic MRI showed the cardiac tumour in the apex, septum and the free wall of the left ventricular without hemodynamic obstruction. A biopsy was realized showing a fibroma on histology as usual in this syndrome. The genetic test was confirmed the Gorlin syndrome in this girl, and the same mutation was observed in her mother and her grandmother. The one year follow up showed no evolution of the tumour without hemodynamic disorder, but few briefs episodes of sinus tachycardia.

The combination of cardiac tumour and cerebral ventricular dilatation in utero should evoke the diagnosis of Gorlin Syndrome that could be confirmed by the appropriate genetic test.



Echocardiography of tumour

P-44

Assessment of Cardiac Axis In Fetuses With Conotruncal Abnormalities And 22q11.2 Microdeletion Using Spatiotemporal Image Correlation Volumes

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Objective: To evaluate the association between the fetal cardiac axis and conotruncal abnormalities (CTA), with and without 22q11.2 deletion, and the thymic-thoracic ratio (TT- ratio)

Methods: Database records were reviewed for cases of fetal conotruncal and arch abnormalities with a known karyotype between January 2007 and September 2013. Cardiac axis (the angle between the midline of the thorax and the interventricular septum in a four-chamber view) and TT-ratio (as previously described) were measured retrospectively in 81 fetuses with CTA using stored 3-dimensional spatiotemporal image correlation volume datasets independently by two authors without knowledge of the karyotype. Inter-observer and intra-observer reproducibility was assessed on a random sample of 20% of fetuses.

The results were compared to a control group of 55 normal fetuses.

Results: The median gestation in the control and CTA groups was 20 and 23 weeks, respectively. Of the 81 cases of CTA, 18 had 22q deletions. The mean cardiac angle in controls was 43.8 degrees (±7.2); in CTA with normal karyotype, it was 58.1 degrees (±13.06) and in CTA with 22q11.2 microdeletion, 67.8 degrees (±14.2). Using one-way ANOVA, this increase in angle is significant ($p < 0.0001$). There was displacement of the angle of the septum to the left in 88% of fetuses with conotruncal abnormalities. The sensitivity of a cardiac axis greater than 60 degrees in diagnosing conotruncal abnormalities was 55% and the specificity was 100%. There was no linear correlation between the cardiac angle and TT-ratio. Inter- and intra-observer variability was 4% and 3% respectively.

Conclusion: Reliability of measurement of the cardiac axis is good. Leftward displacement was most commonly seen in fetuses with 22q11.2 microdeletion. There is overlap of the angle between those fetuses with the deletion and those without, so this cannot be used in isolation as an indicator for which patients to offer cytogenetic analysis for Di George syndrome. Our data does not show a close relationship between the cardiac axis and the TT-ratio. Our data suggests that an abnormal septal axis in the fetus, a characteristic which is readily assessed during routine obstetric ultrasound assessment, is an indication for specialist fetal echocardiography.

P-45

Common origin of the innominate and left carotid artery in prenatally suspected coarctation of the aorta

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Objective: In neonates with prenatal suspicion of coarctation of the aorta, we have noted a frequent occurrence of common origin of the innominate artery and left common carotid artery (LCCA). Our aim was to establish whether branching pattern influenced the diameter of the distal aortic arch.

Methods: The last 37 cases of prenatally suspected coarctation diagnosed at our institution were reviewed. Neonatal echocardiograms performed within 24 hours of birth were analysed. For the purposes of this study, common origin included neonates with a true common origin and those with less than 1millimetre between the origins of the innominate artery and LCCA. The arch diameter immediately distal to the LCCA, arch diameter proximal to the left subclavian artery (LSCA) and isthmus at end-systole were measured. Measurements were taken blinded to the outcome. Student's t test, Fisher's exact test and one-way ANOVA were used for analyses.

Results: 14/37 neonates developed neonatal coarctation requiring surgical repair. Common origin occurred in 6/14 (43%) of those neonates who developed coarctation and 13/23 (57%) of those who did not develop coarctation ($p = 0.508$). Neonatal weight was not significantly different amongst these four groups ($p = 0.811$)

With a normal branching pattern the pre-LSCA arch diameter was wider compared to the pre-LSCA arch diameter in neonates with common origin (mean ± SD: 4.5 mm ± 0.8; 3.3 mm ± 0.8; $p = 0.003$). Fig. 1 shows the distribution of pre-LSCA diameters.

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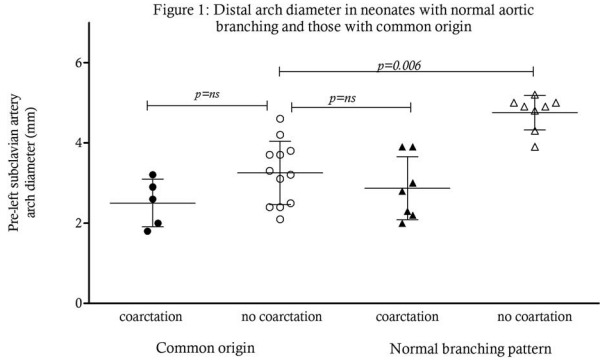
The neonates with common origin who did not develop coarctation had similar arch diameters to those with a normal branching pattern who did develop coarctation: post-LCCA arch diameter ($p = 0.347$), pre-LSCA diameter ($p = 0.331$) and isthmus ($p = 0.117$).

Figure 1: Distal arch diameter in neonates with normal aortic branching and those with common origin



In those neonates who do not develop coarctation, the post-LCCA diameter, pre-LSCA diameter and isthmus are significantly smaller in common origin compared to normal origin ($p = 0.002$, $p = 0.030$, $p = 0.012$, respectively).

Conclusions: In neonates with common origin and no coarctation, the arch measurements were a similar size to those neonates with normal branching pattern with coarctation. Common origin may be a reason for the high false positive rate of prenatal diagnosis. Therefore, we recommend ascertainment of the branching pattern of the aortic arch when disproportion of the transverse and ductal arches is seen in the fetus.

P-46

Antenatal echocardiographic parameters to predict postnatal outcome of neonates with Ebstein anomaly

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Ebstein tricuspide valve anomaly is a rare CHD with uncertain postnatal prognosis. Criteria to predict outcome are still a matter of debate. The aim of this study was to determine antenatal echocardiographic predictive parameters.

Methods: Retrospective multicentric analysis of fetus with diagnosis of Ebstein anomaly. Echocardiographic measurements of ventricles, atria, great vessels and tricuspid regurgitation were collected. Comparisons were made between group I (poor outcome = death occurred in utero or within the first 3 months of life) and group II (favourable outcome: postnatal survival >3 months).

Results: 16 fetuses were included in the study: 10 in group I (62.5%: 2 TOP, 2 fetal deaths, 6 postnatal deaths) and 6 in group II (37.5%). Mean gestationnal age at diagnosis was 29weeks (22 to 38). The mean number of echocardiographic records per patient was 2 (1 to 6). LV to RV ratio, tricuspid valve regurgitation grade and retrograde or anterograde ductal flow did not differ between the 2 groups. Significative differences were found between groups I and II regarding the presence of pulmonary flow (none or mild RV to PA flow: 8 of 9 cases died = 89%), AO to PA ratio (75% death if >97°p vs 25% if 3-97°p), RA diameter (77.3% death if >97°p vs 0%), PA diameter (100% death if <3°p) and pericardial effusion (80% death vs 0%). Only 1 case had arrhythmia and died.

Conclusion: this small sample size study showed that the absence of RV to PA flow and/ or pulmonary valve opening, increased AO to PA ratio, RA and decreased PA diameter and the presence of pericardial effusion might represent prognosis factors in fetus with Ebstein anomaly. These results should be confirmed by large scale prospective study.

P-47

Extracardiac or chromosomal anomalies strongly influence parental treatment decision and postnatal survival of neonates with prenatally diagnosed congenital heart diseases

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Objectives: This study was design to assess the influence of extracardiac or chromosomal anomalies on parental decision of termination of pregnancy and on survival rates in newborns with prenatally diagnosed congenital heart diseases.

Methods and results: 2057 consecutive foetuses with congenital heart disease diagnosed from January 2002 to December 2011 were included: 1258 (61%) in-born neonates and 799 (39%) terminations of pregnancy (TOP). The overall prevalence of major extracardiac or chromosomal anomalies was 18,6%. Of the 1258 newborns, 121 had a major associated anomaly but only 55 were identified before birth. Prenatally identified associated anomalies were significantly lower in the newborn group in comparison with the TOP group (4% vs 31%, $p < 0,0001$). They were also lower in the surviving group at one year of follow up (7,5% vs 20,7%, $p < 0,0001$). A 4-fold increase of death rate was observed if an associated anomaly was identified (IC95%[2,5-6,7], $p < 0,0001$). These associations remained significant after multiple logistic regression analysis including the severity of the heart defect (univentricular or biventricular physiology).

Conclusion: Women are more likely to terminate pregnancy if extracardiac or chromosomal anomalies are associated. Post natal survival is strongly influenced by these associated anomalies.

P-48

Prenatal detection of congenital heart defects after in vitro fertilization

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Objectives: Increasing number of children born by assisted reproductive techniques (ART) is associated with higher risk for unfavourable birth outcome. ART is related to preterm delivery, intrauterine growth retardation and based on recent studies to higher birth defects including CHD. The aim of the study was to determine the incidence of CHD in foetuses after in vitro fertilization (IVF).

Methods: All pregnant women undergo prenatal ultrasound examination in the Czech Republic. In case of any suspected or proven heart abnormality, the foetus is referred for specialized centralized prenatal echocardiography. There is not any special screening programme and follow-up for pregnancies after IVF, thus CHD detection rate after IVF should not be biased.

Results: From 469 671 children born between 2007 and 2010, 8226 (1.75%) were conceived using IVF. CHD was more frequently prenatally detected in IVF+ than IVF- pregnancies: 41/8226 (0.50%) vs. 881/461445 (0.19%). Pregnant women with

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IVF had higher rate of twins (22.0% vs. 1.5%, $P < 0.001$) and were older (22% vs. 8% >35 years, $P < 0.001$). Due to pre-implantation diagnosis, IVF+ foetuses with CHD were less frequently affected by chromosomal aberrations than those conceived spontaneously: 2/41 (4.9%) vs. 140/881 (15.9%), $P < 0.001$. The incidence of non-cardiac congenital defects was, however, similar: 10/41 (25%) vs. 238/881 (27%), P NS. In case of an isolated CHD the parents of IVF conceived foetuses opted for termination of pregnancy less often than in IVF- pregnancies (1/31, 3.2% vs. 202/643, 31%, $P < 0.001$).
Conclusions: In vitro fertilization is associated with 2.5- times higher rate of congenital heart defects detected by prenatal screening. Pregnancy termination for an isolated CHD is less common than after spontaneous conception.
Supported by MH CZ – DRO, University Hospital Motol, Prague, Czech Republic 00064203

P-49
Reference ranges and Z scores of functional parameters in fetal echocardiography

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Background: Fetal echocardiography is the standard-of-care in a variety of obstetrical cardiovascular circumstances. Reference values for basic fetal heart anatomic measurements are available. However, studies that propose reference values for functional parameters (doppler velocities and patterns, m-mode, cardiac output and shortening fraction) are scarce and their quality is often questionable. Furthermore, most existing reference values are not reported as Z-score equations. Improved definition of reference values with adequate statistical validation is needed for proper interpretation of these measurements in clinical settings.
Objective and methods: We aimed to propose robust reference values for fetal functional cardiac parameters. Singleton uncomplicated pregnancies with normal fetal heart were selected. When fetal position and echogenicity allowed it, a set of 62 functional measurements was performed (44 pulsed doppler related measurements and 18 M-mode measurements – see the Table). Several parametric regressions were tested to model each measurement against gestational age (GA). Variation around the predicted mean was also modeled. Z-score equations were computed and the proposed reference values were tested for residual association, residual heteroscedasticity, and departure from the theoretical normal distribution.
Results: Data from 104 fetal echocardiograms were analyzed. Parametric normalization was successful for most measurements analyzed. We were able to compute Z-score equations with minimal residual association with GA, no residual heteroscedasticity and insignificant departure from the normal distribution.
Conclusion: This study allowed computation of Z-scores for several anatomical and functional measurements, most of which did not have any published reference values. These Z-scores equations will allow echocardiographists to more accurately identify measurements that diverge from normal and thus better detect potential alterations in fetal heart function.

Table – list of selected fetal echo measurements

Cardiac output	
	Left cardiac output
	Right cardiac output
	Right/left cardiac output ratio
	Effective pulmonary flow
	Ductus arteriosus flow
	Ductus venosus flow
Peak velocity and time velocity integral	
	Ascending aorta
	Pulmonary artery
	Ductus arteriosus
Inflow dopplers and time velocity integrals	
	Mitral valve E and A waves
	Mitral valve E/A ratio
	Tricuspid valve E and A waves
	Tricuspid E/A ratio
Venous flow doppler and time velocity integrals	
	Inferior vena cava systolic, diastolic and a waves
	Ductus venosus systolic, diastolic and a wave
Other doppler flows and calculations	
	Systemic ve_GoBack_GoBack
	Isthmus systolic ante/retrograde flows
	Isthmic ratios
	Left ventricular myocardial performance index
M-mode short axis and 4 chambers	
	LV and RV end-diastolic dimensions
	LV and RV end-systolic dimensions
	LV and RV shortening fraction
	Wall thicknesses

P-50
The Effects of Pre-pregnancy Obesity on Fetal Cardiac Functions

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Introduction: Obesity is a substantial public health problem with the prevalence increasing rapidly in numerous industrialized nations. The objective of this study was to evaluate the effects of maternal pre-pregnancy obesity on fetal cardiac functions.
Methods: We studied 55 fetuses of obese mothers and 44 fetuses of healthy mothers at 26–38 weeks of gestation. Cardiac functions were evaluated by M-mode, pulsed-wave, and tissue Doppler echocardiography.
Results: The two groups were similar in terms of maternal age, gravidity, parity, gestational age, estimated birth weight, serum lipids and systolic- diastolic blood pressure. Fetal heart rate, diameters of the aortic and pulmonary valve annulus, aortic and pulmonary peak systolic velocities, ventricular systolic function and cardiothoracic ratio were similar in the two groups. Pulsed-wave Doppler-derived E/A ratios in the mitral and tricuspid valves were similar in the two groups. The deceleration time of early mitral inflow was prolonged in the fetuses of the obese mothers. In the interventricular septum, left ventricle posterior wall, and right ventricle free wall the Ea and Aa were higher, and Ea/Aa ratios were significantly lower in the study group than control group. The E/Ea ratio was higher in the obese group than in the control group. The isovolumic relaxation time and the right and left ventricle myocardial performance indices were higher in the fetuses of the obese mothers than in the fetuses of the healthy mothers.

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Conclusions: We believe that maternal obesity has an important influence on fetal cardiac diastolic functions.

P-51

Assessment of Microvolt T-wave Alternans in Repaired Tetralogy of Fallot Patients with 24-hour Holter Electrocardiography

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Objective: In this retrospective study, we aimed to examine microvolt T-wave alternans (MTWA) in Holter electrocardiography (ECG) of children with repaired tetralogy of Fallot (TOF) to assess risk of sudden death and ventricular arrythmias. We tried to define correlation between MTWA and electrocardiographic, echocardiographic parameters, heart rate variability (HRV) as well.
Methods: Holter ECG records and archive files of 56 patients with repaired TOF were analyzed. Subjects' ECG parameters, HRV and MTWA values were compared with a control group with similar age and gender distribution.
Results: Thirty-five subjects (%62,5) were male and 21 (%37,5) were female. Mean age was 123,4 ± 48,3 months. All patients underwent total correction operation and eight (%14,3) patients had Blalock-Taussig shunt procedure before total correction. There was significant difference between patient and control groups, in terms of QRS duration, median QT dispersion, maximum and minimum mean duration of QT and QTc. We found RBBB in 37 patients (%66), monomorphic premature ventricular contractions in 8 (%14,3) and non-sustained ventricular tachycardia in 1 (%1,8). All HRV parameters except LF/HF ratio were found to be significantly lower in patient group than in control group. LF/HF ratio was significantly higher in patient group. Median MTWA value was 55,5 μV in the control group whereas 95,5 μV in patients group. MTWA was found to be significantly higher in patient group. A significant positive correlation was found between presence of premature ventricular contractions and tricuspid regurgitation; SDNN, SDNNi, TP, VLF and RV dilatation; SDNNi, TP, VLF and QRS duration; SDNN, SDNNi, rMSSD, pNN50, TP, VLF, LF, HF and maximum QT; SDNNi, rMSSD, pNN50, TP, VLF, LF, HF and minimum QT; SDNNi, TP, VLF, LF and QT dispersion. There was no correlation between HRV parameters, echocardiographic findings and MTWA.
Conclusions: HRV was reduced and MTWA was increased in children with repaired TOF. HRV and MTWA are significant markers of ventricular arrhytmias and sudden death. Predictions about future RV diameters, functions, likelyhood of ventricular arrhythmias and sudden death may be made by evaluating HRV parameters and MTWA, and thus need for closer follow-up can be defined.

P-52

Dispersion durations of P-wave and QT interval in children treated with ketogenic diet

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Introduction: The ketogenic diet, a high-fat, adequate-protein and low-carbohydrate diet, offers new hope to children with intractable epilepsy. Known adverse effects include anorexia, constipation, renal stones and dyslipidemias. Cardiac complications, including prolongation of the QT interval and sudden cardiac death (SCD) have also been reported in patients treated with ketogenic diet. The association between ketotic conditions and prolonged QTc and/or SCD raises the suggestion of whether ketosis may directly affect cardiac repolarization and be a cause of arrhythmia and/or SCD in diabetic ketoacidosis patients and ketogenic diet patients. There is very limited data for the effects of ketogenic diet on QTD and PWD measures. Therefore we aimed to search for changes in the QTc interval, QTD and PWD with serial electrocardiograms (ECGs) in patients treated with ketogenic diet at our center.
Methods: Twenty-three drug resistant epileptic patients treated with ketogenic diet were enrolled in this study. Electrocardiography was performed in all patients before the beginning and after the sixth month of ketogenic diet. Heart rate, maximum and minimum P-wave duration, P-wave dispersion, maximum and minimum QTc duration and QT dispersion were manually measured from the 12-lead surface ECGs.
All children were started on a standardized 3:1 ketogenic diet with a non-fasting gradual initiation protocol.
Results: A total of 23 patients (13 male and 10 female) with median age of 51 months ranging from 13 to 158 months were included in the study. Electrocardiographic measurements before the beginning of ketogenic diet and after sixth month of treatment are presented in Table 1. Minimum and maximum QTc and QTD measurements showed non-significant increases at 6^{th} month when compared to baseline values. Other previously mentioned ECG parameters showed no significant changes (Table 1).
Conclusion: A six-month long ketogenic diet has no effect on PWD in children with intractable epilepsy. Further studies with larger samples are needed to clarify the effects of ketogenic diet on QTc and QTD.

Table 1. Electrocardiographic measurements of the patients

Variables	Baseline	6^{th} month	p value
Mean HR, bpm	112 ± 25	115 ± 28	0.521
P max, ms	96 ± 14	95 ± 20	0.768
P min, ms	47 ± 11	46 ± 9	0.716
PWD, ms	48 ± 9	51 ± 12	0.242
QTc max, ms	438 ± 26	455 ± 41	0.053
QTc min, ms	381 ± 20	389 ± 28	0.209
QTD, ms	43 ± 16	47 ± 18	0.382

P-53

The short-term effects of ketogenic diet on carotid intima media thickness and elastic properties of the carotid artery and the aorta in epileptic children

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Objective: Currently no data are available for the effects of ketogenic diet on the development of atherosclerosis and/or cardiovascular disease. The aim of this prospective study is to investigate the effect of a 6-month-long ketogenic diet on carotid intima-media thickness, carotid artery, and aortic vascular functions.
Methods: Twenty-three drug resistant epileptic patients who were treated with ketogenic diet at the pediatric neurology clinic were enrolled in this prospective study. Fasting total cholesterol, high-density lipoprotein (HDL), low-density lipoprotein (LDL), triglycerides, total cholesterol and glucose concentrations were measured and echocardiography was performed in all patients before the beginning of ketogenic diet and after the sixth month of treatment. Carotid intima-media thickness and aortic/carotid diameter at systole and diastole were measured after conventional echocardiographic examination.
Results: The body weight, height, serum levels of triglyceride, total cholesterol and LDL increased significantly at month 6 when compared to baseline values. Systolic and diastolic diameters of the carotid artery and systolic diameter of aorta increased significantly at month 6 when compared to baseline measures ($p < 0.05$). Carotid intima media thickness, elastic properties of the aorta and carotid artery did not change at the sixth months of therapy compared to baseline values.
Conclusions: A six-month long ketogenic diet has no effect on carotid intima-media thickness and elastic properties of the carotid artery and the aorta. Further studies with larger samples and longer follow-up periods are needed to clarify the effects of ketogenic diet on carotid intima-media thickness and elastic properties of the carotid artery and the aorta.

P-54

Evaluation of cardiac functions and myocardial performance index in children with iron deficiency and iron deficiency anemia

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Background: The aim of this study was to evaluate the possible effects of iron deficiency (ID) and iron deficiency anemia (IDA) on myocardial functions.
Materials and Methods: Thirty children with ID and thirty children with IDA were included along with 30 age and sex matched control subjects. Cardiac functions were evaluated echocardiographically with Motion-mode (M-mode), pulsed wave (PW) Doppler and PW tissue Doppler methods.
Results: Left ventricular systolic functions showed no statistically significant differences between groups before and after therapy. Significantly higher values of right and left ventricle MPI were found in ID and IDA groups before therapy. After treatment there was no significant difference in left ventricular values except E/A ratio. However, the right ventricular parameters between ID, IDA and control groups revealed that there were statistically significant differences for E flow velocity, E/A ratio, MPI and modified MPI values both before and after therapy.
Conclusion: MPI and diastolic functional changes are better indicators of myocardial dysfunction due to ID and IDA. Right cardiac chambers are affected by ID and IDA more than left ones and it likely takes more time for right chambers to recover after iron therapy.

P-55

Risk Factors for Out-of-Range International Normalized Ratio in Paediatric Patients Receiving Warfarin

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Introduction: Warfarin has a narrow therapeutic window; therefore, out-of range international normalized ratio (INR) often occurred in paediatric population. We aim to identify risk factors for out-of-range INR in paediatric patients receiving warfarin.
Methods: Study population was recruited from a database of 823 consecutive INR measurements (24 patients) which were performed for dose optimization of warfarin between 2004 and 2012. INR measurements which were performed <1 week after initiation of warfarin, those from patients who used combined medicines having an interaction with warfarin except aspirin and heparin and those from patients with liver dysfunction were excluded from the study. No patients experienced pharmacogenomics of warfarin. Age, gender, body weight, warfarin dose, Fontan physiology as an indication of anticoagulation and complications during anticoagulation were analysed. INR values were divided into 3 groups; optimal anticoagulation (INR from 1.5 to 3.0), under-anticoagulation (INR <1.5) and over-anticoagulation (INR > 3.0).
Results: A total of 576 measurements were selected for additional study. Age, female gender, body weight, warfarin dose, INR values and Fontan physiology were 7 ± 3 years, 77%, 20 ± 8 kg, 1.86 ± 0.04 mg, 1.83 ± 0.54 and 82%, respectively. There were neither thrombotic nor haemorrhagic events during the period. Out-of-range INR was observed in 33% of measurements; under- and over-anticoagulation was observed in 29% and 4%, respectively. The under-anticoagulation group comprised significantly more female patients and those with Fontan physiology compared with the optimal and over-anticoagulation groups (86% vs. 74% vs. 64% and 93% vs. 77% vs. 82%, respectively; both $p < 0.003$). In contrast, the over-anticoagulation group showed significantly lower age and lower body weight compared with the optimal and under-anticoagulation groups (5 ± 2 vs. 7 ± 3 vs. 7 ± 3 years and 13 ± 2 vs. 19 ± 8 vs. 20 ± 8 kg, respectively; both $p < 0.03$).
Conclusions: Out-of-range INR often occurred in paediatric patients receiving warfarin. Female, Fontan physiology, lower age and lower body weight were the risk factors for out-of-range INR.

P-56

QTc and QTd changes after Cardiopulmonary Bypass Surgery in Children

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Objectives: The aim of the study was to assess the impact of cardiopulmonary bypass surgery on corrected QT (QTc) and QT dispersion (QTd) intervals. The possible role of inflammation on these variables was investigated.
Background: Systemic inflammation and altered myocardial repolarization are common consequences of cardiopulmonary bypass surgery.
Methods: Electrocardiograms (ECGs) were registered and C-reactive protein (CRP) as well as white blood cell (WBC) count were measured in 36 children with ventricular septal defect

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(VSD) or atrial septal defect (ASD) one day before and 5 days after surgery. QTc and QTd were calculated.
Results: QTc increased after surgery in 24 (67%) patients (mean ± SD = 31 ± 25 ms, range = 7 to 125); whereas QTc decrease was noted in 8 (22%) patients (28 ± 28 ms, range = 1 to 77). After surgery, QTc was abnormally (increments in the QTc above the upper limit of normal) prolonged in 8 (22%) patients (461 ± 18 ms, range = 445 to 487). Only one of these 8 patients had abnormally prolonged QTc before surgery.; Abnormally prolonged QTc returned to normal in 3 of the 4 patients with pre-operatively prolonged QTc. A trend for increased QTd was also noted. The changes did not correlate with CRP, WBC count, bypass time or aortic cross-clamp time.
Conclusions: Twenty-four (67%) of the 36 patients had a post-operative increase in QTc; only 8 of these 24 patients had impaired myocardial repolarization (increments in the QTc above the upper limit of normal). Similarly, 8 (22%) of the 36 patients had a post-operative decrease in QTc. CRP and WBC had no clear correlations with the post-operative ECG changes. Although post-operative QTc changes were common, their natural history and clinical significance remain uncertain.

P-57
Medium-term Follow up of Patients with Vasovagal Syncope
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Medium-term Follow up of Patients with Vasovagal Syncope
Objective: In this study, we aimed to evaluate 3-year mid-term follow-up results that consist of treatment response, frequency of recurrences, predisposing factors that cause recurrence and prognosis in patients with syncope and presyncope. We also aimed to obtain useful information that will contribute to follow-up and treatment of these patients.
Materials and Methods: 285 consecutive patients diagnosed with syncope between January 2008 and December 2009 were enrolled into study. Patients files were rewieved retrospectively. Etiology of syncope in these patients were evaluated. 114 patients were diagnosed with vasovagal syncope. Reccurrences and related conditions were recorded.
Results: Vasovagal syncope 51.2%, pseudo-syncope 22.1%, syncope with neurological origin 14.4%, cardiac syncope 2,1% and unexplained syncope 10.2% were found in 285 patients that were examined for syncope. 114 patients diagnosed with vasovagal syncope. Of those 58.8 female %, 41.2% were male. Female to male ratio was 1,4. Mean age at presentation 11.7 ± 2.5 years, the mean age of first syncope 11.3 ± 5.2 years, and the mean duration of follow-up 42.8 ± 3.4 months were found. During the follow-up period recurrence of syncope was found in 31 patients (27.2%). Any effects of sex, absence or persence of prodromal symptoms, response type to tilt table test at the time of diagnosis was not determined on the reccurence and type of syncope. Significant decrease was seen in the number of syncope attacks after the tilt table test. The recurrence of syncope was observed in 15 of 26 patients that were treated with beta blockers, whereas 16 of 88 untreated patients. Reccurence of syncope were found more frequent in patients with drug therapy.
Discussion: In childhood period, education and recommendations, increasing fluid and salt intake are successful form of treatment for vasovagal syncope. In patients with recurrent syncope attacks β blocker treatment is used besides the recommendations. But effect of the beta blocker therapy for preventing the recurrence is limited. For this reason, medication should be used only in cases with frequently repeated vasovagal syncope attacks.

P-58
The anxiety level, the awareness and the expectations from pediatric cardiologist of parents whose children diagnosed with cardiac murmur during general physical examination
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Objectives: To determine the anxiety levels, the awareness of application to pediatric cardiology clinics and the expectations from pediatric cardiology doctors, of parents whose children have cardiac murmur.
Methods: The children with cardiac murmurs diagnosed during general physical examinations and appliying to pediatric cardiology clinics at first time were included. Both pathological and innocent murmurs were taken. The questionnaire forms consisting of 7 questions were answered by 354 families appliying to pediatric cardiology clinic between May-August 2013.
Results: The anxiety level of parents were 8 out of 10 points when they learned that their children had cardiac murmur. While the parents' awareness of a department dealing with pediatric heart disease was 60.7%, when their expectations were asked; 8.4% of the parents said that cardiac examination would be enough while 82.2% thought that echocardiography should be performed. It was determined that 59% of the parents applied to pediatric cardiology clinic within a day and 29.6% applied within a week. According to 79.9% of the parents the murmur was a sign of a serious heart disease; 51.4% of them researched and obtained information about murmur, and 55.4% of them used internet for that purpose.
Conclusions: The cardiac murmurs cause significant level of anxiety to parents even though they are mostly innocent. The parents' of children with cardiac murmur, who are not informed by a health care provider working at primary or secondary care health centers, typically visit a pediatric cardiology clinic and ask for echocardiographical examination.
Keywords: Children, parent, cardiac murmur, anxiety, expectation.

P-59
The Relation of IVIG Resistance, Laboratory Findings and Coronary Complications in Patients with Kawasaki Disease
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Objectives: The aim of this study is to evaluate the cases diagnosed with KD in our hospital.
Methods: We retrospectively analyzed the medical records of 31 patients, ranging from 3 months to 7 years of age who were admitted to our hospital between January 2007-June 2013 and diagnosed as KD according to EULAR/PrES criteria. The epidemiological, clinical, laboratory and echocardiographic findings; treatment and follow-up results of the cases were evaluated.

Results: The male/female ratio was 1,81. Median age was 24 months and 90,3% of cases were under 5 years of age. The median duration of illness at diagnosis was 7 (5-21) days. 22,6% of cases were diagnosed at subacute stage. 54,8% of cases were incomplete KD. 41,2% of incomplete cases were diagnosed after 10 days ($p = 0,009$). Conjunctivitis, oropharyngeal changes, changes in extremities and rash were significantly more common in classical cases ($p < 0,05$). Coronary artery ectasia or aneurysm were identified in 8 cases (25,8%), all of these cases were male ($p = 0,028$). 20% of cases received IVIG after 10 days of ilness and coronary artery involvement was 57,1% in these cases ($p = 0,05$). IVIG resistance was seen in 23,3% of cases and these cases received second dose of IVIG therapy. High C-reactive protein levels ($p = 0,017$), high platelet levels ($p = 0.043$) and low albumin levels ($p = 0,014$) were related to IVIG resistance. The coronary artery ectasias resolved in all cases. In one of the cases with multiple giant aneurysms, the aneurysms regressed but the other case resulted in thrombosis, obstruction and finally death.
Conclusions: There is relation between IVIG resistance and laboratory values. So, supporting the clinical suspicion with laboratory and echocardiographic findings to diagnose and treat especially the incomplete cases earlier is important in preventing the cardiovascular complications of this disease.
Keywords: IVIG, incomplete, Kawasaki disease, coronary artery aneurysm, resistance

P-60

Intracardiac thrombus in children: We are in need of and must discuss new algorithms

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Objectives: The aim of this study was to determine the risk factors and outcomes of patients with intracardiac thrombus.
Methods: The medical records of patients with intracardiac thrombus between June 2010 and December 2013 were searched thoroughly. The size, location and outcome of thrombus were assessed retrospectively. The patients given the diagnosis of infective endocarditis later on according to the cilinical and laboratory findings were excluded from the study.
Results: 16 patients were enrolled in this study. The median age of patients was 2,2 (2 days-14,1) years. Six patients were newborn and 2 patients were infant. The median size of thrombus was 9 (5-21) mm. The localization was right atrium in 7, right ventricle in 5, left ventricle in 1, pulmonary artery in 1 and superior vena cava in 2 patients. There was prematurity in 5, siyanotic congenital heart disease in 1, blood culture positivity in 3, malignancy in 4, nephrotic syndrome in 1, indwelling (umblical or central venous) catheters in 10 and acquired or genetic thrombophilia (protein C defficiency, factor V Leiden, MTHFR A1298C and MTHFR C677T mutations) in 6 patients as risk factors. The first choice was tissue plasminogen activator in 2 patients because of the risk of embolization, classical heparin in 1 patient because of the acute nature of the thrombus, parenteral antimicrobials in 1 patient because of isolated bacteremia and low molecular weight heparin in remaining 12 patients. In 9 patients therapy included parenteral antimicrobials together with anticoagulants because systemic infection. The result was complete resolution in 15 patients and in 1 patient thrombus was surgically removed during corrective surgery for tetralogy of Fallot. The median time was 16 (2-70) days for 50% resolution and 26 (3-93) days for complete resolution. There was no anticoagulant therapy related major complication.
Conclusions: In patients with risk factors, cardiac thrombosis should be kept in mind. In patients with intracardiac thrombus, selection of anticoagulant therapy may decrease the risk of complications. Surgery is rarely required and thrombolytics are not usually necessary for resolution of thrombus.
Keywords: Intracardiac thrombus, children, risk factors, treatment, new, algorithm

P-61

Naproxen treatment in patients with Acute Rheumatic Fever

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Objectives: To evaluate the effectiveness and side effects of Naproxen in patients diagnosed with acute rheumatic fever (ARF). The freguency of rheumatic valvulitis in the patients who were given Naproxen, was also evaluated at the long term follow up.
Methods: We reviewed clinical, laboratory and therapeutic data of 167 patients who diagnosed with ARF according to revised Jones criterions between 2009-2013 years. 96 cases who had presented with acute arthritis, were included in this study. Patients were classified into two groups; Group1: consisted of 50 patients who treated with Naproxen (15 mg/kg/d)), Group2: consisted of 46 cases who treated with Aspirin (80-100 mg/kg/d). All patients have been followed with echocardiography for a mean time of 2.8 +/- 0.3 years (ranged: 2-4 years). The study protocol was approved by our hospital Ethic Committee.
Results: The mean age of patients was 10.7 years. Four cases in both groups had past history of ARF. Mild degree valvular regurgitation was detected in 40 cases (80%) in groups 1 and 36 cases (78%) in group 2 at the initial examinatian. High grade fever was detected in 27 cases (28%) in all study population. After medication started, fever resolved quickly in both groups (median: 1 day, ranged 1 to 3 days). The clinical activity resolved completely within a few days. The median time to normalization of arthritis was 2 days in both groups. Erythrocyte sedimentation rate normalized at the median of 10 days in both groups (ranged: 3-14 days). Except one case, no side effect was observed in Group 1. The treatment with Aspirin was discontinued in eight cases (17.4%), due to elevated hepatic enzymes levels. Treatment failure was not observed in both groups. None of the patients had developed new valvular regurgitation during follow-up. The rheumatic valvular involvement persisted in 26 cases (52%) in group 1 and 25 cases (54.3%) in group 2, respectively.
Conclusion: Pediatrician can use Naproxen in patients with ARF, as a safe and effective alternative drug option.

P-62

Increased P wave and QT dispersion in children with Down Syndrome without Congenital Heart Disease

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Objective: Previously reported studies have shown that patients with DS without concomitant congenital heart disease (CHD) may exhibit cardiac functional abnormalities, valvular dysfunction, bradicardia and AV block. Thus, due to increase in life expactancy in persons with DS, these persons are nedeed long-term follow-up in cardiovascular field. The aim of the present study to investigate the P-wave, QT and corrected QT dispersions which are reflects the tendency for atrial and ventricular arrythmias in children with DS without CHD.
Method: The standard 12-lead electrocardiograms of 100 children with Down's syndrome without congenital heart defects and 100 age-and-sex matched healthy children were prospectively assessed by a blinded specialist.
Results: Maximum durations of P-wave and QTc, dispersions of P-wave, QT and QTc were found significantly higher in DS group compared to without DS group. A positive correlation was found between P-wave dispersion and age in patients with DS. However, we could not find any association between the QT, QTc dispersion and age in DS group. And also any correlation were not determined between P, QT and QTc dispersions and gender.
Conclusions: In conclusion, our results showed that children with DS are more prone to ventricular and atrial arryhtmias due to the prolonged durations of PW, QT and QT dispersions. Thus, all children with DS should be carrefully assess with electrocardiography according to the possible atrial and ventricular arryhthmias during the cinical follow up even in the absence of concommitant CHD.

P-63

A simple marker in supporting the diagnosis of the incomplete Kawasaki disease: Red cell distribution width

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Objectives: In this study, we aimed to assess whether red cell distribution width (RDW) would be a useful supplementary laboratory marker to diagnose Kawasaki disease (KD), particularly its incomplete form (iKD).
Methods: We retrospectively evaluated the medical records of all the cases diagnosed with Kawasaki disease between 2006 to 2012. The patients were divided into two groups consisting of complete and incomplete forms of KD. Complete KD (cKD) was determined according to previously reported criteria. The patients who had prolonged fever and 2 or 3 clinical criteria together with at least three supplementary laboratory findings or echocardiographic coronary artery abnormalities were diagnosed to have incomplete KD. The complete blood counts of sex- and age-matched healthy children were used as controls.
Results: The number of the cases with KD and controls were 67 and 69, respectively. The groups were similar in terms of age and gender. RDW values were significantly higher in patients with KD compared to controls. Fourty-three cases had complete and 24 cases had incomplete KD according to clinical findings. The numbers of the patients with coronary involvement in those groups were 13 and 10, respectively. When clinical and laboratory findings of complete and incomplete Kawasaki disease patients were compared, age at diagnosis was significantly lower and RDW values were significantly higher in patients with iKD.
Conclusions: Our results showed that elevated RDW levels can be used as a simple, inexpensive laboratory marker in supporting the diagnosis of iKD which is frequently associated with coronary involvement.

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Echocardiographic Assessement of Cardiac Functions in Children with Surgically Induced Right Bundle Branch block after ventricular septal defect closure

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Objective: The aim of this study was to assess left (LV) and right ventricular (RV) functions in patients with surgically induced right bundle brunch block (RBBB) after ventricular septal defect closure.
Materials and Methods: 53 patients that follow up at least one year without any treatment after ventricular septal defect closure and 52 healthy controls were enrolled into study. The conventional and tissue Doppler echocardiographic measurements of patients with and without right bundle branch block were compared with each other and healty controls.
Results: RBBB was detected 37.7% of operated patients (20/53). Age at surgery mean: 25.2 ± 24.7 months, follow up duration mean: 5.3 ± 3.8 years were in RBBB(+) group. There were not significant statistical differences between RBBB(+) and (-) groups for surgical age and follow up duration, however in branch block group mean surgical age were lower than RBBB(-) group. In RBBB (+) group LV end-diastolic area (EDA) and LV end-sistolic area (ESA) were found higher than control, no significant differences were detected for other parameters. RV-TAPSE (Tricuspit annular plane systolic excursion) values were detected lower than control group. RV-FAC (Fractional area change) value was detected lower in RBBB (+) group. Pulsed wave Doppler measurements of the both ventricles in operated group; MPI (myocardial performance index) ratios were higher than controls. Tissue Doppler measurements of the LV and RV in operated patients; MPI and E/E′ values acquired from MLA (Mitral lateral annulus), TLA (Tricuspit lateral annulus) and S(septum) were detected higher than control group. No differences were found between E′/A′ ratios.
Conclusion: Our results showed that systolic and diastolic ventricular functions are decrease over time regardless of persence of RBBB. Therefore, ventricular functions should be periodically assessed with conventional and tissue Doppler echocardiography in order to determine early detection of ventricular dysfunction in children after ventricular septal defect closure.

P-65

Usefulness of an elevated neutrophil to lymphocyte ratio in predicting coronary artery involvement in children with Kawasaki Disease

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Objectives: In this study, we aimed to evaluate the usefulness of NLR in the diagnosis of any form of KD and the correlation between NLR and the persence of coronary artery lesions.
Methods: We retrospectively evaluated the medical records of all the cases diagnosed with Kawasaki disease between 2006 to 2012. The patients were divided into two groups consisting of complete and incomplete forms of KD. Complete KD (cKD) was determined according to previously reported criteria. The patients who had prolonged fever and 2 or 3 clinical criteria together with at least three supplementary laboratory findings or echocardiographic coronary artery abnormalities were diagnosed to have incomplete KD. The complete blood counts of sex- and age-matched healthy children were used as controls.
Results: The number of the cases with KD and controls were 72 and 71, respectively. The groups were similar in terms of age and gender. Twenty-five of those (34.7%) had iKD and twenty-two (30.6%) had coronary involvement. WBC, neutrophil, platelet counts, and NLR were significantly higher and conversely hemoglobin levels were lower in patients with KD compared to controls. The comparison of clinical and laboratory findings of complete and iKD revealed no difference in age, sex, WBC, neutrophil, lymphocyte, and platelet counts, NLR ratio, ESR and CRP levels ROC (reciever operator characteristics) analysis was performed in order to determine sensitivity and specifity of NLR value in predicting the KD and 1.34 value, was determined for predicting the KD with 70.8% sensitivity (95% CI 59.8-81.0), specificity 97.18% (95% CI 90.2-99.7), respectively. There were no significant differences between the patients with and without CALs groups in terms of age, gender, WBC, neutrophil, lymphocyte and platelet counts, NLR ratio, ESR and CRP levels.
Conclusion: In conclusion, it was found that NLR was increased in KD patients. Although we did not find any difference between complete and incomplete KD patients in terms of NLR, our results demonstrated that the diagnosis of KD can be supported by elevation of the NLR over a cut-off value of 1.34 especially in the cases with incomplete clinical features

P-66
Hypertension and Improved left ventricular mass index in children after renal transplantation

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Introduction: Hypertension and left ventricular hypertrophy (LVH) are observed in most children with end-stage renal disease (ESRD). The aim of this study was to evaluate LVH in patients with ESRD on dialysis and to compare after renal transplantation, and to evaluate the impact of BP parameters on LVH.
Methods: The study comprised 39 patients with renal transplantation (21 M, 18 F; mean age 14.54 ± 4.42 years). Medical records were reviewed for demographic features; casual blood pressure (BP) measurements, and echocardiographic evaluation were applied to all patients before and 6-9 months after renal transplantation.
Results: The mean Left ventricular mass index (LVMI) of patients before transplantation was $50.18 \pm 16.94\ g/m^{2.7}$, and 25 (64%) patients had LVH. The mean LVMI of patients after transplantation was $33.52 \pm 7.94\ g/m^{2.7}$, and 14 (36%) patients had LVH. The mean LVMI of patients before transplantation was significantly higher in patients than in the after transplantation ($p < 0.001$). The mean Systolic BP (41%) was 123.84 ± 11.59 mmHg and the mean diastolic BP (59%) was 78.20 ± 9.31 mmHg in the patients before renal transplantation. After renal transplantation; The mean Systolic BP (36%) was 117.10 ± 14.07 mmHg and the mean diastolic BP (51%) was 73.73 ± 8.72 mmHg. The mean systolic BP and mean diastolic BP were not significantly different before and after renal transplantation period.
Conclusion: LVH in children with ESRD is potentially reversible after renal transplantation, but hypertension may be permanent. There are several factors contributing to the development of hypertension in patients with ESRD; thus, it is possible that we did not fully control for all factors.

P-67
Persistent left superior vena cava as marker of anomalous pulmonary venous connection in patients with isolated atrial septal defect: Descriptive analysis of 441 pediatric cases

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Introduction: Atrial septal defects (ASD)are among the most common congenital heart diseases. The persistence of a left-sided superior vena cava (LSVC)is the most common variant of systemic venous drainage (0.5% of the general population, and up to 10% of those with established congenital heart disease), with a typical drainage to the coronary sinus. LSVC has been associated with an increased relation to atrial and ventricular septal defects, and several other malformations, and an increased risk of arrhythmias, most commonly atrial fibrillation in adult patients. We present 441 pediatric cases of isolated ASD and its association with anomaly of pulmonary venous connections and LSVC.
Methods: Retrospective observational study of isolated ASD in pediatric population and a description of the pulmonary venous connections and LSVC, from January 2003 to December 2013. Anatomic and surgical characteristics are also described.
Results: A total of 441 pediatric patients with isolated ASD were identified. Other heart congenital diseases were excluded. Range age was 1 month-18 years (mean 4,95), and data showed predominance in females (58%). Of these isolated ASD, had a surgical correction in 52% (232/441), and the other cases (209/441) had interventional catheterisation with a device or had a spontaneous closure. The most common reasons for the surgical correction were the anomalous pulmonary drainage and the impossibility to closure with a device due to anatomic incompatibility. Of all 441 isolated ASD, 69 patients (15%) had a partially or totally pulmonary anomalous drainage.
LSVC was present in 19 cases (4,3%)and no arrhythmia has been described, but a 24h cardiac-Holter was not in the cardiac protocol when a LSVC was detected. Of these 19 ASD combined with LSVC, we found an anomalous pulmonary venous connection in 9 cases (47%, sinus venous ASD), and corresponding to right pulmonary veins draining to right atrium (5), right pulmonary veins draining to right superior vena cava next to the venous sinus (3), and left inferior pulmonary vein draining to inferior vena cava next to the venous sinus (1). In these 19 patients, and the others with a partially or totally pulmonary anomalous drainage, surgical correction of the venous drainage and the ASD with a pericardial patch was required.
Conclusions: When an isolated ASD is related to LSVC, an anomalous pulmonary drainage is most frequently detected, and surgical repair will be required. Because the relation of LSVC to arrhythmias published in several reviews, a cardiac-Holter study seems recommendable in these patients and we added it at our protocol.

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Outpatient referrals to a tertiary paediatric cardiology service: evidence of increasing workload

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In the present era, demands on the specialist services provided in paediatric cardiology centres have increased dramatically. This study aimed to determine the volume and range of outpatient referrals to the paediatric cardiology service in a tertiary teaching hospital.
Information regarding new outpatient patient referral activity in the Department of Paediatric Cardiology, RBHSC was obtained over 2 separate time periods for comparison: January to April (inclusive) 2007 and January to April (inclusive) 2012. The total number of referrals for period January to April (inclusive) 2002 was also obtained.
In 2012 there were 423 new referrals made. This represents a four-fold increase from 2007. The most common reason for referral across both time periods was assessment of a murmur. The average age of referral was just over 4 years (4.7 years in 2007 and 4.3 years in 2012). There was a significant increase in the number of infants (under 1 year old) referred for assessment ($p < 0.05$). Overall the proportion of patients referred with no cardiac pathology has increased. Of the referrals made by GPs in 2012 90.2% had normal hearts compared to 80.3% in 2007. Of referrals made by paediatricians in 2012 82.7% had normal hearts compared to just 28.6% in 2007 ($p < 0.05$). In 2012 64.3% of patients were discharged following the initial outpatient assessment. New referrals to paediatric cardiology centres continue to increase at an alarming rate. The majority of patients referred have structurally normal hearts and over half can be discharged following the first out patient attendance. There appears to be an increasing reluctance to exclude cardiac pathology among GPs and more worryingly paediatricians in Northern Ireland over recent years. This may have implications for future training and planning of tertiary centre resources.

P-69

Timely diagnosis of congenital heart disease – did we improve?

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Background: 12 years ago, a study was published showing that in our referral population, cardiac diagnosis was made late in 10% of all patients with congenital heart disease (CHD) requiring therapy, resulting in too late initiation of therapy according to accepted standards of treatment timing. The rate of late diagnosis was the same in cyanotic and acyanotic CHD with 10% in each.
Methods: An identical study was performed as 12 years ago with a prospective evaluation of the time of diagnosis of CHD during a 3-year period ending in june 2011. Only patients with newly diagnosed CHD that required either catheter-interventional or surgical therapy were included. Of note that in between the two studies came the nationwide recommendation for neonatal pulse oximetry (POX) screening of all newborns starting in 2006.
Results: A total of 209 patients were included. 41% of these had cyanotic, 59% acyanotic CHD. According to the study criteria, late diagnosis was observed in 21 patients (10%), 6% (5 of 85) of cyanotic and 13% (16 of 124 patients) of acyanotic CHD. The 2 most frequent heart defects with delayed diagnosis were atrial septal defect and coarctation (7 and 6 patients, respectively). Delayed diagnosis resulted in one patient dead (undiagnosed interrupted aortic arch).
Compared to the historical study in our referral population, the striking finding was that still 10% of all CHD diagnosis was made late. The main difference was that late diagnosis in cyanotic CHD decreased from 10 to 6%, whereas in acyanotic CHD an increase from 10 to 13% was seen. Moreover an increase in fetal diagnosis was not surprisingly seen to currently 26% of all children with relevant CHD included in the study.
Conclusion: After 12 years of referring physician education and introduction of a nationwide POX screening, the rate of late diagnosis of CHD in our referral population remained stable at 10% of all CHD, only the rate of delayed recognition of cyanotic CHD showed a decline.

P-70

Cardiac Manifestations in Propionic Acidemia

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Introduction: Propionic acidaemia (PA) is an autosomal recessive disease that results from deficiency of propionyl-CoA carboxylase (PCC). In the majority of reported cases, the phenotype includes metabolic acidosis and/or neurological deficits. Our aim was to evauluate the relationship between propionic acidemia (PA) and cardiac involvement, which has been identified in literature.
Materials and Method: 12 year retrospective analysis of data in a single centre of patients with PA; electrocardiographs (ECG) and echocardiograms of all patients with PA analysed looking for patterns of presentation and progression of cardiac disease.
Results: 11 patients with confirmed diagnosis of PA. 10 had ECGs, all had echocardiograms. In our cohort, all the patients were of Asian origin, and there were 4 girls, and 7 boys. 2 children died, one following an attempted septal ablation for hypertrophic obstructive cardiomyopathy, and the other following a liver transplant. ECG analysis was undertaken on 10 patients as one child did not have an ECG. Of these, 4 had a normal corrected QT interval (QTc); 4 others had a normal ECG to start with but progressed to have QT prolongation, while the other two had prolonged QTs on theie first ECG. In two of the children, we identified intermittent QT prolongation on the 24-hour holter when there baseline 12-lead ECG was normal to start with. On echocardiography, one child had a small muscular VSD, while two others had small PFOs. All of them had a normal global systolic function with a normal fractional shortening; two had hypertophic cardiomyopathy of whom in one, the pathology was present at the first cardiac assessment and in the other, it developed at 3 years of age. Both the children who died had LV hypertrophy.
Conclusion: We have shown that there can be progression of ventricular repolarisation abnormalities and cardiac function. It is also important to undertake regular holter monitoring as in some cases the QT prologation is intermittent.

P-71

Noonan's syndrome: Cardiac pathology, outcomes and practice in a single institution

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Introduction: Noonan's syndrome is the commonest genetic syndrome associated with CHD after Trisomy 21, often requiring cardiac intervention. We reviewed our practice in the last

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13 years with regards to the type and rate of interventions as well as arrangements for follow-up and transition to adult services.
Methods: This was a retrospective review of patients' electronic records referred with Noonan's phenotype.
Results: Out of 128 patients, 105 patients were suitable for analysis (complete records). Only 17 (16%) patients had no cardiac abnormalities, of which 13 were discharged, some as early as at 3 years of age. There were 139 abnormalities in 87 patients, with valvar pulmonary stenosis the commonest type (64% of all patients). Other abnormalities included supravalvar pulmonary stenosis (9.5%), branch pulmonary stenosis (4%) and subpulmonary stenosis (2%). There were 15 cases of ventricular hypertrophy (14%) of which 4 were of the obstructive type (4%). ASDs were identified in 17%, aortic valve abnormalities in 8.5%, 2.8% had mitral valve abnormalities, PDA in 1% and VSD in 4.7%. There was one case with a partial AVSD, one with ccTGA and one with aortic coarctation.
A total of 57 interventions were undertaken in 47 patients (overall intervention 54%). 56% of patients with valvar PS required intervention (50% had a transcatheter procedure and 6% surgery). One quarter of those patients needed re-intervention. There were 9 more interventions for LVOTO relief, ASD closure, aortic coarctation and AVSD repair.
There were 6 deaths (5.7%). The cardiac findings in these patients were ventricular hypertrophy, dysplastic aortic valve and multi-level pulmonary stenosis.
Most of our patients with a known cardiac abnormality remain under follow-up by a paediatric cardiologist. At 16-18 years their care is transferred to the cardiology service in peripheral hospitals (17% patients) or the Grown Up Congenital Heart disease programme (12%).
Conclusions: In patients with Noonan's syndrome there is a high rate of cardiac abnormalities and interventions, therefore suitable follow-up arrangements during childhood and adulthood should be in place.

P-72
Mean platelet volume and the ratio of mean platelet volume/platelet count in acute rheumatic fever
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Objective: Acute rheumatic fever (ARF) is an endemic disease especially in developing countries. Due to an autoimmune response to group B streptococcus throat infection ARF develops in susceptible children. Mean platelet volume (MPV) reflects the platelet size and the rate of platelet production. It is important in cardiovascular events and rheumatic diseases. MPV/platelet count ratio was detected more sensitive than MPV alone in patients with hepatocellular carcinoma, deep vein thrombosis and myocardial infarction. The aim of this study was to investigate the alterations in MPV and MPV/platelet count ratio at the active and remission periods of ARF compared with healthy controls.
Methods: This study population consisted with 70 ARF patients and age – gender matched 70 healthy controls. In all subjects, complete blood count; including hemoglobin, white blood cell count (WBC), platelet count, MPV and C-reactive protein (CRP), erythrocyte sedimentation rate (ESR) were measured at the active stage and during the remission period in comparison with healthy subjects.
Results: There was no statistically significant difference between the ARF and control groups for the sex and age ($p > 0.05$). Forty-one patients of ARF had carditis. ARF patients at the active stages had significantly higher WBC, CRP and ESR values ($p < 0.05$). Although no significant difference was observed in MPV between the groups ($p > 0.05$); MPV/platelet count ratio was decreased at the active stage and increased again at the remission period as a similar the healthy controls ($p < 0.001$).
Conclusion: We did not find any relationship between MPV and ARF. However decreased MPV/platelet count ratio was detected at the active stage of ARF. The present findings emphasize the association between MPV/platelet count ratio and ARF. MPV/platelet count ratio may be used to determine activity of ARF disease.

P-73
Pulse Oximetry Screening for Critical Congenital Heart Disease in the Nordic Countries – implementation progress, update of current practice and a proposal for uniform guidelines
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Introduction: Pulse oximetry screening of newborn infants has been shown to increase the early detection rate of critical congenital heart disease and to minimize the risk of circulatory collapse before surgery. In spite of this, few countries yet have a national recommendation to screen. This study provides an update of the implementation of pulse oximetry screening in the Nordic countries and proposes standardized guidelines across the Nordic countries.
Methods: A questionnaire containing 28 items exploring pulse oximetry screening, clinical examination routines and availability of echocardiography was distributed to all delivery units ($n = 149$) in the Nordic countries in June 2013.
Results: The results describe the situation in September 2013. In Finland pulse oximetry screening had been implemented in 97% of all delivery units, in Sweden in 91% and in Norway in 90%. In Denmark 8% of delivery units were screening while no unit was screening in Iceland. Pre- and postductal screening was consistently used in Sweden and in 34% of delivery units in Finland. Postductal screening alone was used in 72% of units in Norway and in 55% in Finland. Screening was performed before 24 hours of age in 76% of the screening units in Sweden, 97% in Finland and 88% in Norway. Four Nordic countries lacked national guidelines for pulse oximetry screening, while Norway endorsed universal screening and guidelines in June 2013. As a result of the questionnaire a consensus was reached to propose uniform Nordic guidelines using pre- and postductal screening before 24 hours of age.
Conclusion: In Finland, Norway and Sweden the implementation of pulse oximetry screening is currently the highest in the world and coverage will be close to 100% before the end of 2013. Uniform guidelines across the Nordic countries will promote future collaboration and enable accurate comparisons.

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P-74

Dose Efficiency of Enteral Ibuprofen in Treatment of Patent Ductus Arteriosus in Premature Infants

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Hemodynamically significant patent ductus arteriosus (PDA) in premature infants must be closed due to the risk of serious morbidities. Enteral ibuprofen (EIBU) looks innocent compared with surgery and indomethacin but is not completely harmless. The aim of this study was to evaluate the efficiency of EIBU after each dose administered for PDA closure.
Sixty premature infants (≤33 week) who have hemodynamically significant PDA included in the study and treated with enteral ibuprofen. The first dose of IBU is administered via nasogastric tube in a dose of 10 mg/kg. Afterwards, 2 more doses of OIBU in a dose of 5 mg/kg are given at every 24 hours. Echocardiographic examination was performed after 24 hour of each doses. Treatment was stopped when the PDA is closed. Patients whose PDAs closed with ibuprofen treatment were followed in terms of recanalization. Treatment was continued in the absence of complications and if PDA wasn't closed until three courses were completed.
The closure rate after the first dose was 29.3%, second doses 32.7% and third doses 22.4%. At the end of first course, total closure rate was 84.4%. Among patients whose PDAs were closed after the first second and third doses, recanalization rates were 3.3%, 1.6% and 1.6%, respectively. Complications were infrequent, fairly mild and improved spontaneously in a short time. The mean PDA diameter of patients who did not respond to EIBU was significantly larger than that of responders ($p = 0.042$). Also, patients with larger PDA diameter needed more EIBU doses for closure ($p = 0.012$). Our results indicate that a in high number of preterm infants with hsPDA, ductal closure can be achieved with one or two doses of EIBU. Performing echocardiography after each dose can minimize unnecessary drug exposure and so possibly the side effects.

P-75

A Prospective Observational Cross-sectional Study on the Prevalence of Congenital Heart Malformations associated to Hirschsprung's Disease

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Objectives: to define the prevalence of associated congenital heart malformations (CHM) in patients with Hirschsprung's disease (HSCR) and to subsequently implement a personalized diagnostic algorithm.
Background: associated CHM have been detected in 5-7% of HSCR patients according to literature and, among them, septation and conotruncal development defects appeared the most frequent.
Method: all HSCR patients admitted between January 2009 and December 2013 were included in this prospective observational cross-sectional study. Cardiovascular screening included medical history, physical examination, a twelve lead electrocardiogram and an echocardiogram. Cardiac anatomy was routinely assessed by a segmental approach. Echocardiographic measurements respectively of the left ventricular dimensions and wall thickness, of the aortic root and of the left ventricular systolic and diastolic functions were obtained. CHM requiring a percutaneous or surgical intervention was described as major CHM.
Results: 133 consecutive HSCR underwent the cardiac screening. Mean age at enrolment was $5,3 \pm 6,1$ years. Eleven patients (8,3%) presented an associated CHD (Table 1). All patients who underwent an intervention are asymptomatic and without any residual cardiac defects at a mean follow up of $9 \pm 5,8$ years. We observed mild dilatation of aortic root in 3 patients whereas in other 2 measurements were within the superior normal limits.
Conclusions: in our series the prevalence of associated CHMs was slightly higher than in the previous papers, and were mostly represented by septal defects. Noteworthy, no one presented conotruncal heart defects. Six patients (4,5%) had major CHM and 4 of them had chromosomal abnormalities. If we do not consider the subpopulation of patients with HSCR and associated chromosomal anomalies, cardiac defects were still present in approximately 3.8% of the patients, which means that the prevalence of CHM in isolated HSCR population is higher than that of the general population. Basing on these results we suggest to perform routine echocardiogram in all HSCR patients, with or without associated chromosomal syndromes.

Patient	Sex	Age	Type of Heart Disease	Management	Syndrome
1	M	59	OS ASD + small PDA	F-up	
2	M	58	OS ASD + small PM VSD	F-up	Down
3	F	54	OS ASD	F-up	
4	M	51	OS ASD + moderate PM VSD	Intervention	
5	F	309	OP ASD	Intervention	Down
6	M	171	OS ASD + moderate PM VSD	Intervention	Down
7	M	192	large PM VSD	Intervention	Down
8	F	27	CoA	Intervention	Turner
9	M	96	OS ASD	Intervention	
10	M	52	tiny PDA	F-up	Down
11	M	107	small PDA	F-up	

P-76

Monitoring Epstein-Barr viral load assay after pediatric heart transplantation

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Introduction: Posttransplant patients are at risk of developing posttransplantation lymphoproliferative disorder (PTLD) associated with Epstein-Barr virus (EBV) infection. PTLD is potentially life threatening, but can be treated with success. At our unit monitoring of EBV viral load assay in serum and whole blood has been routinely used since 2010.
Objective: To study the correlation between EB viral load assay and development of PTLD at our unit.
Methods: A retrospective study of the medical history and laboratory parameters has been conducted. This study has involved: EBV serology, EBV viral load assay in plasma and whole blood after pediatric heart transplantation in Gothenburg, Sweden, between January 2010 and December 2013.
Results: The cohort consists of 30 subjects, two -18 years of age. Heart transplantation was performed at median 4 years (3 weeks–17 years) of age. During the observation time 10 subjects remained seronegative for EBV. Twenty subjects were seropositive for EBV; two subjects with PCR showing up to 3 log copies/ml, ten subjects had 3-5 log copies/ml, eight subjects more than 5 log copies/ml. A mismatch where the donor was seropositive and the recipient was seronegative for EBV was

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found in six cases (unknown in three). Two of them developed PTLD within a year after transplantation; both had 3-5 log copies/ml at the time before diagnosis. Another two subjects were diagnosed with PTLD 4 and 15 years after transplantation, one had less than 3 log copies before diagnosis, the other had between 3-5 log copies/ml. Both died during the study period. In all four subjects where PTLD was diagnosed, suspicions was raised due to symptoms as pain and organ failure, EB viral load varied between two and 5 log copies/ml at diagnosis.
Conclusions: The majority (20/30) of posttransplant children at our unit had EB viral load in blood. In children with diffuse symptoms PTLD should be suspected even when PCR in serum is below 3 copies/ml.

P-77
FBN1 mutation does not influence handling of paediatric patients with confirmed Marfan syndrome (MFS)

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Objectives: There is no single molecular or clinical test to demonstrate or rule out MFS. Clinical diagnosis of MFS is established according to Revised Ghent Criteria (RGC) and molecular diagnosis relies on fibrillin-1 (FBN1) mutation. In fact, although FBN1 mutation is the cause of classical MFS there are cases FBN1 mutation could not be detected. Therefore we evaluate whether there is a difference concerning clinical manifestation between paediatric Marfan patients with FBN1 mutation and those without and whether there is a need to distinguish those patient groups concerning follow-up, prophylaxis and therapy.
Methods: This study includes a cohort of 262 patients (10.9 ± 5.3 y) with confirmed or assumed MFS. They were subjected to standardised diagnostic programme including echocardiography and examination according to RGC. In addition molecular analysis was assessed whenever indicated. Prevalence and age of manifestation of the three cardinal symptoms of RGC were analysed.
Results: After all MFS was diagnosed in 103 patients (genetic analysis n = 86). The detection rate for FBN1 was 73.8%. We did not find any significant differences concerning prevalence or age of manifestation of the three cardinal symptoms of RGC by comparing patients with or without FBN1 mutation. Also analysis of different types of mutation did not show any significant variation.

Table 1. Prevalence and age of manifestation of dilatation of sinus of Valsalva (SV), ectopia lentis (EL), systemic manifestation (SysM) in patients with and without FBN1 mutation

	FBN1 mutation positive Prevalence	FBN1 mutation negative Prevalence	FBN1 mutation positive Age (years±SD)	FBN1 mutation negative Age (years±SD)	p-value
SV	65.6% (42/64)	68.2% (15/22)	8.9 ± 0.8	11.1 ± 1.5	ns
EL	25.0% (16/64)	13.6% (3/22)	7.2 ± 1.2	3.9 ± 0.2	ns
SysM	40.6% (26/64)	77.3% (17/22)	11.3 ± 1.0	13.7 ± 0.9	ns

Conclusions: All three cardinal symptoms did not show any significant variation between patients with or without FBN1. Both prevalence and mean age of manifestation were almost similar in the two patient groups. We conclude that there is no need to differentiate between paediatric patients with and without FBN1 mutation concerning follow-up, prophylaxis and therapy.

P-78
Epidemiological and clinical features, microbiological findings and prognosis of pediatric infective endocarditis in a teaching hospital in Tunisia

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Introduction: Infective endocarditis is a serious disease caused by the attachment and multiplication of germs in the endocardium. Mortality and morbidity of this disease are still significant despite advances in microbiological diagnosis, medical and surgical treatment.
Aim of the study: Study the clinical, microbiological, therapeutic and outcome features of infective endocarditis in children.
Methods: Retrospective study including 24 children with infective endocarditis collected in the pediatric department of Sahloul Hospital over a period of 19 years (from 1995 to 2013). Duke criteria was used to establish the diagnosis of infective endocarditis.
Results: 24 patients were involved in the study; 15 girls and 9 boys. The average age at diagnosis was 5 years old (40 jours-13 years). The first symptom was a fever in all cases. Clinical findings-signs showed a heart murmur in 17 cases and acute heart failure in 7 cases. Infective endocarditis had occurred on a plot of congenital heart disease in 17 cases, rheumatic heart disease in 2 cases and on healthy heart in 5 cases. Endocardium structures affected by endocarditis, identified with ultrasound, was mitral valve in 7 cases, 4 cases in aortic, tricuspid in 6 cases, pulmonary in 2 cases, on ventricular septal defect in 5 cases, at the ductusarteriosus, in a case of the ventricular wall in one case and a tube RV-PA in a case. The infectious agent was identified in 14 cases: it was a satphylococcus in 7 cases, Gram-negative bacilliin 5 cases, in one case a streptococcus and candida in one case. Antibiotics (cefotaxime+ fosfomycin ++ aminoglycoside) was administered in most cases. Cardiac surgery was used in 3 cases, the outcome was favorable in most of our patients. However eight children had complications kind cerebral stroke (7 cases), pulmonary embolism (1 case) and seven children had fatal outcome.
Conclusion: Infective endocarditis stills a serious disease by its local and general complications and by its mortality rate remaining high. Several questions are asked regarding the prevention of infectious diseases and the application of asepsis in our daily practice.

P-79
The Evaluation of Double Inlet Left Ventricle with Ventriculoarterial Discordance: Four Years Experience from a Single Center

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Objectives: Double inlet left ventricle is the most common form of single or common ventricle. The restriction of the bulboventricular foramen (BVF) and subaortik obstruction are important complications in these patients. We undertook a review of the data of patients with double inlet left ventricle (DILV), ventriculoarterial discordance (VAD).
Patients and Methods: Thirty-one patients diagnosed as DILV were evaluated between 2009 and 2013. The patient records reviewed retrospectively.
Results: Twenty-four of 31 patients have functionally single ventricle, VAD and systemic outflow through a BVF; no patient had outflow obstruction at birth. Thirteen were female (54%), 11

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were male (%45). Two patients have atretic pulmonary valve, 6 have pulmonary stenosis, 16 have pulmonary hypertension. One of the patients had arcus, isthmus hypoplasia at initial evaluation, where coarctation developed in another patient after pulmonary banding operation (PAB). Fourteen patients were operated and 3 patients planned to be operated, 2 patients were inoperable and 5 patients were lost follow up. Among the operated patients initially shunt operation was performed in 3, PAB in 9, bidirectional cavapulmonary connection (BCPC) in 2 patients. Initial palliation with PAB was performed in association with other procedures and the median age of initial palliation was 1,75 (0,5 to 2,5) months. The median follow-up duration was 7,5 (0,13-51) months. During follow up BCPC were completed in 6 of the patients (BCPC was performed as initial operation in 2) and the median age was 12,5 (8 to 156) months during BCPC operation. BVF restriction was not reported in our patients at admission or after PAB procedure. Arcus reconstruction and pulmonary banding was planned in one patient at admission and the coarctation developed on follow up was repaired in another patient 5 months after banding operation. Two patients died during follow up.
Conclusion: Norwood operation or pulmonary artery banding with aortic arch repair can be performed as the initial palliation in DILV, VAD patients. The latter procedure and early BCPC after pallative banding can be the procedure of choice to prevent early BVF restriction.

P-80
Determining the factors affecting the knowledge levels of ECG interpretation of the pediatric residents and interns (last year medical students) in ECG learning program
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Electrocardiography occupies a significant place in the diagnosis of cardiovascular diseases. In this study, it was aimed to determine the factors affecting the knowledge levels ECG interpretation of the pediatric residents and interns (last year medical students) before and after ECG learning program. 24 pediatric residents and 35 rotating interns were included. Interns and assistants were randomized into two groups. They were asked to complete a questionnaire (comprised of 20 EKG samples lacking clinical data other than age of the patients and of 30 findings) in 20 minutes. After training the same test presented to participants at one month and at one year. 16 (69.5%) of the residents were females and 7 (30.5%) were males. 3 (13%) of the residents were one-year residents, 8 (34.7%) were two-year residents, 6 (26%) were three-year residents and 6 (26%) were four-year. The rates of correct answering to the total findings pre-training were 24.2% in residents and 17.1% in interns; however post-training rates were 38.5% in assistants and 41.8% in interns; at one month they were 33.4% in residents and 42.1% in interns; at one year they were 32.7% in residents and 34.37% in interns. At the assessments performed during pre-training and at one month a statistically significant difference was found between groups. It was observed that the most correctly answered EKG sample was normal EKG; residents rate increased from 60.8% pre-training to 78% at one year; however, interns rate did not change significantly, 52.9% pre-training and 52.4% at one year. Pre-training, post-training and one-year total finding rates known by assistants who received cardiology rotation were higher than those who did not receive. However, similar values were observed at the end of one year and a statistically significant difference could not be found between two groups. In first three sessions, there was an increase in assistantship years and known total finding rates. At the end of one year the total finding rate known by 1-year assistants was detected as 15%; this rate was similar (30%) in 2-3-4-year residents and a statistically significant difference could not be found.

P-81
A rare cause of central cyanosis and nose bleeding: hereditary haemorrhagic telangiectasia with large pulmonary arteriovenous fistula
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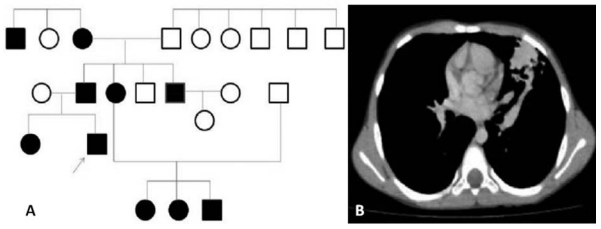
Introduction: Pulmonary arteriovenous fistula (PAVF is a rare cause of right-to-left shunting. The most of them are congenital, 50% of these cases are associated with hereditary haemorrhagic telangiectasia (HHT). We reported a 10-year-old boy who was presented with cyanosis and clubbing and diagnosed PAVF associated with HHT.



Case: A 10-year-old boy patient admitted to our clinic with cyanosis and clubbing for 6 years. There was frequent nose bleeding in family history (in his father, sister, grandmother, uncle, aunt and cousin) (Figure A). Physical examination revealed telangiectasia in the lips, central cyanosis, and clubbing. Oxygen saturation was 78%. The telecardiogram revealed irregular opacity in the middle and basal zone of the left lung. Echocardiographic examination was normal; contrast echocardiography showed bubble contrast in the left chambers, raising suspicion of PAVF. Computed tomographic arteriography confirmed multiple PAVF originating from the left inferior segmental pulmonary artery and draining into the left lower pulmonary vein. (Figure B). The PAVF was closed successfully with multiple coils by transvenous embolization. Oxygen saturation was 95% after embolization.
Conclusion: In this case, the patients who are presented with cyanosis and frequent nose bleedings in the family history should be considered as HHT. PAVF should be suspected during a contrast echocardiography study. Confirmation of micro bubbles entering the left atrium directly from a pulmonary vein suggests that a PAVF is present. Contrast computed tomographic arteriography is the modality of choice for defining the precise anatomy of PAVF.

P-82
Early repolarization and myocardial bridging in an adolescent with chest pain
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We present a case with chest pain who had ST-segment elevation (STE) and increased troponin and who was found to have

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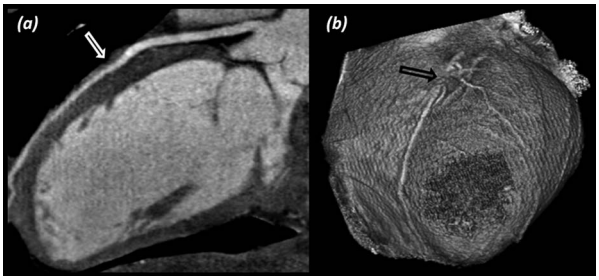
myocardial bridging (MB). Early repolarization pattern in the inferior leads was thought to result from ischemia caused by MB which adds uniqueness to presentation.
Case: A 17-year-old boy was admitted to the emergency department with burning, exertional chest pain that persisted for 2 hours. Initial ECG showed normal sinus rhythm with 2-mm STE in the inferior leads. On admission, his troponin was increased at 0.42 mg/ml and during 2 days it was undulant. Echocardiography showed no abnormality. Coronary artery imaging with multidetector computed tomography (MDCT) revealed a coronary myocardial bridging with a length of 12 mm in the middle tract of the left anterior descending artery (LAD). Magnetic resonance imaging (MRI) showed a reduced perfusion on segments 7 and 12 consonant with ischemia and a weak contrast uptake consonant with subendocardial infarct. These lesions matched the areas supplied by bridged segment of LAD which was defined by MDCT. An exercise stress test, based on the Bruce protocol, revealed no ischemic changes or arrhythmias. The patient was restricted from strenuous exertion. Metoprolol was prescribed and he was discharged without any problems. After 6-month follow-up, he had no cardiac symptoms, and his ECG remained normal.



Conclusion: Differentiating STE caused by acute myocardial infarction (AMI) from all other non-AMI etiologies, especially acute pericarditis-myocarditis and ER, can be challenging. In our patient, anginal chest pain was thought to be due to myocardial bridging of the LAD considering the possibility of a systolic narrowing of the coronary artery with subsequent ischemia. MB should be included in the differential diagnosis of children presenting with chest pain and signs of ischemia even in the absence of ventricular hypertrophy.

P-83
Comparison of two different head-up tilt test protocols to evaluate vasovagal syncope in children

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Introduction: Vasovagal syncope, is a common clinical problem in childhood which has higher incidence in adolescents. Head-up tilt test (HUTT) is the gold standard test in patients without organic heart disease. Different tilt test protocols are used for the evaluation of patients with syncope. In this study we aimed to compare the results of Westminster and Italian HUTT protocols. The passive phase of Italian protocol is 20 minutes which is 45 minutes in Westminster protocol.
Methods: 293 patients aging 6-18 years with a history of recurrent syncopes were involved in our study. Physical examination, laboratory parameters and echocardiographic examination were normal in all patients who were divided into two groups according to HUTT protocol. Westminster protocol (Group 1) and Italian tilt test protocol (Group 2) were performed in 150 and 143 patients respectively, the results were evaluated comparatively. Nitroglycerin was used sublingually in the active phase of HUTT protocols. Moreover, the results were analyzed according to different protocols and age. Statistical analysis was evaluated by using the T-Student's test, Chi-square test and Mann-Whitney U test.
Results: There was no significant difference between two groups by means of age, sex, age at first syncope and duration between first syncope and tilt test ($p > 0.005$). In Westminster protocol, HUTTs were 67%, 33% and 64.7% positive in passive phase, active phase and total respectively. These ratios were 46.3%, 53.7% and 56.6% in the Italian protocol respectively. There was not any significant difference between two protocols by means of positivity of the tests ($p = 0.160$). We found a significant difference in Westminster protocol in passive phase according to positivity of HUTT ($p = 0.002$). The type of syncope during the HUTT was shown in the table.

Table	Negative (n,%)	Cardioinhibitor (n,%)	Vasodepressor (n,%)	Mixt (n,%)
Group 1	53(35.3%)	15(10%)	34(22.7%)	48(32%)
Group 2	63(44.1%)	17(11.9%)	20(14%)	43(30.1%)

Conclusion: We think that Italian protocol has the advantage of time-saving. Thus, it seems to be more preferable than Westminster protocol. However, it should be considered that the positivity of HUTT in passive phase is higher in Westminster protocol.

P-84
Possible Involvement of Eicosapentaenoic Acid in Anti-platelet Therapy Effects and Possible Development of Artheriosclerosis in Chronic Kawasaki Disease patients

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Background: There has been reported that Eicosapentaenoic acid (EPA) sorting of n-3 fatty acid and cardiovascular disease are relevant. EPA is commonly contained in blue-skin fish and incorporated into platelet membrane phospholipid by ingestion. The incorporated EPA is metabolized to thromboxane A3 (TXA3) and leukotriene B5 (LTB5) by phospholipase A2. These metabolites respectively antagonize TXA2 and LTB4 which are metabolites of Arachidonic acid (AA). Because TXA2 can promote platelet aggregation and LTB4 can enhance inflammatory action, the state that EPA/AA is high will be extremely important. We thought that it would become an interesting research issue even in Kawasaki disease and focused on it in this study.
Methods: Twenty-four KD patients who were followed up more than five years were enrolled. EPA/AA ratio was measured by fasting blood sampling, Furthermore, platelet aggregation for an assessment of the effect of anti-platelet therapy and hydroperoxide suggesting oxidative stress as a relevant marker of arteriosclerosis, and %FMD as endothelial function were measured.
Results: According to the results of platelet aggregation (five grades; from class -2 to +2), patients are classified into three groups (Group A: from class -2 to -1, Group B: class 0, Group C: from class +1 to

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+2). Although the EPA/AA values were distributed in a wide range in group A, those in group B and C were going to be intensively distributed in low values. Moreover, although the patients with enhanced oxidative stress were included in those with low EPA/AA values, we could not find such patients in those with high EPA/AA values. Furthermore, the %FMD in patients with high EPA/AA values tended to be higher than those with low EPA/AA values.
Conclusions: In this study, the state that EPA is inferior to AA could be a reason for refractoriness of anti-platelet therapy in some chronic KD patients. The high values of EPA/AA possibly suppress the oxidative stress and EPA might contribute to inhibition of its development and progression of arteriosclerosis.

P-85
The catheterization and angiographic variations of coronaries in patients with Tetrology of Fallot

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Background: Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease and the catheterization and angiography still considered (in most centers) as essential preoperative diagnostic step. The prevalence of anomalous coronary artery in tetralogy of Fallot (TOF), has been reported as being up to 9%. These anomalies are not always detectable intraoperatively, particularly when they are covered by epicardial fat, pericardial-epicardial adhesions or by overlying myocardium.
Objective: To determine coronary artery anomalies in tetralogy of Fallot in children as seen on angiography.
Place and Duration of Study: The Children hospital & Institute of Child health, Lahore, Pakistan from January 2006 to December 2012.
Methodology: This is retrospective descriptive study. Children under 6 months to 16 years of age with echocardiographic diagnosis of tetralogy of Fallot were included in the study. All patients had pre-operative cardiac catheterization and angiography. Coronary arteries were studied with a non- selective aortic root angiogram in standard 45 (LAO) left anterior oblique & 20 cranial and 30 (RAO) right oblique views. The frequency of a normal and an anomalous coronary was determined by analyzing in SSPS-19.
Results: Of the 662 patients, 65.4% were male and 34.6% were female. The mean age was 69 (±43.14) months. Six hundred and twenty five (94.4%) had a normal coronary anatomy while 37 (5.6%) patients had anomalous coronary arteries. Among the patients with coronary anomalies, the commonest was a single origin coronary artery in 19 (2.9%) cases. Seventeen patients (2.6%) had coanal branch crossing right ventricular outflow track (RVOT) anteriorly and one (0.2%) had coanal branch crossing RVOT posteriorly.
Conclusion: Coronary artery anomalies were detected in 5.6% of the cases with tetralogy of Fallot. Single origin coronary artery anomaly was the commonest anomaly.

Keywords: Coronary anomaly. Angiography. Aortogram. Right coronary artery. Left coronary artery. Tetralogy of fallot. Single origin coronary artery

P-86
Paediatric ASD closure in the United Kingdom; practice vs demographics?

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In March 2013, the NICOR database was interrogated for paediatric ASD closure; from 2009-12. These publicly available data were analysed as a proportion of overall centre activity, & ratios of device vs surgical closure.
+/-400 ASDs are closed per annum in the UK and Ireland (6% of total activity). Indications for closure are (in theory) uncontroversial. The ratio of surgical to device closure nationally is 1:1 (0.85 to 1.05). Over the study period, Oxford ceased surgery, Irish practice also changed. These units are therefore excluded. NICOR has recently updated its website but sampling suggests no material changes to this dataset.

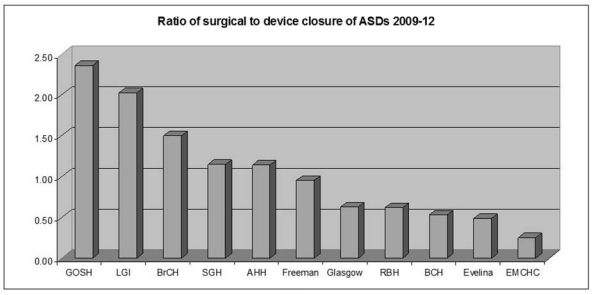

Figure 1

There are apparent regional variations in total ASD (surgical+‴ device) closure rates as a proportion of unit activity (3–11%). This is similar year on year with variation around the mean. It appears that ASD closure is less commonly performed in the Midlands and the North-East than other areas. There is wide variation across London.
There is much greater variation between units in the device/surgical ASD closure ratios (0.33:1 vs 5:1). This does not correlate with differences in percentage per unit activity.
Data extraction from CCAD/NICOR public-portal is time consuming. However, even for relatively straightforward lesions, it reveals differences either in demographics or clinical practice which merit further study. Variations may be due to patient differences or team preference; this should be examined with reference to physical and emotional outcomes. Differences in total ASD closure rates may have demographic implications and may skew complexity scoring; this needs further examination.

P-87
Unusual Manifestations of Kawasaki Disease

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Introduction: We report four unusual manifestations of Kawasaki disease occurring in our centre between 2000 and 2013.
Case 1: A 20-month-old boy was referred for high fever persisting for 48 hours with poor general condition. Secondarily, he had all major clinical criteria of Kawasaki disease associated with pulmonary, meningeal and gastrointestinal symptoms leading to initial misdiagnosis. Three courses of γ-globulins were needed but with persistent fever and poor general condition. Aneurysm of coronary, axillary and femoral arteries, and partial thrombosis of the right popliteal artery were revealed. Aneurysms of pulmonary arteries were also found, explaining the initial radiographic imaging misdiagnosed as pneumonia.
Case 2: The diagnosis of Kawasaki disease with coronary aneurysms was made in a 6-month-old girl with good response to γ-globulins. On evolution, intestinal occlusion revealed mesenteric ischemia and partial splenic ischemia combined with

coronary artery aneurysms. An ileostomy and resection of a large part of the small bowel were required. Prolonged parenteral nutrition and gastrostomy were also needed. Two years after initial presentation, she presented inferior myocardial infarction which was medically well controlled.
Case 3: A 2-month-old girl was admitted for fever 38.5°C with poor general condition. She was presented on day 2, acute abdomen with peritonitis. Surgical exploration concluded to medical peritonitis without appendicitis. Appendectomy and peritoneal drainage were performed. She was discharged home on day 8. She was readmitted on day 10 with the recrudescence of fever, impaired general condition and clinical signs of Kawasaki disease. Assessment found dilatation of coronary arteries. γ-globulin therapy was successful.
Case 4: A 5-month-old girl was referred for high fever persisting for five days, with major criteria of Kawasaki disease associated to an intense systolic murmur. Dilatation of the coronary arteries and high grade aortic insufficiency were found. General status remained poor despite γ-globulins. Echocardiographic control showed a worsening of aortic regurgitation caused by prolapsus of cusps with a total lack of coaptation and haemodynamic compromise. The child had a cardiac arrest and died before any surgical management.
Conclusion: Unusual manifestations of Kawasaki disease are pitfalls that may cause misdiagnosis with high rate of morbidity and/or mortality.

P-88

Kawasaki Disease, Clinical Characteristics and Risk Factors for Coronary Aneurysm in Children Hospitalized in Three Hospitals in level IV 2000-2012

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Introduction: Kawasaki disease is a vasculitis of unknown etiology, affecting blood vessels of small and medium caliber, the main complication is the formation of coronary artery aneurysms. It is the leading cause of acquired heart disease in children in developed countries. In 2011 an outbreak was observed in our environment, so it is important to understand clinical manifestations, complications and seek risk factors associated with the development of coronary artery abnormalities.
Objectives: To determine the clinical and laboratory features that relate to the development of coronary artery abnormalities in patients hospitalized with the diagnosis of Kawasaki disease in three hospitals in level IV between 2000-2012.
Material and methods: Observational, descriptive and retrospective study, between the years 2000-2012. Cases through medical record review were identified.
Results: 70 cases were observed in twelve years, being more common in males. The largest number of cases in children under 2 years. The main clinical manifestations fever, changes in the oral mucosa, bilateral conjunctival hyperemia, exanthem, changes in the extremities and cervical lymphadenopathy less frequent. In laboratory data include C-reactive protein, leukocytosis, trombocitocis, anemia and increased transaminases Cardiac complications occurred in 38% being the most common coronary aneurysms and second coronary dilation and thirdly spill pericardial. Male sex (OR = 2.5), exanthem (OR = 1.3) and CRP >10 mg/dl (OR = 6.4) were identified as risk factors for coronary aneurysms.
Conclusions: The typical Kawasaki disease is the most common. Kawasaki disease is on the rise in recent years, early diagnosis and early treatment prevents complications.

P-89

Long-term monitoring by MRI of coronary arteries after arterial Switch operation in D transposition of the great arteries

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Introduction: Transfer of the coronary arteries during arterial switch operation (ASO) is the principal step; the long term morbidity and mortality of this operation depend on the status of the coronary perfusion, and coronary obstruction is an important cause of death with an incidence around 3-8%. However, severe coronary artery injuries may occur in asymptomatic patients and its diagnosis is still a challenge. This study aims to assess the incidence and sensitivity of magnetic resonance imaging (MRI) in the diagnosis of the coronary obstruction after ASO.
Method: Retrospective study of the patients with ASO since 1997 in our hospital. The coronary study was performed using MRI. Patients with suspected coronary lesion were performing a 64 multislice computed tomography (MSCT). The presence of symptoms, the time of diagnosis and other diagnostic tests was recorded.
Results: 157 patients were operated with ASO. Follow up MRI was performed in 88 patients (mean age of 8 years). Five patients have an image of coronary stenosis in MRI, three of them were confirmed with MSCT (incidence of 4,3%). The first was a girl of 16 years old, asymptomatic, with normal EKG and TTE. Severe stenosis of left trunk coronary was shown in both noninvasive image techniques (MRI and MSCT). A stent was implanted to treat the obstruction. The second patient was a 15 years old boy with normal stress test, EKG and echocardiography. Although he was asymptomatic, MRI and MSCT dropped out a severe obstruction of DA. This severe obstruction was treated with a stent implantation too. The third one, a twelve years old boy with a previous image of stenosis in right coronary artery in MRI, was confirmed by to MSCT present a mild stenosis. He is asymptomatic without treatment. The other 2 patients have images suggestive of coronary stenosis in MRI but MSCT did not confirm them.
Conclusion: After arterial Switch operation coronary events are not rare. Asymptomatic patients with an uneventful course after ASO may have coronary obstruction. While other noninvasive diagnostic tests are inconclusive, MRI and MSCT are a good diagnosis method. Protocol of diagnosis needs to be established.

P-90

NT-pro BNP after Fontan Anastomosis: Early versus Late Post-operative Value

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Introduction: NT- proBNP has become a diagnostic marker for the diagnosis aiding prognosis and therapeutic guidance of cardiac conditions in adults. However, its assessment remains limited in congenital cardiology, particularly in patients with univentricular heart physiology who underwent total cavopulmonary connection (TCPC) where long-term prognosis is greatly related to ventricular function.

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Aim: To evaluate the NT- proBNP serum level after TCPC and seek a possible correlation with clinical and laboratory monitoring.
Methods: A retrospective study was initiated on all TCPC patients born after 1991. Demographic, anatomical and echocradiography data were collected for all subjects following the early post-operative period. Routine biochemical data including NT-proBNP were similarly collected. Routine NT-proBNP screening for these patients was initiated in our institution in 2008. NT –proBNP z –score was calculated based on our published equation from healthy children.
Results: Of a total of 46 patients who completed TCPC, 5 died post-operatively. All remaining 41 subjects had available NT-proBNP beyond the immediate post operative period and constituted the study population. Mean TCPC age was 4.8 ± 1.1 years, with a mean follow-up of 6.24 ± 5.0 years. Serum NT-proBNP level was significantly elevated during the first post-operative year (Z-score 1.9 ± 1.17) compared to mid-term follow-up (3-5 years) (Z-score 1.23 ± 0.72), $p = 0.03$. Mean NT-proBNP Z-score increased in the subsequent years to 1.6 ± 1.1 ($p = 0.21$ v.s. year-1 post TCPC, and 0.06 v.s. mid-term). Z-scores >2.0 were associated with a lower serum albumin (39.2 ± 3.3 v.s. 44.4 ± 4.4 mg/dl; $p = 0.04$), but not with other laboratory tests. There were however no identifiable predisposing clinical factors (i.e., ventricular morphology, age), preoperative hemodynamic data (e.g., PVRi, mean pulmonary artery pressure, Nakata index), surgical specifics (type of TCPC, presence of fenestration, pacemaker implantation), or postoperative events.
Conclusions: The TCPC postoperative year is marked by a significant increase in the levels of NT-proBNP. This is probably related to an adaptation period to the uni-ventricular physiology assuming a full cardiac output. The observation does not seem to vary between left and right dominant ventricular anatomy, neither did it correlate with various parameters. The NT- proBNP improved in the mid-term follow-up.

P-91
Report of two observations of left ventricular systolic disfunction in children with mucopolysaccharidosis (MPS) type 1
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Introduction: Our purpose is focusing pediatricians and cardiologists on the problem of early diagnosis of rare genetic diseases in children with heart disease.
Methods: In the department of cardiology Scientific Centre of Children Health RAMS were admitted 2 children with a diagnosis of dilated cardiomyopathy. On physical examination both children at an early age (3 and 7 months) were observed to have coarse facial features (wide and depressed nasal bridge, large nostrils, hypertelorism, gingival hypertrophy), hirsutism, short neck, hypersalivation, noisy breathing, joint stiffness, hepatosplenomegaly. Also both children had symptoms of congestive heart failure. An electrocardiogram showed atrial hypertrophy, hypertrophy of both ventricles, marked disturbances of repolarization. According to the ultrasound examination – dilatation of the left chambers of the heart, a reduction in myocardial contractility (EF < 35%). We paid attention to uncharacteristic for dilated cardiomyopathy hypertrophy of the left ventricular wall (6-7 mm). Based on clinical findings we suspected a storage disease, in both cases, the diagnosis of MPS type 1 was confirmed by the analysis of urine glycosaminoglycans and genetically. The specific enzyme replacement therapy with laronidase (Aldurazim©) was initiated at early age.
Result and conclusions: At MPS type 1 (Hurler syndrome) heart is always affected, manifesting with ventricular hypertrophy, thickening of the heart valves, but preserved myocardial contractility, and chronic heart failure usually occurs only at late stages of the disease. In this case, the two children have shown a dramatic dilation of the left heart and decreased myocardial contractility, which led to the development of symptoms of congestive heart failure caused, apparently, by myocarditis. The presence of a cardiac dilatation and reduced ejection fraction in conjunction with myocardial hypertrophy and specific clinical features is the ground for a deep survey on storage diseases, which helps to diagnose a rare genetic disease at an early age and start specific therapy at the proper time.

P-92
Only Hepatic Venous Blood Closes Intrapulmonary Shunts after Cavopulmonary Connection
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Objectives: Intrapulmonary shunts in congenital heart disease lead to significant morbidity and mortality. The exact pathophysiology is still under discussion although the most favourable hypothesis involves the lack of an unknown liver factor in pulmonary perfusion. This study compares therapeutic options of intrapulmonary shunts in patients with single ventricle.
Methods: We retrospectively evaluated nine patients with intrapulmonary shunts and a functional single ventricle, four of them in combination with heterotaxy.
Results: All patients had surgical management by means of a Fontan operation or cavo-pulmonary connection. In each of the patients the hepatic venous blood did not reach the lung or was angiographically unequally distributed between the lungs. The side with diminished hepatic venous blood flow showed a more frequent appearance of shunts. After the Fontan operation or cavopulmonary connection the shunts evolved after different periods of time, ranging from months to several years. Patients treated only with pulmonal vasodilators (oxygen or sildenafil) showed no substantial increase in oxygen saturation. Of the four patients treated successfully, in two the Fontan circulation was completed by total cavopulmonary connection. One had already a Fontan circulation, but hepatic venous blood was unequally distributed due to stenosis of the left pulmonary artery which was managed successfully by stenting. And one patient needed pneumonectomy due to massive shunts in one lung.
Conclusion: The appearance of intrapulmonary shunts in congenital heart disease is linked to pulmonary perfusion without hepatic venous blood. The only causative therapy seems to be redirecting hepatic venous blood to the lungs. If this is not possible, other therapy options, such as selective pulmonary vasodilation, provide low chances of success.

P-93
High levels of uric acid predicts latent heart failure and need for earlier operation in infants with ventricular septal defects
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Background: High levels of uric acid (high-level UA) were reported to be a predictive indicator for prognosis in adult patients with chronic heart failure. If infants with VSD have to undergo surgery earlier, they have more serious heart failure. We predicted VSD infants with high-level UA had to undergo surgery earlier. We attempted to identify clinical presentation and cardiac function in VSD infants with high-level UA.
Methods: Between 2005 and 2012, consecutive 105 infants with VSD were studied. Cardiac catheterization was performed in preparation for surgery. Venous blood sample for analysis of UA was obtained at the same point in time. We defined high-level UA as uric acid levels in the top fifth of 105 patients (UA ⩾ 5.7 mg/dl). First, the timing of operation was compared between patients with high-level UA and those without high-level UA. Second, cardiac performances were determined which affected high-level UA.
Results: Within 4weeks after catheterization 59% of patients were operated in high-level UA group, whereas 30% of patients in no high-level UA ($p = 0.032$); within 6weeks 77% operated in high-level UA, whereas 40% in no high-level UA ($p = 0.0038$). However, descriptive features which were turned up by means of daily clinical practice, such as standard deviation of body weight and cardiothoracic ratio, were not different between two groups. After multiple logistic regression analysis high-level UA was independently associated with odds ratio of 7.6 for high levels of brain natriuretic peptide (⩾175pg/ml), 5.5 for elevated end-diastolic pressure of left ventricule (⩾12 mmHg), 13.1 for increased levels of creatinine (⩾0.28 mg/dl) and 13.3 for high dose of lasix (⩾2.2 mg/kg/d).
Conclusion: Our study shows VSD infants had to undergo surgery earlier with high-level UA, although their clinical statuses were not different from those in patients without high-level UA. In addition, high-level UA was related to elevated end-diastolic pressure of left ventricle, high levels of brain natriuretic peptide, reduced renal function and high dose of diuretics. These factors were indexes which reflect worsening of heart failure in VSD infants. We could use high-level UA as the method of picking up VSD infants whose heart failure is worsening and who needs earlier surgical intervention.

P-94
Systemic pulmonary hypertension in Down syndrome infants with ventricular septal defect is associated with hypoventilation and large various shunts

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Introduction: Systemic pulmonary hypertension (systemic PH) often resides in Down infants with ventricular septal defects (VSD). However, there are few reports to describe risk factors for systemic PH in Down infants with VSD. The purpose of this study is to investigate why systemic PH often subsists in Down infants with VSD.
Methods: The medical records of 222 infants with VSD were reviewed (44 with Down syndrome and 178 with non-Down). All had no significant stenosis of pulmonary artery. They had cardiac catheterization in view of surgical intervention between 1993 and 2012. Arterial carbon dioxide pressure (PaCO2) was measured during cardiac catheterization. Maximum dimension of VSD on echocardiography was calibrated by the body surface area. We calculated the right-to-left pressure ratio on end-systole point (RV/LV). We defined systemic PH as value of RV/LV in the top of fifth (RV/LV ⩾ 0.99). First, independent factors which influenced systemic PH were determined in all 222 infants. Second, we investigated relationship between these independent factors for systemic PH and Down syndrome group.
Results: The ratio of patients with systemic PH was significantly higher in Down infants than in non-Down infants (38% vs. 16%). After multiple logistic regression analysis systemic PH in 222 infants was independently associated with large dimension of VSD (⩾43 mm/m2), existence of patent ductus arteriosus, existence of atrial septal defect/patant foramen ovale, and increased levels of PaCO2 (⩾47 mmHg). The ratio of patients who had each factor was significantly higher in Down infants respectively. However, these four factors were not associated with systemic PH only in Down group, although they were all associated with systemic PH only in non-Down group.
Discussion: This study revealed large VSD, accompaniment of other left-to-right shunts and hypoventilation were related to systemic PH, each of which more existed in Down infants. This would cause systemic PH more frequently in Down infants. However, these independent factors were not related to systemic PH only Down group, although those were associated with systemic PH only in non-Down group. Some other factor relevant to Down syndrome itself, which was not disclosed in this study, would be responsible for systemic PH.

P-95
Patients misdiagnosed as persistent pulmonary hypertension of neonate are especially severe cases of total anomalous pulmonary venous connection

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Introduction: Total anomalous pulmonary venous connection (TAPVC) was occasionally misdiagnosed as persistent pulmonary hypertension of neonate (PPHN). Both disorder had severe pulmonary hypertension and interatrial right-left-shunt. If TAPVC patient is in particularly serious condition, its common chamber is hardly-detectable by echocardiogram. We predict PPHN-misdiagnosed patients were especially severe cases of TAPVC. We investigated clinical characteristics of PPHN-misdiagnosed patients with TAPVC.
Methods: The medical records of six patients with TAPVC who was diagnosed as PPHN initially were reviewed. They underwent surgery between 1 day and 12 days after birth. We used 13 patients with TAPVC as control who underwent surgery between 1 day and 12d days. Clinical findings were compared between two groups.
Results: In PPHN-misdiagnosed patients symptoms appeared within 4 hours after birth (83% vs. 7%, $p = 0.0095$). Before surgery PPHN-misdiagnosed patients were more on respirator (100% vs. 7%, $p = 0.00025$). In PPHN-misdiagnosed group inotropic drugs administered more (83% vs. 7%, $p = 0.029$). Half of patients in PPHN-misdiagnosed group died after TAPVC repair, whereas only 15% died in correct-diagnosed group. The differences on images were two factors in PPHN-misdiagnosed group: pneumothorax (50% vs. 0%, $p = 0.02$); small cardiothoracic ratio 47% or less (66% vs. 7%, $p = 0.021$). It is difficult to distinguish two groups in following factors: levels of oxygen saturation; degree of interstitial opacity; intensity of pulmonary hypertension; dimension of left ventricle. Types of TAPVC and

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existence of pulmonary venous obstruction are also not different between two groups.
Conclusion: The patients misdiagnosed as PPHN were especially serious in cases where TAPVC repair was needed earlier after birth. They had smaller hearts on chest X-ray with pulmonary vein flow potentially decreased. Furthermore, they had pneumothorax. Possibly small common chamber were hard to be detected by echocardiography because of decreased pulmonary-vein flow and pneumothorax. There was no easy-to-understand information on images except for these two factors in PPHN-diagnosed patients. We should review infants to suspect TAPVC who were diagnosed as PPHN and medicated intensive care with small heart or pneumothorax.

P-96
Syncope Unit in pediatric population, a single center experience
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Introduction: Syncopes are frequent in the pediatric population. The majority is benign but, for a minority of children, a cardiac disease is the underlying cause and has to be recognized promptly as it can be fatal. Syncope units developped in adult population have demonstrated major improvement in diagnostic process, hospitalisation reduction time, with favourable long-term outcome. We report our experience of syncope management in chidren and adolescents through a dedicated syncope unit.
Methods: This ongoing prospective study enrolled 98 consecutive patients (12 ± 3 years old, 52% male), referred for loss of consciousness (LOC) in a dedicated pediatric syncope unit involving a pediatric cardiologist, a nurse, a physiotherapist and a psychologist. All patients underwent initial evaluation including medical history assessment, physical examination, 12-lead ECG and echocardiography to exclude non-cardiogenic syncopes. If initial assessement was abnormal, they underwent targeted tests that differed according to suspected aetiology. Patients with neurocardiogenic syncope underwent specific physiotherapy training and a consultation with a psychologist.
Results: The most common causes of LOC was neurocardiogenic syncope – 70 patients (71%) and psychogenic LOC – 20 patients (21%). Cardiac syncope was present in 5 patients (5%), 2 had long QT syndromes and received beta blocker therapy, 2 presented third-degree atrioventricular block and had pacemaker implantation and one had cathecholergic polymorphic ventricular tachycardia and received beta blocker therapy. Two patients had typical epileptic seizure and were transferred to neurologic department. Mean hospitalization duration was 0,9 ± 0,5 days. Head-up tilt testing was positive in 60% neurocardiogenic syncope. Echocardiograms and exercise tests were not contributive for diagnosis. After a mean follow up of 11 ± 5 months, including physiotherapist and/or psychologist specific care, syncope reccurence occured in 11% patients.
Conclusion: Syncope unit in pediatric population with dedicated team improves diagnostic process, reduces hospitalisation and decreases syncope recurrence when adapted follow up is proposed.

P-97
Cardiac involvement in paediatric patients with mitochondrial disease
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Introduction: Mitochondrial diseases have a wide clinical spectrum, with multisystemic manifestations and can present at any age. Cardiac abnormalities are often present and include hypertrophic or dilated cardiomyopathy, heart conduction defects and ventricular pre-excitation. These manifestations may be late, subtle or can be the first presentation of the disease. The aim of this study was to review the cardiac manifestations and its impact in the outcome of the patients followed in a tertiary hospital.
Methods: The authors made a retrospective analysis of clinical, laboratorial records and cardiac exams (EKG, Doppler echocardiography, Holter monitoring) of children with probable mitochondrial disease (MD), according to the modified Walker criteria (Bernier et al., 2002).
Results: The study includes 22 patients, aged 1 day to 4 years at presentation of MD, with a median follow-up of 72 months (min 9 months, max 156 months). Eight patients (36%) had cardiac involvement, with a male-female ratio of 5:3. The cardiac diagnosis preceded the mitochondrial disease diagnosis in 2 cases presenting hypertrophic cardiomyopathy in the newborn period (one was asymptomatic but had left ventricular hyperechogenic foci in fetal ultrasound and the other developed respiratory distress syndrome and cyanosis after birth), while the other patients developed the cardiac abnormalities during follow-up (age of cardiac diagnosis varied from 7 days to 14,5 years). EKG abnormalities were found in all eight patients.
Five had primary myocardial disease: hypertrophic cardiomyopathy (n = 5), and systolic dysfunction of the left ventricle (n = 1); and two had conduction abnormality (atrioventricular block and WPW syndrome). One patient had implantation of pacemaker at the age of 14 years due to advanced atrioventricular block. Mortality was 12,5% in the cardiac group vs 7% in the non cardiac group.
Conclusions: Cardiac abnormalities are often present in MD (36% of our children) but the patterns of heart involvement are very heterogeneous. They may occur as the principal clinical manifestation, may have an early or late onset, and carry a worse prognosis. The authors emphasize the need of a periodic cardiac evaluation and follow-up of these patients.

P-98
The effect of exercise training on cardiac remodelling in adolescents with corrected tetralogy of Fallot and Fontan circulation: a randomized control trial
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Introduction: Exercise can improve physical fitness in children and adults with congenital heart disease. Cardiac effects have hardly been studied. We hypothesized that exercise training would not lead to adverse cardiac remodelling.
Methods: This multi-centre randomized controlled trial included adolescents (10 to 25 years) with either corrected tetralogy of Fallot or Fontan circulation. The exercise-group was enrolled in a 12 week standardized aerobic dynamic exercise training program. The control-group continued their life-style as usual. Both groups underwent cardiopulmonary exercise testing, cardiac magnetic resonance imaging (MRI), echocardiography and neurohormonal assessment, 2 weeks before and within 2 weeks after the intervention period.
Results: Fifty-six patients were randomized to the exercise-group, 37 to the control-group. We assessed changes between the pre- and post-intervention period for the exercise group compared to the changes in the control-group. Peak load increased significantly in the exercise-group compared to the control-group (exercise-group 6.9 ± 11.8 Watt; control-group 0.8 ± 13.9 Watt; $p = 0.047$). There were no adverse events linked to the study. Ventricular systolic parameters, cardiac dimensions and neurohormonal markers did not change during follow-up in patients allocated to the exercise-group and control-group. There were some isolated minor but statistically significant changes in inflow parameters. The pattern of these changes was not consistent, indicating a lack of true change in the diastolic function.
Conclusion: We demonstrated that no adverse cardiac remodelling occurred after 3 month exercise training in adolescent patients with either a corrected tetralogy of Fallot and Fontan circulation. This data may provide support for improved clinical guidelines.
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P-99
Changes of Pattern and Incidence of Childhood Rheumatic Fever in Sabratha Teaching Hospital
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Fifty seven Libyan children suffering from Rheumatic Fever were diagnosed and treated in Sabratha Teaching Hospital in the periods of June 1986–May 1988 and from June 2000–May 2002. The number of patients in the first period was forty five patients and in the second period was twelve patients.
There were twenty three boys and twenty two girls in the first period and seven boys and five girls in the second period. The mean age in the first period was 10.8 years (3-16 years) and the mean age in the second period was 9.3 years (6-12 years).
Concerning the first period, Arthritis was diagnosed in 93% of patients (42 patients); ankle joint Arthritis was the commonest joint involved followed by knees and wrist joints. Carditis was diagnosed in 66% of patients (30 patients); the commonest valvular lesion was mitral regurgitation. There were four patients presented with Rheumatic Chorea as the only manifestation and in two patients there was Erythema marginatum and in one patient there were subcutaneous nodules.
Concerning the second period, Arthritis was diagnosed in all patients. The commonest joints involved were the ankles followed by the knees & wrists. Carditis was seen in 7 patients (58%); the mitral regurgitation was the commonest valvular lesions. There were no recorded cases with rheumatic chorea, Erythema marginatum or subcutaneous nodules.
In conclusion it appears from this comparative study that the incidence or Rheumatic Fever in our region is declining during the recent years and its pattern becomes much less severe than before.

P-100
Results of Treatment with Endothelin Receptor Antagonist and Prostacylin Analogs for Patients with Pulmonary Arterial Hypertension (PAH)
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Introduction: The short and long term consequences of treating PAH patients with endothelin antagonist and prostacyclin analogs are evaluated in this study.
Methods: 25 PAH Patients (16 girls, 9 boys), followed from 2001 to 2012, are included in this study. Patients have been evaluated pre and post treatment with the following tests: cardiac catheterization (flows ratio, average pulmonary arterial and systemic arterial pressure, pulmonary and systemic vascular resistance index), echocardiography (right ventricular pressure), functional capacity (NYHA) and 6-minute walking distance (6-MWT).
Results: The average age of patients is 12.5 years (2-28 years) with an average treatment start age of 9 years. The average weight is 30 kg. The average duration of treatment is 18 months and the average period between two hemodynamic studies is 24 months. 16% of the cases have idiopathic pulmonary arterial hypertension (IPAH), while %84 of the cases have innate heart disease with shunt. 32% of the patients have down syndrome. 52% of the cases have been treated with 52% prostacyclin analog (Ilioprost) while 48% have beentreated with endothelin antagonist (Bosentan). The treatment of 3 patients (%12) have been switched from Ilioprost to Bosentan (1 due to disphony and resistance to treatment,1 due to resistance to treatment and 1 due to treatment nonconformity) There have been statistically significant drops in average pulmonary arterial pressure, PVRI and NYHA functional capacity when compared to pre-treatment levels, while a statistically significant increase in 6-WMT is observed (p-value = 0.05). There is not a significant difference detectable pulmonary arterial pressure detectable via echocardiography. The difference in clinical and hemodynamic parameters between patients treated with Ilioprost and those treated with Bosentan is not found to be statistically significant
Conclusion: Specific PAH treatment improves quality of life of patients while the difference in the effects of various medications to the clinical and hemodynamic parameters is not significant. The medication preference should therefore be based on the age of the patient, the conformity of the medication to the patient and/or the family of the patient, ease of use, the period it takes until the medication starts becoming effective and side effects of the medication.

P-101
Role of late gadolinium enhancement magnetic resonance imaging in the management of patients with primary cardiomyopathy
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Introduction: Late gadolinium enhancement magnetic resonance imaging (LGE-MRI) has been widely accepted for detection of myocardial fibrosis, which has been implicated as a factor in cardiovascular events including sudden cardiac death. We introduce

some cases of primary cardiomyopathy that MRI study including LGE-MRI was useful for management.
Methods: We performed MRI study for five patients with primary cardiomyopathies. Two siblings with hypertrophic cardiomyopathy (HCM) caused by an alpha-tropomyosin1 (TPM1) mutation. Twin sisters and a girl with overlap of restrictive and hypertrophic cardiomyopathy (RCM/HCM) caused by a cardiac troponin-I (TNNI3) mutation and a myosin light chain (MYL2) mutation, respectively. Elder brother with HCM (patient 1) was diagnosed at 13 years old, and younger brother (patient 2) was diagnosed at 6 years old. Twin sisters with RCM/HCM (patient 3 and patient 4) were diagnosed at 11 years old. A girl with RCM/HCM (patient 5) patient was diagnosed at 7 years old. The extent of LGE was expressed as the percentage of the total left ventricular mass (the % of LV mass).
Results: The extent of myocardial fibrosis by LGE-MRI (age at examination) in patients 1 to 5 was 8% (18), 1% (10), 18% (11), 7% (11), and 6% (18) of LV mass, respectively. Life-threatening events were found in patient 1 (syncope due to ventricular tachyarrhythmias) at 18 years old and patient 3 (sudden death) at 11 years old. Patient 2, 4, and 5 have been uneventful. The extent of LGE increased to 16% of LV mass by re-examination two years later in patient 2.
Discussion: Implantable cardioverter defibrillator (ICD) was implanted for secondary prevention of sudden death in patient 1. Furthermore, we decided to place a primary prophylactic ICD in patient 2 because the extent of LGE in patient 2 was larger than that of the patient 1. In the case of RCM/HCM, we arrange heart transplantation because myocardial fibrosis might be in progress with patient 4 as in the case of patient 3.
Conclusions: LGE-MRI could be useful to manage patients with cardiomyopathy and to estimate their prognosis.

P-102
Analysis of clinical characteristics and causes of chest pain in children and adolescents
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Objectives: Chest pain is a common problem to visit pediatrician in children and adolescents and is the second most common symptom after cardiac murmurs which is referred to pediatric cardiologists. Serious diseases such as coronary ischemia are very rare in children and adolescents.
Although most of chest pain in children and adolescents are benign and need no treatment, timely diagnosis is important to not to miss several diseases that require prompt treatment.
Methods: To investigate diverse causes of chest pain in children and adolescents and to descriminate diseases that need further evaluation and prompt management of chest pain, retrospective medical records were reviewed during the period of July 2006~October 2013. A total of 517 patients of chest pain were referred to Department of Pediatrics, Kyung Hee University Hospital at Gangdong.
Results and Conclusions: Most of chest pain in children and adolescent was idiopathic or musculoskeletal origin (80.5%), followed by respiratory (10.7%), cardiac (4.5%), gastrointestinal (2.9%), psychiatric (1.4%). In case of respiratory disease, pneumothorax (4) and pneumomediastinum (2) was in 6 case (1.2%), and all the patients with pneumothorax had been done chest tube insertion. In such air-leak syndrome, the pain was abrupt, continuous and has short duration after the onset of 1-2 days in older adolescents. In the patients of pneumomediastinum, radiologic diagnosis was not easy without careful searching, therefore caution is required in patients with pain of these characters. In patients with pain of cardac origin, 13 patients had cardiac arrhythmias (56.5%), 8 had congenital heart diseases (34.7%), 2 had coronary aneurysms caused by Kawasaki disease (8.7%). One patient with atrial flutter had symptome of syncope and chest pain only. One should not neglect syncope and further work-up is essential.

P-103
Spectrum of childhood heart disease in Tunisia: rheumatic heart disease or congenital heart disease?
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Introduction and aim: Spectrum of childhood heart disease varies from country to another and may change over time. Through this study we aimed to provide the pattern of childhood heart disease in our public institution.
Methods: All patients under the age of 14 and referred to our department of cardiology in Hedi Chaker Hospital in Sfax for echocardiography, during three years, were included and analyzed. A focus on new referrals was made
Results: During the period of study, among the 1971 patients studied, 1607 were new referrals. In this latter group, the proportion of normal scans was 56%. By far, the most frequent reason of the exam was cardiac murmur, which accounted for 40.8%.In patients with abnormal finding, we counted 539 patients with congenital heart disease and 71 patients with acquired heart diseases. Left to right shunts were the commonest defect in 292 patients (54%), caused by ventricular septal defect in 156 patients, atrio-ventricular septal defect in 41 patients, atrial septal defect in 57 patients and patent ductus arteriosus in 38 patients. Tetralogy of Fallot was the commonest among the cyanotic defects (5.1%) followed by transposition of great arteries (3.7%).
In acquired heart disease, Rheumatic heart disease, dilated cardiomyopathy and Kawasaki disease were the cause in 18%, 29% and 21% respectively.
Conclusion: Heart diseases in Tunisian children are dominated by congenital heart disease. Rheumatic heart disease was not the majority cause of acquired heart disease and was lower than expected. A population based prevalence study is required to determine the full extent of this problem.

P-104
Chromosomal abnormalities in Tetralogy of Fallot do not contribute to increased short term morbidity
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Background: There is an increased incidence of chromosomal abnormalities in TOF particularly 22q11 micro-deletion. Our aim was to determine the incidence of chromosomal abnormalities in our TOF population and assess their impact on short term morbidity.
Method: The medical records over a five year period from January 2007 to December 2011 were retrospectively reviewed. Ninety consecutive patients were included in the study.
Results: Antenatal diagnosis was made in 24.4%. The mean age at surgery was 5.2 months with a mean weight of 5.8 +/- 1.5 kg. The mean hospital stay was 15.1 ± 3 days including an intensive

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care stay of 4 ± 4.1 days. Twelve patients were found to have a chromosomal abnormality out of which 9 (10%) were found to have a 22q11 micro-deletion. Other diagnosis included Trisomy 21, a partial duplication of chromosome 2 and a balanced pericentric inversion of chromosome 1. There was a significant increase in the finding of a disconnected LPA in those with a 22q11 micro deletion. Other co-morbidity included cleft palate (2) and umbilical hernia. Those with chromosomal abnormalities did not have a significantly longer hospital stay (17.5 days vs. 14.8 days, p = 0.39). Multiple regression analysis revealed sepsis and chylothorax as significant factors prolonging hospital stay for the entire cohort, patients with 22q11 micro deletion did not have a significant increased incidence of wound infection or sepsis.
Conclusion: Tetralogy of Fallot is significantly associated with 22q11 micro-deletion, this association does not increase short term morbidity or duration of hospital stay.

P-105
Clinical and echocardiographic predictors of recurrence of subaortic stenosis

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Introduction: Obstruction of the left ventricular outflow tract beneath the aortic valve and resultant sub-aortic stenosis (SAS) could represent an acquired condition, being rarely recognized in the newborn period, but common in infancy and childhood. Although surgery for SAS is very effective, recurrence occurs in up to 55% of patients, necessitating reoperation in many. There are conflicting data regarding factors predictive of recurrence of SAS after surgical resection. This study aimed to determine clinical, anatomical and echocardiograhic pre- and post-operative parameters predictive of SAS recurrence and requirement for further surgery after initial SAS resection.
Methods: Demographic, clinical, anatomical and pre-operative echocardiographic characteristics of 52 consecutive patients with SAS were retrospectively recorded. Type of surgery and immediate post-operative echocardiographic measurements were also recorded and used for the analysis. T-test or the Mann Whitney test were used for parametric or non-parametric variables respectively, to identify differences between patient groups. Multivariate regression analysis was used to determine the predictors for requirement for re-operation.
Results: Recurrence of SAS was more common in females (57.9% vs 27.3%, p = 0.040) and in those who had residual SAS on echocardiography during the immediate post-operative period compared to those without residual SAS (78.6% vs 22.2%, p = 0.001). Patients with recurrent SAS had undergone initial surgery at an earlier age in contrast to those without SAS relapse (7.3 ± 13.1 vs 10.4 ± 13.2 years, p = 0.036). Echocardiographic characteristics of patients who subsequently underwent second SAS resection compared to those without SAS recurrence included higher post-operative peak and mean trans-aortic gradients (81.1 ± 20.6 mmHg vs 69.6 ± 28.7 mmHg, p = 0.021; 16.2 ± 8.4 mmHg vs 10.3 ± 6.7 mmHg, p = 0.007) and lower left ventricular ejection fraction and fractional shortening (71.7 ± 6.4% vs 76.2 ± 6.2%, p = 0.017; 36.4 ± 4.7% vs 40.6 ± 5.3%, p = 0.018). Multivariate regression analysis showed that the presence of residual SAS post-operatively was the single independent predictor for requirement for re-operation after initial SAS resection, adjusted odds ratio (OR) = 9.782 (2.066-46.320, 95% confidence interval).
Conclusions: SAS commonly recurs following surgical resection, but it is hard to predict which patients will be affected. This study aids clinicians regarding future prognosis by identifying risk factors for patients most likely to require further surgery.

P-106
Cardiovascular involvement in children with oncologic pathology and in bone marrow transplanted patients

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Aim: To determine the type of cardiovascular involvement in patients with cancer and in bone marrow transplanted patients, treated in our clinic between 2010-2013.
Methods: A number of 268 patients were diagnosed with different types of cancer, as: acute lymphoblastic leukemia, Hodgkin lymphoma, myeloblastic acute leukemia, nonHodgkin malignant lymphoma. All this patients performed: clinical examination, ECG, cardiopulmonary X-ray, echocardiography. The investigations were done post radiotherapy, post corticotherapy and after anthracycline treatment. Regarding bone marrow transplanted patients, from 161 cases, 125 were autologus and 36 were allogeneic. The cardiovascular examination was before, at the moment and after transplant.
Results: From the total number of patients with cancer, 13 (4.8%) presented pericardial fluid at diagnose. Post radiotherapy, 15 (5.59%) patients presented pericardial fluid, from which two developed cardiac tamponade. Post corticotherapy, 86 (32%) patients developed hypertension and were treated. Regarding the anthracycline group of patient, 18 (6.7%) developed tachycardia and no other cardiac involvement, because the patients were treated after protocols that counted the total amount of anthracycline, to prevent overdose. Carvedilol and coenzyme Q10 were used for cardio protection. Post bone marrow transplant, 36 (22.3%) patients presented cardiovascular involvement, like: toxic cardiomyopathy 4 (11.1%) cases, pericarditis in 4 (11.1%) patients, pericardial tamponade 1 (2.77%) case, atrial fibrillation 2 (5.5%) cases, hypertension 11 (30.5%) cases, venous thrombosis 6 (16.6%) cases and veno-occlusive disease in 8 (22.2%) cases. Radiotherapy and chemotherapy induced pericarditis and tamponade. Tachycardia was the first sign post rapid injection of stem cells. Myocardial reaction was transient post chemotherapy. Central venous catheter and infection were the trigger for atrial fibrillation. After anthracycline therapy, patients were monitorised almost 1 year, to prevent dilated cardiomyopathy or heart failure.
Conclusions: Cardiovascular involvement in oncologic pathology of children is secondary to radiotherapy, corticotherapy, anthracycline therapy and autolog or allogeneic bone marrow transplantation. Pericarditis was secondary to unprotected radiotherapy. Anthracycline treatment did not severely affect the heart, because of protocol counted dosage. Hypertension regressed at the end of the corticotherapy. The cardiovascular involvement in transplanted patients was secondary to the treatment administrated previous the transplant. Stem cell transplantation did not severely affect the children. The cardiology examination is very important in oncologic pediatric patients.

P-107
Coarctation of the aorta presenting as dilative cardiomyopathy: Exception or distinct mode of clinical presentation?

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While neonates with coarctation frequently present with congestive heart failure and critically reduced perfusion of the descending aorta following ductal closure, infants and older children with coarctation are usually oligosymptomatic and are diagnosed due to weak femoral pulses, heart murmur or arterial hypertension. We encountered several infants with coarctation, who presented with dilative cardiomyopathy (DCM) and reviewed our patients to determine, whether this presentation represents a rare exception or is relevant for the differential diagnosis of children with DCM.
101 infants were referred to our hospital for treatment of coarctation between 1/2001 and 12/2013. 24 of these patients were older than 4 weeks. Among them five children with severe coarctation presented at an age of 2.1-5.9 months under the clinical picture of DCM. These children accounted for 5% of those, who were diagnosed with coarctation in the first year of life and 21% of those, who presented in infancy. Echocardiography revealed a dilated left ventricle and markedly reduced function with shortening fractions between 9-19%. The aortic arch was normal in diameter with a circumscribed severe coarctation with some distance to the left subclavian artery. All patients underwent resection of the coarctation within 24 hours. Significant postoperative improvement of the left ventricle resulting in a normal shortening fraction occurred within a median interval of 1.7 months.
Coarctation of the aorta presenting as DCM in infancy has been rarely reported in the literature. In our study however this specific clinical presentation accounted for 21% of infants with discrete coarctation, who became symptomatic beyond 4 weeks of age. The stenosis was difficult to detect due to its distal location, the normal configuration of the aortic arch and the low isthmic gradient resulting from low cardiac output. Indirect hints are bicuspid aortic valve, thickened myocardium and reduced flow in the celiac artery. Following treatment of the coarctation left ventricular function appears to recover completely. Since these patients require urgent treatment differing completely from other forms of DCM, careful examination of the isthmic region should be included in the primary assessment of all infants presenting with DCM.

P-108
Aortic coarctation and infantile hemangioma: coincidence or PHACES syndrome?

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Objective: To determine the prevalence of PHACES syndrome (Posterior fossa malformations, Hemangiomas, Arterial anomalies, Cardiac defects, Eye anomalies, Supraumbilical raphe and/or Sternal pit) in patients with obstructive aortic arch pathology (OAAP) and to achieve more insight in the possible association between infantile hemangiomas (IH) and cardiovascular anomalies.
Study design: Pediatric patients diagnosed between 1999 and 2013 with OAAP in our tertiary referral center were included. Questionnaires focusing on the (past) presence of an IH and other symptoms fitting the diagnostic criteria of PHACES syndrome were designed. Data of deceased patients were analyzed separately.
Results: Questionnaires send to 286 patients with OAAP, were returned by 175 subjects (response rate 66%). In 9 cases an IH was diagnosed. One child met the criteria of PHACES syndrome. This child demonstrated a segmental hemangioma and an atypical interrupted aortic arch with atresia of the left common carotid artery, which fits the complex vasculopathy seen in PHACES syndrome. All other children did not meet the PHACES criteria due to characteristics of IH or aortic arch and thus were thought to be normal distribution. Deceased children suffered from severe congenital anomalies though none of them fitted the PHACES syndrome criteria.
Conclusion: In this retrospective cohort, one child met the PHACES criteria, indicating that PHACES syndrome in AC patients is less common than expected. Our study was not able to provide further evidence for the suggested association between IH and obstructive aortic arch pathology.

P-109
Etiology of hypertrophic Cardiomyopathy in Young childhood

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Introduction: Hypertrophic cardiomyopathy (HCM) in young childhood is a rare and heterogeneous disease. The prognosis is variable and depends upon the underlying disorder. Unfortunately the etiology of HCM remains unclear in the majority of reported cases. This study aims to describe the diversity of etiologies and the prognosis of HCM in infants under the age of one year.
Methods: In this retrospective single center study all patients under the age of one year born from 2007-2012 with cardiac hypertrophy measured by echocardiography (defined as interventricular septum thickness with Z-score >1.64) were included. Patients with underlying congenital heart disease or hypertension were excluded. Diagnosis and clinical follow up were extracted from patient files.
Results: During the studied period 32 children with HCM matching the inclusion criteria were identified. Malformation syndromes (including Beckwith-Wiedemann syndrome, Costello syndrome, Noonan syndrome, Cantu syndrome, Leprechaunism, Bernadelli-Seip congenital lipodystrophy and Trisomy 18) were most frequently diagnosed (31%). Other underlying causes were maternal diabetes mellitus (19%), metabolic disease (6%), sacromeric disease (3%), toxic disease (3%) and congenital hyperinsulinemia (3%). In 34% of the patients no underlying cause of the cardiac hypertrophy was identified. Overall, the underlying etiology of HCM was related to hyperinsulinemia in more than half of the HCM patients. In this studied group 25% of the patients died, all during their first year of life (mean age of death: 4 months). During follow-up HCM normalized in 80% of the surviving patients. Patients diagnosed with malformation syndromes had the highest chance of dying and persistent HCM.
Conclusion: An underlying etiology can be found in 2/3 of infants with HCM under the age of 1 year. Hyperinsulinemia is a likely causative factor in more than half of these patients and may have direct consequences for prognosis and treatment. Infants with HCM surviving their first year of life have a good prognosis as the HCM resolves in most of the cases.

P-110
Doppler echocardiography imaging and prognosis in children with hypertrophic cardiomyopathy

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Background: Assessment of left ventricular (LV) diastolic function by Doppler echocardiography imaging (DEI) has been reported

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to be useful for predicting the prognosis in patients with hypertrophic cardiomyopathy (HCM). The aim of this study was to evaluate the clinical significance of DEI parameters for prediction of cardiovascular events in children with HCM.
Methods: Retrospective analysis of 96 children, mean age 10,1 ± 5,2 yrs (ranged from 1 month to 18 yrs) with HCM diagnosed from 1991 to 2013. The combined end-points were HCM-related death; resuscitated cardiac arrest; appropriate ICD discharges; admission for cardiac arrhythmias; new episode of syncope; and worsening of heart failure symptoms (NYHA class III and IV). In echocardiography mitral inflow was assessed (wave A, E, E/A ratio), left atrial dimension (LAD) in the parasternal long axis view. The values of E, A waves, E/A ratio were considered abnormal if they were lowered or raised in relation to the standards for the age of the patient. LAD was assessed with respect to the patient BSA standards.
Results: During a follow-up period mean 7,9 yrs (ranged from 3 months to 22 yrs) 38 (40%) pts (group I) achieved the combined end-points while 58 (60%) pts (group II) did not. The left atrium was enlarged in 68% pts in gI vs 42% in the gII ($p = 0.027$). Children who experienced cardiovascular events had larger LAD (mean 153% of the average of the standard for BSA) compared with those who did not (mean 123% of the average of the standard for BSA). Analysis of mitral inflow showed that in gI, LV diastolic function was abnormal in 28 (74%) pts vs 28 (48%) pts in gII ($p = 0.0195$). E/A ratio was abnormal in 16 (42%) pts in the gI vs 25 (43%) in the gII. Wave E was abnormal in 23(61%) pts in gI vs 21(37%) pts in the gII ($p = 0.035$) and wave A in 17(45%) children in gI vs 16(28%) in the gII ($p = 0.124$).
Conclusions: (1) Assessment of diastolic function by Doppler echocardiography imaging is useful for risk stratification in children with HCM. (2) Doppler echocardiography imaging parameters should be included into the clinical management of HCM patients.

P-111
Coarctation of aorta, 10 years of clinical epidemiological study, diagnostic and therapeutic considerations

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Introduction: Our study is a clinical epidemiological retrospective analysis of coarctation of the aorta in a 10 year follow-up (2001-2011).
Methods and results: The study includes 201 children, 35,8% girls and 64,2% boys, with an average age of 28,57 ± 49,37 months (0,1-204). They are categorized in 4 age groups: <1 month, 1 month – 1 year, 1 year – 6 years, >6 years. Isolated CoA was found in 62,2% patients; 16,42% preductal and 45,77% postductal. CoA with an additional heart defect was found in 37,81% patients; 15,32% with VSD, 13,93% within a complex heart defect, 5,47% within the Schone syndrome, and 2,49% with a dilatative cardiomyopathy. Clinical symptoms are analyzed in detail according to age groups; tiredness, intense perspiration and dispnoic difficulties are a dominant feature in lower age groups (newborns and infants), while claudications, headaches and epistaxes are typical in older children. In the case of as many as 61 (30.35%) patients diagnosis was missed on the previous cardiological examination. In 20 (9,95%) patients coarctation is found within the known syndromes (Turner, Noonan, Williams Beuren, Ellis van Creveld, partial trisomy 18, fetal valproate syndrome). Echocardiography was performed in all patients, and in 45 (22, 38%) it was the only diagnostic procedure. Altogether, 123 heart catheterizations, 38 MSCTs and 15 MRs were performed. The gradient on the place of coarctation before surgery or emergency procedures measured in 132 (65,67%) patients was 57,99 ± 18,68 mmHg (20-100 mmHg). In 82 (40,8%) patients a bicuspid aortic valve was found. Average age at the time of surgery was 27,92 ± 47,98 months (0,1-204). In 84,1% patients a cardiosurgical intervention was performed, in 54.2% T-T anastomoses, in 15.9% balloon dilatation or stent implantation. Fatal outcome occurred in 1,99% children, all newborns or infants.
Conclusion: Coarctation of aorta is still diagnostic and therapeutic chalenge depending of its different expressions, range of pathological changes on the aortal arch, relation towards other organs (ductus, subclavian arteries), and association with other complex heart defects, including complex anomalies. All these factors determine the clinical picture, diagnostic and therapeutic approach, as well as the course of the illness itself.

P-112
Congenital Coronary Artery Fistulas: Clinical and Therapeutic Considerations

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A coronary artery fistula is a link between one or more coronary arteries with another heart cavity or a segment of systemic or pulmonary circulation. These are very rare anomalies which constitute approximately 0.2–0.4% of all congenital heart defects. Still, they are clinically significant if they are of medium or large size and are manifested with a series of clinical symptoms such as angina pectoris, arrhythmias, myocardial infarction, endocarditis, progressive dilatation, heart failure and cardiomyopathy, pulmonary hypertension, thrombosis of the fistula and formation of aneurysms with possible ruptures.
We present six patients with a coronary arterial fistula, their history, diagnostic procedures and outcomes. Therapeutic closure of coronary artery fistulas is recommended in symptomatic, but also in asymptomatic patients, if there are significant clinical symptoms with roentgenographic and electrocardiographic abnormalities. In recent times transcatheter closure of coronary fistulas has become a possible alternative to surgery and is becoming increasingly used thanks to improved diagnostic possibilities and technology. If possible, interventional closure of fistulas is precisely the method preferred in pediatric patients. Choice of method depends on the anatomy, presence or absence of additional defects, and experience of an interventional cardiologist or a heart surgeon. This work presents two children with a fistula between the right coronary artery and the right ventricle (RV), one child with a fistula between LAD and RV, child with a fistula between the main tree of the left coronary artery (LCA) and RV, one child with a fistula between LCA and the right ventricular outflow tract (RVOT), and one child with a fistula between LCA and the right atrium (RA). The last one (LCA-RA) is not described in the latest classification of anomalies of coronary blood vessels in children based on MSCT coronarography, so we consider our presentation to be a contribution to the new classification. Along with descriptions

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of fistulas and presentations of interventional and cardiosurgical interventions, we are also presenting a rare case of spontaneous closing of the fistula within the first six months and of a reopening of the fistula between the right coronary artery and the right ventricle after six years.

P-113

The association between the head circumference and the central venous pressure in children undergoing the Fontan procedure

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Introduction: High central venous pressure (CVP) may cause communicating hydrocephalus and macrocephaly. We examined if higher pulmonary pressures (PAP) in children undergoing bidirectional cavopulmonary anastomosis (BDCPA) leads to an increased growth of the head circumference (HC) as the central venous pressure (CVP) interferes with cerebral venous drainage.
Methods: Seventy-four consecutive children with single-ventricle physiology (24 HLHS, 11 DILV, 10 DORV, 9 unbalanced AVSD, 7 PA IVS, 5 TA, 4 PA VSD and 4 unbalanced ccTGA) were included in a retrospective analysis. Patients with other causes of a macrocephaly were excluded. CVP and PAP were measured during the routine catheterization before BDCPA and Fontan completion. The occipital-frontal circumference was measured using a flexible non-stretchable measuring tape at the same time and at the routine clinical visits and compared with longitudinal age dependent percentiles in normal children measured in our own institution.
Results: Median age at BDCPA and Fontan were 4.8 (1.6-12) and 27.9 (7-40.6) months respectively. HC was more than 1 SD below 50th percentile at the time of the BDCPA and showed significant growth between pre-BDCPA- and pre-Fontan catheterization (7 (0-100) vs. 36 (3-56)th percentile, $p < 0.001$), while PAP decreased significantly (14.8 ± 2.7 vs. 13.2 ± 2.1 mmHg, $p = 0.012$). There was no correlation between PAP and HC pre-Fontan ($R2 = 0.001$). Children with lower differences in CVP pre-BDCPA and PAP pre-Fontan showed increased growth of HC. Higher cerebral venous drainage may explain this weak correlation ($R2 = 0.19$, $p = 0.012$).
Conclusions: Moderately elevated CVP in children with single ventricle physiology does not lead to macrocephaly. Between BDCPA and Fontan, HC increased significantly and achieved values close to norm for age. The lack of direct correlation between PAP before Fontan and HC may be explained by a catch-up growth of HC in patients with better pulmonary vascular bed. Further studies with focus on high PAP are needed to exclude or prove a direct correlation.

P-114

Quality of life and social situation following Fontan operation (FO) vs. correction of tetralogy of Fallot (TOF)

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Introduction: Nowadays patients with a single ventricle as well as those with tetralogy of Fallot reach adulthood without cyanosis. Nevertheless, health and psychosocial problems emerge in the long-term. Therefore we conducted a comparative analysis of cardiopulmonary performance, quality of life and social situation in patients (pts) after FO and TOF correction.
Patients: 47 adults ($m = 57.4\%$) after FO (median 21 years) and 97 after TOF correction (median 24 years) answered standardized questionnaires from 2006-2012 on their quality of life (SF-36) and social situation. Spiroergometric tests performed around the same time an the maximal oxygen consumption (VO2max) was compared between patients after FO ($n = 27$) and TOF ($n = 25$).
Results: VO2max was lower for FO pts. than for TOF pts. (median 51% vs. 64% of the norm) and correlated strongly ($p < 0.005$) with FO with "physical limitations", "pain" and "vitality" and with the patient's own health satisfaction ($r = 0.597$, $p = 0.002$) and life satisfaction ($p = 0.014$). On a scale from 1 to 7 high levels of satisfaction were reported (mean health: 5 for FO vs. 4.8 for TOF/mean life: 5.7 for FO vs. 5.24 for TOF).
Despite differences in social situation (long-term relationships: 47.8% of FO vs. 58.3% of TOF pts/ own children: 4.4% of FO vs. 33.3% of TOF pts), the levels of psychological well-being in FO pts were similar to those in TOF pts ("role emotional" FO $83.3 + 33.3$ vs. TOF $82.1 + 32.8$; "mental health" FO $69.4 + 17.5$ vs. TOF $73.4 + 14$ and "vitality" FO $55.6 + 21.4$ vs. TOF $54.5 + 20.1$).
Conclusion: Despite physical and social limitations adults after FO achieve a high quality of life and life satisfaction. In many dimensions of the SF-36, particularly in the areas of psychological well-being, FO and TOF pts. show only small differences. It appears that quality of life and life satisfaction depend less on performance capacity than on attitude.

P-115

The advantage of a shared care service in improved access to pulmonary hypertension therapy for adult congenital heart disease associated pulmonary hypertension

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Background: Historically, pulmonary hypertension management has been delivered in the UK by the National Pulmonary Hypertension service centres (NPHS). However, many regions are geographically removed from such centres making it difficult for patients with adult congenital heart disease associated pulmonary hypertension (ACHD-PH) to receive therapy. This is especially true if there are associated learning difficulties such as Trisomy 21.
Methods: We reviewed our clinics run as a shared care with an NPHS centre since 2008 to current date to determine the effect on access to therapy. We measured the mileage and uptake of therapy per head of population for remote areas of the UK covered by our clinic.
Results: 235 new patients were seen, with mean (sd) age 45 (19) years, 98 (42%) were male. Almost all had ACHD-PH with only 7.9% of patients with chronic thromboembolic pulmonary hypertension and 7.1% with idiopathic pulmonary arterial hypertension. 29.9% had learning difficulties. A full range of diagnostic facilities were present locally, the patients rarely having to travel to the NPHS centre. For therapy, 54.3% have been treated with single medication, 40.94% dual therapy and 13.4% on triple therapy compared to 59.4%, 36.5% and 14.9% respectively in the national database. A higher proportion of those on therapy (55.1%) received Endothelin receptor

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antagonist (ERA) therapy compared to 32.5% nationally and 58.0% receiving phosphodiesterase inhibitors vs 63.0% nationally. The number of our patients on therapy per 100,000 population increased from 0.7 to 4.7. The average mileage for clinic attendance was 48.8 miles compared with 163.7 miles if the journey had been made to the nearest NPHS centre, saving an average of 115 miles (3 hours of travelling for each clinic) and the cost of taxi fares to the NHS of £108,000.
Conclusion: We have shown that our clinic achieves the national average for the number of patients on therapy. The increased proportion of patients on ERA is due to the increased numbers of patients with ACHD-PH. The clinic improves access to disease modifying therapy and improves the ease of access to clinic for patients with learning difficulties whilst saving the NHS significant costs.

P-116
Pulse Wave Analysis in Adult Patients with Congenital Heart Disease
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Introduction: Based on advance of surgical procedure and medical therapy, most patients with congenital heart disease can be expected to survive into adulthood today and it means that the patients are faced with problems associated with aging. The aging process of cardiovascular system starts with fracture of aortic elastic lamellae and results in elevation of pulse wave velocity (PWV) and enhanced pressure wave reflection. In some clinical course in congenital heart disease, similar changes had been reported. The purpose of the present study is to clarify the risk factors of the elevation of PWV and the enhanced pressure wave reflection.
Methods: Brachial-ankle PWV (baPWV) and radial augmentation index (rAI) were examined in 99 patients with congenital heart diseases over 20 years of age. The factors affect the two parameters were analyzed. The patients' age was 36.0 ± 14.5 (20-76), and male/female was 63/36. Thirteen patients demonstrated oxygen saturation under 90%. NYHA I: 67, II: 26, III: 6. The systolic blood pressure was 118.1 ± 19.2 mmHg and 13 patients were hypertensive.
Results: The rAI was 77.1 ± 19.1% and it had a significant correlation with the history of aorto-pulmonary shunt ($t = 4.194$; $p < 0.0001$), age ($t = 4.091$; $p < 0.0001$), height ($t = -3.580$; $p = 0.0010$) and the history of direct aortic surgery ($t = 2.253$; $p = 0.027$). Forty-four patients (44.4%) demonstrated high rAI (>1SD of age- and gender matched control) and the determinants of the elevated rAI were the history of aorto-pulmonary shunt (odds ratio, 21.319; 95% confidence interval, 5.467-83.142; $p < 0.0001$) and direct aortic surgery (4.183; 1.376-12.721; $p = 0.012$). The baPWV was 1227 ± 339 cm/s and it had a significant correlation with reflected blood pressure ($t = 6.764$, $p < 0.0001$), age ($t = 4.216$, $p < 0.0001$), male ($t = 2.823$, $p = 0.006$). Twenty-three patients (23.2%)demonstrated high baPWV (>1SD of age- and gender matched control) and the determinants of the elevated baPWV were reflected blood pressure (1.122; 1.067-1.181; $p < 0.0001$) and administration of renin-angiotensin-aldosterone system blockers (0.109; 0.013-0.020, $p = 0.019$).
Conclusions: The aorto-pulmonary shunt and the direct aortic surgery could enhance the pressure wave reflection and the enhanced pressure wave reflection could elevate the PWV. The blockades of renin-angiotensin-aldosterone system might prevent the elevation of the PWV.

P-117
Treatment of coronary anomalies in adults
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Objectives: Prevalence of coronary anomalies is unknown. Symptoms in adults occur rarely prior to the 4th decade except in competitive sports; incidence of these is also unknown. Sudden cardiac death has been reported. Conventional treatment with CABG (intern thoracic arteries or veins) has proven ineffective.
Methods: Seven patients (4 male, 48 ± 14.4 years (24-68, one <40 years) with variable symptoms of angina had normal coronary angiography except for the difficulty to inject into the RCA (6) or the LAD (1). 3D-MRI/CT scan showed an atypical origin of the RCA (6) or LAD (1) of the respective other sinus crossing the commissure between the left and right cusp running between the aorta and pulmonary artery. Surgery addressed all three components of the anomaly (orifice without a funnel, intramural course crossing the commissure, course between pulmonary artery and aorta). Surgical steps under CPB were: aortotomy, unroofing of the ostium distally to the pulmonary artery throughout the intramural course and additional in-situ RIMA Bypass to RCA or LIMA bypass to LAD, respectively. The latter were performed to protect against early occlusion or thrombosis of the reconstructed orifice.
Results: Intraoperative course was uneventful. Patency of the reconstruction was proven by increase of the RIMA/LIMA flow in ultrasonic flow measurement under transient occlusion of the proximal RCA/LAD. All patients were extubated on the day of surgery. One patient had sudden cardiac fibrillation on the ICU with emergent ECMO implantation. Cardiac cath. revealed no filling of the RCA, so that emergent revision of the distally dissected RITA-bypass was performed. The patient was weaned from ECMO and recovered completely being discharged with normal (60%) LV-function. At follow-up of 19.3 ± 11 months (7-36 months), all patients are clinically well without cardiac symptoms. Control CT in two patients showed an occluded ITA graft and a 'normal' reconstructed coronary ostium.
Conclusion: Angina in young or mid-aged patients without coronary artery disease may be caused by coronary anomalies. Anatomical reconstruction with protective bypass grafting is a convincing surgical strategy with excellent results protecting against sudden cardiac death. However, indication is still intriguing as neither prevalence nor incidence of clinical events are known.

P-118
Usefulness of B-type natriuretic peptide and pro-inflammatory interleukin 6 levels to predict adverse cardiac events in adolescents and adults with congenital heart disease
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Introduction: Natriuretic peptides, neurohormones and inflammatory cytokines, all products of the failing heart, are present in elevated concentrations in the circulation of patients with congenital heart disease. Their prognostic value, though, remains unknown.

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Methods: Sixty consecutive clinical stable patients, mean age 28.9 ± 11.4 years old, 53% male, with various forms of CHD, were recruited from a tertiary center. B-type brain natriuretic peptide (BNP) and interleukin 6 (IL-6) were measured prospectively and the patients were followed for major cardiovascular events (MACE), including death or hospitalization for 5.1 ± 1.1 years. Cox proportional hazard ratio analysis was used to determine the relation of BNP and IL-6 concentrations to all cause mortality. Receiver operating characteristics curves were used to determine the cut-off values for BNP and IL-6 that would best predict all-cause mortality.
Results: Most patients were symptomatic (48.3% with NYHA II and 36.7% with NYHA III), seventeen (28.3%) of them were cyanotic at rest and sixteen (26.6%) had ventricular dysfunction. Mean plasma concentrations of BNP and IL-6 were 106.6 ± 98.6 pg/ml and 2.4 ± 2.6 pg/ml respectively. Twenty-two patients (36.6%) experienced a MACE during the follow-up period, among them 8 patients (13.3%) died. Both BNP and IL-6 were proved to be strong predictors of survival (hazard ratio for every pg/ml unit increase, 2% and 12.9% respectively, 95% confidence interval 6.5% to 19.8%, $p < 0.05$) respectively. A BNP value >241 pg/ml predicted MACE with a sensitivity of 65.38% and a specificity of 73.53% (Area Under the ROC Curve, i.e. AUC = 0.693, $p < 0.0001$). An IL-6 value >1.54 pg/ml predicted MACE with a sensitivity of 61.53% and a specificity of 73.53% (AUC = 0.627, $p < 0.0001$). No patient with a BNP level <241 pg/ml and IL-6 level <1.54 pg/ml died during the follow-up period.
Conclusions: Both BNP and IL-6 levels strongly predicted MACE in symptomatic ambulatory patients with various forms of congenital heart disease, during mid-term follow up and could be used as easy applicable risk stratification markers in this population.

P-119

Supravalvular pulmonary stenosis (SVPS) in patients with transposition of the great arteries (TGA) after arterial switch operation (ASO)

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Introduction: ASO is currently the method of choice for transposition of the great arteries. Despite excellent early and long-time survival, there are still important complications in postoperative period like SVPS, neoaortic insufficiency and coronary ostial stenosis. SVPS remain the most frequent indication for reoperation and reinterventions after ASO irrespective of technique of neopulmonary artery reconstruction. The aim of this study was to establish the frequency of significant SVPS in patients with TGA after arterial switch procedure, its pattern during follow up, risk factors and necessity of reinterventions and reoperations.
Methods: We reviewed all 665 arterial switch procedures performed between years 1991 – 2013 in Cardiosurgery Department of Polish Mother's Memorial Hospital including patients with simple TGA (412pts-62%), TGA associated with VSD (175pts-26%), TGA with aortic arch anomalies (59pts-9%) and 2 stage operation with pulmonary artery banding prior to the ASO (19pts-3%). The SVPS gradient was acquired from echocardiographic examinations performed during routine control follow-up visits in the Cardiology Department.
Results: The overall mortality was 7,2%; average clinical follow–up duration for survivors (617pts) was 10,2 years. In majority of the cases neopulmonary trunk reconstruction was performed with direct pulmonary artery anastomosis (584pts-88%), in the remaining cases the risk of coronary arteries compression has forced to use the pericardial patch for pulmonary reconstruction (81pts-12%). The significant SVPS (flow speed over 2,5 m/s; PG > 25 mmHg) was observed in case of 54 patients (8%); 7 patients required reintervention because of significant SVPS (PG >40 mmHg), 1 patient had balloon plasty of SVPS twice. None of the patients from our study group required reoperation because of pulmonary stenosis. Among analyzed risk factors patch reconstruction ($p < 0{,}001$), non-facing commissures ($p = 0{,}02$) and pulmonary banding ($p < 0{,}001$) were correlated significantly with significant SVPS (cox-regression model).
Conclusion: Although the risk of significant SVPS after arterial switch operation is relatively low, relief of supravalvular pulmonary stenosis represents about half of indications for postoperative reinterventions. Lowest risk of SVPS occurs in patients with anatomic correction performed during the neonatal period with direct pulmonary artery anastomosis.

P-120

Infective endocarditis in patients with ventricular septal defect

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In the context of recent change in guidelines for prophylaxis of infective endocarditis (IE), the objective of this study was to assess the features and outcomes of IE in children and adults with non-significant ventricular septal defect (VSD).
Methods: A retrospective analysis of records of patients with non-significant VSD. Clinical, echocardiographic and microbiological data, and outcomes were assessed.
Results: From 1980 to 2013, 57 IE occurred (1 to 4 per year), in patients aged 14.2 ± 11.3y (med12.1), 29males (51%) and 39 were <18y of age. VSD was membranous in all cases, isolated (39 = 68.5%) or associated with mild aortic regurgitation or pulmonary stenosis. VSD was native in 39 (68.5%) and not diagnosed before IE occurred in 4cases (7%). The cause of infection was unknown in 36% of the cases, while 23% were from dental, 13% from cutaneous, 9% from ENT or digestive origin, and 19% occurred in the early postoperative course of patch closure, i.e. 81% of the cases occurred in native mild VSD. Streptococcus from dental origin was the most frequent causal agent (54.5%), staphylococcus was found in 35% of cases, Gram-negative bacillus in 3.5%. Hemocultures were negative in 7% of the cases. Vegetation was the most frequent echographic lesion, and located either on VSD, and/or tricuspid valve and/or RV free wall and/or pulmonary valve. Aortic valve location occurred in 8cases. Embolic event occurred in 28cases (49%): multiple pulmonary embolia in 21 (37%), systemic embolia in 6. Eighteen patients were operated (31.6%): early surgery in 11 (19.3%), delayed patch closure in 7. Six patients died (10.5%). Death was not related to early surgery. FU was 13.4 ± 11.2y (med 10.2y).
Conclusion: Infective endocarditis can severely impair prognosis of mild membranous VSD and dental events are the most frequent origin of infection. Despite recent recommendations, preventive surveillance and management of any dental lesions are probably to be emphasized in these patients.

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P-121

Long term follow-up after biventricular repair of pulmonary atresia with intact ventricular septum and critical pulmonary valve stenosis

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Long term follow-up of repaired pulmonary atresia with intact ventricular septum (rPAIVS) and critical pulmonary valve stenosis (rCPVS) is not well described. Residual lesions such as atrial septal defect, pulmonary and tricuspid regurgitation (PR and TR) may lead to right ventricle (RV) enlargement. We sought to determine long term follow-up of patients with rPAIVS or rCPVS.

Methods: Retrospective study of patients with biventricular rPAIVS or rCPVS attending our centre. Cardiac magnetic resonance (CMR) imaging was performed using standardized protocols. Numbers are expressed as median [interquartile range]. Pearson correlation analysis and X^2 test were used to assess the relationships between different parameters.

Results: Eleven patients were studied at a median age of 13.2 years [10.3-16.2]. Two patients had previous palliation. Repair was a transannular patch in 4 patients and a percutaneous dilatation in 7. Four patients had atrial septal defect requiring surgery. Age at repair was 10 days [2.5-22.7] and delay between repair and CMR study was 13.1 years [9.9-16.2]. RV was dilated 9 patients 133 ml/m^2 [110-164] of which 6 patients had decrease RV ejection fraction. RV volume, ejection fraction and QRS duration were normal in 2 patients. More than mild TR was present in 7 patients. All but 1 patient had PR (32% [30-39]). Late gadolinium enhancement was found in 3 patients, at infundibular level in 2. All patients had normal left ventricle volume and function. QRS duration was $\geqslant$120 ms in 5 patients with right bundle branch. RV dilatation was associated with age at CMR ($r = 0.62$, $p = 0.04$), decrease RV ejection fraction ($r = 0.78$, $p = 0.006$), as a trend with TR ($r = 0.57$, $p = 0.06$) but not with PR ($r = 0.38$, $p = 0.2$) or late gadolinium enhancement ($r = -0.26$, $p = 0,5$). QRS duration was not associated with the type of repair, the presence late gadolinium enhancement or RV dilatation ($p = 0.8$, $p = 0.4$ and $p = 0.5$ respectively) but was associated with RV ejection fraction ($r = -0.7$, $p = 0.02$).

Conclusions: RV dilatation, decrease RV ejection fraction and QRS enlargement are common in rPAIVS and rCPVS. Mechanisms of RV dilatation and decrease RV function appear to be multifactorial. T1 mapping studies should be performed. Determining the optimal timing for pulmonary valve replacement and tricuspid valve surgery is challenging.

P-122

Outcomes and long-term results of complete atrioventricular septal defect repair in infants with Down syndrome

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Objectives: A retrospective cohort study was made as well as a comparative analysis of the immediate (up to 30 days) and long-term (56 ± 15; 12-75 months) results of the repair of complete atrioventricular septal defect (CAVSD) in infants with normal karyotype/chromosome (NK) set and with Down syndrome (DS).

Methods: Surgical correction of congenial heart disease during the first year of life was performed on 593 children (8.3%) with DS in the Bakoulev CCVS. Of this number, 349 infants aged 4.8 ± 2.5 months were diagnosed with atrioventricular septal defect. CAVSD occurred in 279 infants. According to Rastelli's classification, CAVSD of type A occurred in 71% (198/279); of type B, in 14% (40/279); and of type C, in 15% (41/279) infants. CAVSD repair has been performed on 163 patients with DS using the single-patch ($n = 40$) and the two-patch ($n = 123$) methods. The control group consisted of 214 infants aged 6.49 ± 3.03 months with CAVSD and NK.

Results: In infants with DS abnormalities of the left AV valve (doubling of the mitral valve, single papillary muscle, leaflet or chordal dysplasia, etc.) occur as statistically significant (8%DS vs 12%NK; $p < 0.05$) which is rarer than in NK children. The presence of DS increases the risk of complications (mainly in the respiratory area) in the early postop. (48%DS vs 63%NK; $p < 0.05$) and significant co-morbid conditions in the long-term period of observation. Squeal structures in groups were differentiated. The early postop. in the DS group was characterized as rarer with high-class heart failure (14%DS vs 37%NK; $p < 0.05$). CAVSD in the DS group was presented preferentially in the respiratory system combined with infective pathology with basic immunodeficiency (21%DS vs 8%NK; $p < 0.05$), which was delimit the postoperative pulmonary ventilation time (DS 5.1 ± 2.8 days vs NK 1.7 ± 0.8 days; $p < 0.05$).

Conclusions: The infants with DS and CAVSD, who underwent surgical repair during the first year of life, have a good prognosis. The presence of chromosomal imbalance in them significantly increases the risk of severe co-morbidities that has a significant impact on the duration of the recovery period, as well as the duration of their life even after successful correction of CHD. For these patients, individual training programs are particularly important.

P-123

Increased Arterial Stiffness in Patients with Congenital Heart Disease – A Cross Sectional Study of 1106 Patients with Various Congenital Heart Disease

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Objective: Multiple studies have demonstrated the predictive value of aortic stiffness parameters like central systolic blood pressure (cSBP) and augmentation Index (AI) for cardiovascular events. Since some studies suggested that there is an increased aortic stiffness in patients with congenital heart disease (CHD), this study aims to investigate cardiovascular stiffness of patients with CHD by oscillometric measurement.

Patients and Methods: From June 2011 to November 2013, we prospectively examined 1106 consecutive patients with various CHD (27.4 ± 12.1 years, 459 female) referred for cardiopulmonary exercise testing (CPET) in our institution, and 332 healthy volunteers (29.6 ± 18.6 years, 173 female). CSBP and AI was estimated in supine position after 5 minutes rest using the oscillometric Vicorder device (SMT medical, Würzburg, Germany). Afterward patients performed a CPET.

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Results: CSBP was higher in patients with CHD in comparison to healthy peers (CHD: 119.1 ± 14.1 mmHg, $p < .001$ vs Healthy: 115.3 ± 13.2 mmHg, $p < .001$). This was also observed for AI (CHD: 16.1 ± 9.0% vs Healthy: 14.1 ± 10.5%, $p < .001$). Mean peak oxygen uptake in patients with CHD was 30.5 ± 10.0 ml/min/kg or 84.8 ± 22.2%predicted, respectively. After correction for age and body mass index, higher AI in patients with CHD was associated with lower peak oxygen uptake ($r = -.118$, $p < .001$) and vice versa, higher cSBP with higher peak oxygen uptake ($r = .150$, $p < .001$).
Conclusions: Central blood pressure and augmentation index is increased in patients with CHD. A better understanding of pathophysiologic mechanisms, genetic predisposition, and the role of surgical aortic scars, implanted conduits, stents or patches is needed in this cohort to define the predictive value.

P-124
Noninvasive assessment of liver function in adults with congenital heart disease (ACHD) by transient elastography (Fibroscan), Acoustic Radiation Force Impulse Imaging (ARFI) and biochemical markers
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Objectives: ACHD may have hemodynamic features, which cause liver congestion, fibrosis or cirrhosis. This study aimed to assess in ACHD the impact of ultrasound based procedures (USBP) (i.e.: Fibroscan, ARFI), and specific biochemical markers on non-invasive staging of liver fibrosis/cirrhosis (LF/LC).
Methods: Patients after atrial switch operation (ASO), after Fontan-Operation or with Eisenmenger syndrome (ES) were studied prospectively in 2 tertiary care centres (Munich, Berlin) to assess hepatic damage noninvasively using clinical data, Fibroscan, ARFI, and abdominal sonography. Blood samples were taken for detailed laboratory analysis, including conventional liver tests (the aspartate aminotransferase (AST)-to-platelet ratio index (APRI), the FIB-4 test and biochemical fibrosis markers, such as hyaluronic acid.
Results: 50 adults (27 after ASO, 10 after Fontan procedure, 13 ES; median age 33,5 years; 18 female) were enrolled. As a result, 16 patients (8 ASO, 8 Fontan) showed signs of hepatic damage in USBP, while laboratory data were altered in >50% of all enrolled cases. Interestingly, 13 of 16 patients with increased hepatic stiffness in the USBP had normal biochemical markers of LF/LC. Overall, regarding imaging and laboratory analysis, 62% (31/50) patients had signs of liver pathology. According to USBP, 34% (17/50) seem to have LF, and 28% (14/50) are suspect to have LC (thereof most with Fontan hemodynamics).
Conclusions: Adults after ASO, Fontan-Operation or with ES are prone to develop liver congestion, fibrosis and cirrhosis. Noninvasive USBP, such as Fibroscan and ARFI, are useful noninvasive methods for early detection of liver stiffness in ACHD. Biochemical fibrosis markers were not indicative for LF/LC in these patients.
For targeted screening of hepatic involvement in CHD, it seems to be crucial that not a single analysis, but several complementary methods, such as ARFI, Fibroscan, abdominal sonography and liver serotests, are performed. Only combined consideration of such noninvasive procedures lead to a meaningful diagnosis of hepatic damage. As special expert knowledge is essential, patients should be transferred for diagnosis to centres of excellence, where congenital cardiologists and hepatologists cooperate.

P-125
Plasma sodium levels predict exercise capacity in postoperative adults with congenital heart disease
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Objectives: Hyponatremia is closely associated with an adverse outcome in adults with congenital heart disease (ACHD) and may be a reflection of the activated neurohormones.
However, its association with exercise capacity remains unclear in those patients.
Methods: We prospectively studied 292 consecutive clinically stable postoperative ACHD patients, including 181 patients with biventricular and 112 with Fontan circulation (18 to 62, median 25 years) between December 2005 and October 2013. We measured plasma sodium (Na) levels and compared those with neurohormonal factors (plasma levels of brain natriuretic peptide, norepinephrine, and plasma renin activity; BNP, NE, and PRA, respectively), hemodynamics (central venous pressure; CVP, cardiac index; CI and systemic ventricular ejection fraction; SVEF) and the diuretic dose (furosemide dose/kg/day). We also measured peak oxygen uptake (pVO2, ml/kg/min) during cardiopulmonary exercise testing and expressed as %predicted value: %).
Results: Hyponatreimia (≤137 mEq/L) was observed in 59 patients (20.2% of the total patients) and was associated with high levels of BNP, NE, PRA, impaired hemodynamics (low SVEF and high CVP), and high dose of diuretic use ($P < 0.05$). The Na levels were positively correlated with %pVO2 ($r = 0.32$, $p < 0.0001$).
Conclusions: Hyponatremia is relatively common in ACHD and predicts the low exercise capacity. Thus, hyponatremia is a simple and useful marker of evaluating heart failure severity in ACHD that reflects the neurohumoral activation, impaired hemodynamics and lower exercise capacity.

P-126
Abnormal Menstrual Patterns in Women with Congenital Heart Disease
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Background: Because of advances in treatment, most women with congenital heart disease (WCHD) have reached childbearing age. Recently, the influences of CHD on ovarian function with subsequent menstrual abnormalities have been described.
Objective: To investigate major menstrual disorders, age at menarche and menstrual cycle, and the determinant factors in WCHD.

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Methods: From June 2011 to November 2013, after exclusion of patient with chromosomal disorders, 143 consecutive WCHD at National Cerebral and Cardiovascular Center (median age 27 years, ranged 16 to 77) completed a questionnaire about their menstrual patterns. Clinical variables included physique index (height, weight, body mass index: BMI, body fat percentage), diagnosis (cyanotic, non-cyanotic), New York Heart Association (NYHA) class, arterial oxygen saturation (SpO2), atrial and brain natriuretic peptides (ANP and BNP, respectively), peak oxygen uptake (PVO2), serum sex hormone levels, and thyroid function (TSH, fT3, fT4). We analyzed data using the JMP statistical program.
Results: The mean age of menarche was 13.2 ± 2.11 years. Precocious (<10 years) and delayed menarches (>15 years) were shown in one and 9, respectively. The mean age at menarche was older in the cyanotic WCHD than those without cyanotic (13.9 ± 0.2 vs. 12.1 ± 0.3 years, $p < 0.0001$). On univariate analysis, the delayed menarche was associated with greater NYHA class ($p = 0.02$), lower SpO2 ($p < 0.0001$), high levels of elevated ANP and BNP ($p = 0.01$, $p = 0.04$), low PVO2 ($p = 0.002$), low levels of testosterone ($p = 0.05$), high levels of TSH ($p = 0.003$), and body height ($p = 0.05$). Menstrual cycle disorders were seen in 58 WCHD (n = 40.8%), consisting of primary amenorrhea (n = 3, 2.1%), secondary amenorrhea (n = 8, 5.6%), polymenorrhea (n = 13, 9.2%), oligomenorrhea (n = 19, 13.3%), and the irregularity (n = 15, 10.6%). On univariated analysis, the prevalence of cycle disorders was significantly higher in the cyanotic WCHD than those without cyanotic diagnosis.
Conclusion: WCHD have a high prevalence of major menstrual disorders. The high proportion of menstrual disorders in WCHD suggests that ex-hypoxic hemodynamics in their childhood has a significant impact on the later ovarian function.

P-127
Quantitative assessemnt of right ventricular function by speckle tracking imaging in Eisenmenger syndrome

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Background: Pulmonary arterial hypertension (PAH) is characterized by progressive pulmonary vascular remodeling and increased right-sided cardiac pressures, leading to right heart failure. Speckle-tracking echocardiography (2DS) has emerged as a reliable noninvasive technique for assessing global and regional RV function in PAH. Eisenmenger syndrome (ES) differs significantly from other types of PAH in its physiology and prognosis. 2DS data are lacking in this specific population.
Aim: To assess the RV function by 2DS in ES and determine whether there are differences with other types of PAH.
Results: Clinical and echocardiographic variables were assessed in 14 consecutive patients with ES (mean age 39 ± 12), and 15 patients with other types of PAH (mean age 61.1 ± 12). 17 Patients were in functional class (FC) II (10 ES) and 11patients were in FC III (4 ES). No difference was found between the 2 groups regarding 6MWTD (380 ± 82 vs 384 ± 149, $p = 0.93$). mPAP and PVR assessed by right heart catheterisation were higher in the ES group (71.7 ± 13 mmHg vs 41.6 ± 9 mmHg, $p < 0.01$ and 12.9 ± 2 vs 8.3 ± 3 UWood, $p = 0.05$).
Correlation between 2DS and conventional parameters was good (: (TAPSE [$r^2 = -0.91$], FAC [$r^2 = -0,89$], RA/LA surface ratio [$r^2 = -0.88$], peak systolic velocity ([$r^2 = -0.84$], excentricity index [$r^2 = -0.81$], Tei index [$r^2 = -0.81$]). No statistical difference was found between ES and other types of PAH regarding conventional parameters except for TAPSE ($p = 0.02$, FAC $p = 0.13$, S' $p = 0.77$)), whereas 2DS showed a markedly less impaired RV function in the ES group (-18.87 ± 1.66% vs -13.82 ± 2.89%, $p < 0.001$), even when analysis was adjusted to FC in each group (FCII: -20.4 ± 2.5 vs -17.8 ± 4%, $p = 0.12$; FC III: -15.6 ± 1 vs -9.3 ± 2%, $p = 0.003$). 2DS segmental analysis of the RV free wall showed more pronounced difference in the apical segments ($p < 0.001$) in comparison with basal segments ($p = 0.57$) between the 2 groups, emphazing the importance of RV apical function in PAH.
Conclusion: 2DS provides a new tool to quantify global and regional longitudinal RV function in ES. Despite additional cyanosis-related comorbidities, the specific pathophysiology and hemodynamic conditions of ES are associated with relatively preserved RV systolic function.

P-128
MAPSE and TAPSE values in Healthy Children and Adolescents

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Objectives: Tricuspid annular plane systolic excursion (TAPSE) and mitral annular plane systolic excursion (MAPSE) are two echocardiographic measure to assess the right and left ventricular longitudinal myocardial function in adults and children. In recent years some investigators reported reference values and z-scores of MAPSE and TAPSE in children. These studies also found positive correlation between age and body surface area (BSA) with MAPSE and TAPSE. In the current study, we aimed to evaluate right and left ventricular systolic functions from infant to adolescents period. And to propose refence values.
Methods: We prospectively evaluated MAPSE, TAPSE, left ventricular ejection fraction (LVEF) and fractional shortening (FS) by two dimensional echocardiography in 1241 healthy children (age day 1 to 15 years). We determined the effects of age and BSA on MAPSE values and a possible correlation of MAPSE values with LV ejection fraction values.
Results: The MAPSE and TAPSE values range from 0.72 cm and 1.13 cm in term neonates from 1.76 cm and 2.74 cm in 15-year-old adolescents respectively. While, the MAPSE and TAPSE values showed a positive correlation with age ($r = 0.86$, $p < 0.001$ and $r = 0.65$, $p < 0.001$), there were no correlation was found with BSA. In all age groups, LVEF values showed positive correlation with FS values. But we could not find any correlation between MAPSE and LVEF. The regression equation relating age and MAPSE and TAPSE are: MAPSEpred = 9.162+ (Age (year) × 0.64), TAPSEpred = 14.55 + (Age (year) × 0.933).
Conclusions: In this study, the MAPSE and TAPSE values were calculated by age, and percentile charts were established to serve as reference data for patients with congenital heart disease.

P-129
Evaluation of Right Ventricular Function in Patients Operated for Tetralogy of Fallot by 2D Strain: Comparison with Magnetic Resonance Imaging

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Aim: Right ventricular (RV) function is a one of the major prognostic factor in patients with operated tetralogy of Fallot (TOF). Two-dimensional speckle tracking (2DST) is a relatively new and objective echocardiographic technique to quantify myocardial strain, providing comprehensive information on ventricular myocardial contractility. In this study we aimed to evaluate RV systolic functions in patients with operated TOF by conventional, Tissue Doppler (TD) and 2DST echocardiography. We also aimed to investigate possible correlation between RV systolic functions and cardiac magnetic resonance imaging (CMRI) parameters.
Methods: A prospective controlled study was conducted including patients operated for TOF. RV functions were evealuated in patients and controls at transthoracic echocardiogram (Echo) by measurement of RV fractional area change (RVFAC), tricuspid annular plane systolic excursion (TAPSE), velocity of S-wave (Sa) and isovolumic acceleration (IVA) at tricuspid annulus with TD. Additionally, RV global longitudinal systolic strain using 2D-STE was performed in patients and controls. Results of patients were compared to RV indexed end-diastolic volume (EDV), indexed end-systolic volume (ESV), and RV ejection fraction (EF) on CMRI.
Results: Twenty-seven patients (17 male) aged 15-34 years (mean $22.7 \pm 6,7$) and twenty-seven healthy controls age and sex matched were included. Parameters of RV systolic function were significantly lower in patients compared to controls (RVFAC $37 \pm 6\%$ versus $50 \pm 8\%$, TAPSE 18.6 ± 2.4 versus 24.4 ± 2.3 mm, Sa 10.5 ± 2.6 versus 12.7 ± 2 cm/sec, IVA 1.93 ± 1.02 versus 3.75 ± 0.87 m/s^2, all $p < 0.05$). Mean RV EF value on CMRI is $50.6 \pm 8.7\%$. Global RV systolic strain value was significantly lower in patients compared to controls ($-17.3 \pm 3.1\%$ versus -25.68 ± 3.03). Global RV 2D strain value was correlated well with RV EF and ESV on CMRI (respectively $r = 0.60$, $p < 0.05$, $r = 0.55$, $p < 0.05$). Feasibility, intra- and inter-observer reproducibility of 2D strain weas adequate.
Conclusions: Speckle tracking is a promising method to estimate RV systolic function in patients operated for TOF.

P-130
Evaluation of cardiac functions in the children with type 1 diabetes mellitus

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Introduction: Several studies have pointed out the existence of cardiac dysfunction in patients with type 1 diabetes mellitus (DM) even in the absence of ischemic, valvular or hypertensive heart disease. The purpose of this study is aimed to evaluate cardiac dysfunction and the relationship between severity of disease and degree of cardiac dysfunction in children with type 1 DM.
Methods: In this prospective study, 31 patients with type 1 DM and sex- and age-matched 33 healthy children were evaluated with conventional and tissue Doppler echocardiography. A correlation was examined between cardiac functions and HbA1C.
Results: According to conventional echocardiography, all parameters reflecting cardiac functions did not differ between patients with type 1 DM and healthy children. However, in tissue Doppler echocardiography, mitral valve early diastolic annular peak flow rate (E'), mitral valve systolic flow rate (S'), ratio of mitral valve early diastolic peak flow rate to mitral valve early diastolic annular peak flow rate (E/E'), and left ventricle myocardial performance index (MPI) were found higher and left ventricle ejection time (ET) was found shorter in patients with type 1 DM ($p < 0.05$). In addition, tricuspid valve E' and right ventricle MPI were found higher, right ventricle ET and tricuspid E/E' were found lower in patients with type 1 DM compared to healthy children ($p < 0.05$) by tissue Doppler echocardiography. It was shown that mitral E', tricuspid E', and right ventricle ET had a positive correlation with HbA1C level in patients with type 1 DM.
Conclusions: This study showed that although there was no difference between patients with type 1 DM and healthy children by conventional echocardiography, tissue Doppler echocardiography showed dysfunctions of both ventricles. This influence was closely related with the control degree of blood glucose level. These findings signify the diagnostic value of tissue Doppler echocardiography in the early detection of cardiac effects among patients with type 1 DM.
Keywords: cardiac function; children; tissue Doppler echocardiography; type 1 diabetes mellitus.

P-131
Left ventricular muscle mass in CHD by realtime three dimensional echocardiography: Quick, cheap, precise, reproducible, ready for clinical use

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Introduction: Calculation of muscle mass is one important aspect in the follow up of increased pressure load. Problems of current two dimensional echocardiographic techniques are a lack of precision and bad reproducibility. We tested the hypothesis if realtime three dimensional echocardiography (RT3DE) is far behind the gold standard cardiac magnetic resonance (CMR) concerning calculation of left ventricular muscle mass.
Methods: 20 individuals (5-44ys, 9 f, median 15ys, 10 patients with LV disease) were consecutively investigated by CMR (3T TX Achieva, Philips Healthcare, Cine steady-state free precession technique, TR/TE/flip = 2.7/1.35 ms/40°, slice thickness = 5-6 mm, 25 cardiac phases) and RT3DE (IE 33, X5-1 transducer Philips, 4 subvolumes, resolution 20-24 cardiac phases). Calculation of CMR data was done using the summation of disks method excluding papillary muscles (HDZ MR-Tools software package, HDZ, Bad Oeynhausen, Germany), RT3DE data were processed using 4D LV Analysis 3.1 software (Tomtec, Germany) by two separate expert investigators blinded to each other. For interobserver variability a third investigator was introduced. Statistical analysis by Bland Altmann, correlations by Pearson-Bravais.
Results: All data could be evaluated, muscle mass by CMR ranged from 38-204g, mean 110g. RT3DE provided very mild overestimation of $1,7 \pm 5,6\%$($r = 0,991$) with low intraobserver-($3,2 \pm 5,5\%$, $r = 0,993$) as well as interobserver-variability ($1,1 \pm 5,5\%$, $r = 0,994$). Mean calculation time for RT3DE was <3 minutes.

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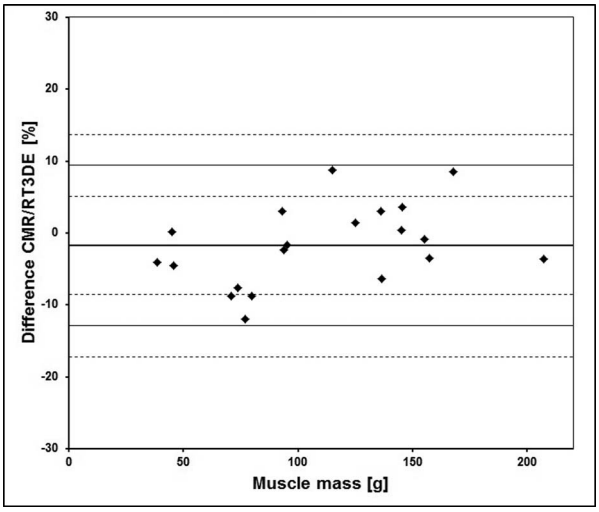
Conclusions: RT3DE allows quick and precise calculation of left ventricular muscle mass for a wide spectrum of LV size which makes the method an interesting tool for clinical use in patients.



P-132

Cardiopulmonary developmental characteristics in healthy children at high altitudes in China

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Objectives: Altitude-hypoxia induces pulmonary arterial hypertension and altered cardiac morphology and function, which is little known in healthy children at high altitude. We compared the cardiopulmonary measurements between the healthy children at 16 m and those at 3700 m in China, and further compared these measurements between the Han and Tibetans at 3700 m

Methods: Echocardiography was assessed in 477 children (15 day-14 years) including 220 at 16 m and 257 at 3700 m. Children were divided into 7 age groups (<1 m-6 m-1y-3-6y-10y-14y). The dimensions of the right and left were measured. Systolic and diastolic function included ejection fraction and fractional shortening of RV and LV and cardiac output (CI). Diastolic function included of E and A waves and E wave declaration time of tricuspid and mitral valves (EDTTV and EDTMV), isovolumic relaxation time of d RV and LV (RIRT and LIRT). Mean pulmonary arterial pressure (mPAP) was estimated.

Results: As compared to the 16 m group, children at 3700 m had higher mPAP, increasing dilatation of the right heart, and slower decrease in RV hypertrophy in 14 years ($p < 0.05$). The left heart morphology was not different ($p > 0.20$). Systolic and diastolic function of both ventricles significantly reduced, but CI was higher ($p < 0.0001$). There was no difference in any measurement between the Han and Tibetans ($p > 0.05$).

Conclusions: The cardiopulmonary developmental values in the Han and Tibetan children at high altitude provide references for the care of healthy children and the sick ones with cardiopulmonary diseases at high altitude.

P-133

Assessment of global & regional ventricular function in children with Friedreich's Ataxia: a single centre experience

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Introduction: Friedreich's ataxia (FA) is an autosomal recessive neurodegenerative disorder, caused by unstable GAA expansions in the FA gene encoding for the 210-amino-acid protein frataxin. Cardiac associations described are hypertrophic cardiomyopathy (HCM), dilated cardiomyopathy (DCM), ventricular dysfunction and arrhythmias. There is dearth of literature in paediatrics of cardiac presentation and progression in FA.

Methods: 13 year retrospective analysis of data in a single centre; clinical features, electrocardiographs (ECG) and echocardiograms of all patients with FA looking for patterns of presentation and progression of cardiac disease.

Results: 21 patients with confirmed diagnosis of FA were seen (10 male and 11 female). Analysis was available in 18 patients (insufficient data in 2 patients) for assessment of global ventricular function, and advanced (synchronicity & strain) functional assessment in 10 of these. Median age of presentation to cardiology was 11 years (5-15 years).

ECG abnormalities in 12 (63%) with isolated T wave inversion in lateral leads in 8(42%), left ventricular hypertrophy with strain pattern in 4, normal in 6 (no data in 1). None had palpitations/syncope.

Echocardiograms assessed for left ventricular mass (107–209G), global and regional function. 15 (83%) had concentric HCM; of these 13 had normal global systolic function, three had diastolic dysfunction. One had DCM, and one (5%) had hypertrophic obstructive cardiomyopathy (HOCM), and no hypertrophy in two. Synchronicity parameters were within normal limits in all 10 patients we analysed; all segment delay varied from 45-165 ms (SD: 6-45 ms). Longitudinal strain (ε) showed a reduction in all patients (average ε range was -9.1% to -20.6%) with segmental variation that was not consistent to a particular region.

Nine patients were on idebenone; three were on additional Vitamin E and Coenzyme q10; and one on beta-blocker (HOCM). There were no arrhythmias noted in our case series.

Conclusion: Children with FA have some evidence of cardiac dysfunction, but are asymptomatic. The dominant pattern is concentric LVH, but other patterns are also present, and there are regional changes even when global function is normal. Regular, serial assessment is essential in this group preferably with advanced analysis looking for progression.

P-134

How does HIV affect cardiac function in children admitted for management of severe acute malnutrition in rural Kenya?

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Background: Approximately 3.4 million children are infected with HIV worldwide, 90% in Sub-Saharan Africa. They suffer from severe acute malnutrition (SAM) and mortality amongst this group remains high. We wish to determine the cardiac risk factors for such children in order to target the treatment at those most likely to succumb.

Methods: As part of the CArdiac Physiology in MALnutrition (CAPMAL) Study we recruited children with SAM (marasmus

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or kwashiorkor) who presented to a rural Kenyan hospital. We used a Vivid I portable echocardiograph to assess cardiac function at day 0, day 7 and day 28 from admission. Bedside observations, co-morbidities and blood tests including HIV status were also recorded.
Results: 52 marasmic and 36 kwashiorkor cases were recruited. 20 (22.7%) were HIV positive (HIV+), more with marasmus (32.7% vs 8.3%; p = 0.007). Between HIV+ and HIV-, age (23.5 vs 18.5 months), gender (60 vs 53% male), mid upper arm circumference (10.5 vs 11.0 cm) and co-morbidities were similar but a higher proportion presented with chest infections (30 vs 7.4%, odds ratio (OR) 5.4; range 1.4-20.2) in the HIV+ group. HIV+ children had higher heart rates at day 28 (143 vs 131/min, $p < 0.05$). HIV+ and HIV- children showed normal average fractional shortening, mitral annular plane systolic excursion (MAPSE), E/A ratio and E'/E ratio. Right ventricular systolic long axis function was reduced (TAPSE 12.8 vs.14.9 mm; p = 0.0123) and Tei Index was higher in HIV+ at day 7 (0.4 vs 0.29, $p < 0.001$). We found no difference in indexed LV mass (g) at any time point (Day 0: 64.8 vs 63.5 g day 7: 78.8 vs 69.8 g day 7, day 28: 80 vs 77 g, all $p > 0.05$). In all groups, the cardiac index increased proportionately with the increase in left ventricular dimension in diastole. However, children were more likely to die if they were HIV+ (OR 3.2; CI 0.96-10.76).
Conclusions: Admission with SAM appears to be linked to the presence of HIV, especially in those with marasmus. Despite an improvement in cardiac index and no difference in myocardial performance or structure, malnourished children are more likely to die if HIV positive.

P-135
Evaluation of cardiac functions in children with liver cirrhosis using Brain natriuretic peptide and tissue Doppler imaging

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Introduction: Patients with liver cirrhosis suffer various degrees of cardiac dysfunction. Brain type natriuretic peptide (BNP) is a cardiac neurohormone released in response to increased ventricular wall tension. Cardiac dysfunction in cirrhotic children has been rarely investigated.
The aim of the study was to evaluate the level of BNP and its relationship with cardiac functions in children with cirrhosis.
Patients and Methods: Prospective longitudinal study of 52 patients with hepatic cirrhosis and 53 age and sex matched healthy children as controls. Patients' ages ranged from 9 months up to 15 years. Cardiac function was assessed using BNP, conventional echocardiography and Tissue Doppler Imaging (TDI) for systolic and diastolic functions. Results were analyzed by using the Guidelines of American Society of Echocardiography. BNP plasma level was measured using quantitative ELISA technique for BNP supplied by WKEA MED SUPPLIES CORP.
Results: The BNP levels were significantly higher in cases compared to controls (5.25 ng/l versus 4 ng/l, $p < 0.04$) but without significant difference compensated & decompensated patients. Compared with controls, the patients had larger left atrium and right ventricle diameters (P value 0.01, 0.02 respectively) and increased posterior wall thickness (P value 0.04). The patients had higher late atrial diastolic filling velocity (A wave) of tricuspid valve (TV) inflow (P value 0.001) and lower ratios between the early diastolic filling velocity (E wave) and E/A ratio of both mitral & tricuspid valves inflow (P value of 0.005 & 0.0008 respectively) The patients had significantly longer Isovolumetric Ventricular Relaxation Time (IVRT) of the left ventricle (P value 0.008) and higher early diastolic peak myocardial velocity (E′) of the right ventricle (P value of 0.0003). The findings are compatible with diastolic dysfunction. The high level of the BNP was only correlated with the E wave velocity of the TV inflow (P value 0.004). BNP level showed no significant correlation with any of the clinical or laboratory findings.
Conclusion: Cirrhotic patients have subtle cardiac dysfunction. BNP is a useful marker of cardiac dysfunction yet it could not be correlated with specific clinical or laboratory findings and still further studies are required to correlate it with echocardiographic findings.

P-136
Abnormal longitudinal cardiac rotation is a new marker of regional LV myocardial dysfunction in children and young adults with end-stage renal failure undergoing hemodialysis

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Background: Cardiac dysfunction frequently complicates the clinical course of patients with end-stage renal failure (ESRF). Recently, we observed abnormal longitudinal cardiac rotation (LR) among patients with ESRF. In this study we sought to quantify LR mechanics in patients undergoing hemodialysis (HD).
Methods: Twenty-four subjects, 12 patients (58% male; age 17.5 ± 4.4 years) with ESRF undergoing HD, and 12 aged-matched controls, were prospectively studied. All patients underwent echocardiographic studies before and 1 hour after HD. LR mechanics were quantified with two-dimensional speckle tracking echocardiography using a 5-segment-rotation-model (2 lateral and 2 septal segments, 1 apical segment). We also assessed longitudinal strain, displacement and mechanical dyssynchrony.
Results: A typical pattern of LR was noted in all controls. This physiological LR pattern was characterized by a predominant early-systolic counterclockwise rotation (CCWR) of the lateral segments, and a late-systolic clockwise rotation (CWR) of the septal segments. A small end-systolic apical rotation was noted in all controls. LR abnormalities were identified in 5/12 (41.6%) of ESRF patients, and included an apical counterclockwise motion pattern (n = 2), or a delayed septal clockwise rotation pattern (n = 3).
Time to peak CCWR increased significantly after HD (preHD: median 35.6% (range 29.2–39.0%) vs. postHD: 41.0% (32.7–49.3%), p = 0.008). Moreover, CCWR of the lateral segments significantly increased after HD (p = 0.019). Timing of late systolic CWR occurred significantly earlier in systole in patients than in controls (preHD: 72.0% (60.0–84.3%) vs. controls 87.6% (82.1–91.8%), p = 0.033), and showed a significant prolongation after HD (p = 0.003).
Longitudinal strain was significantly reduced in patients, both before and after HD. Similarly, patients showed more dyssynchrony (p = 0.009). In contrast, septal displacement was significantly reduced in patients after HD (p = 0.003). A negative correlation was observed between regional lateral strain and septal displacement (p = 0.01). Moreover, septal displacement correlated significantly with timing of CCWR and CWR.

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Conclusions: LR abnormalities were demonstrated in a large proportion of patients with ESRF. Abnormal LR motion seems a feasible and reliable new marker of regional LV myocardial dysfunction in ESRF patients.

P-137

Abnormal aortic arch morphology in Turner syndrome patients is a risk factor for hypertension

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Hypertension in Turner syndrome (TS) is a multifactorial, highly prevalent and significant problem that warrants timely diagnosis and rigorous treatment.
We aimed to investigate the association between abnormal aortic arch morphology and hypertension in adult TS patients.
Methods: single centre retrospective study in 74 adult TS patients (age 29.41 ± 8.91 y) who underwent a routine cardiac MRI. Patients were assigned to the hypertensive group (N = 31) if blood pressure exceeded 140/90 mmHg and/or if they were treated with antihypertensive medication. Aortic arch morphology was evaluated on MRI images and initially assigned as normal (N = 54) or abnormal (N = 20), based on the curve of the transverse arch and the distance the left common carotid – left subclavian artery. We propose a new more objective method to describe aortic arch abnormality in TS by determination of the relative position of the highest point of the transverse arch (AoHP).
Results: logistic regression analysis showed that hypertension is statistically and independently associated with age, BMI and abnormal arch morphology, with the size effect of the new AoHP method being larger than the classical method. TS patients with hypertension and abnormal arch morphology more often had dilatation of the ascending aorta.
Conclusion: there is a strong significant association between abnormal arch morphology and hypertension in TS patients, independent of age and BMI, and not related to other structural heart disease. We suggest that aortic arch morphology should be included in the risk stratification for hypertension in TS and propose a new quantitative method to express aortic arch morphology.

P-138

Mitral annular peak systolic velocity (Sm) in healthy children, calculation of reference values, of z-score values, and comparison to the mitral annular plane systolic excursion

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Background: The mitral annular peak systolic velocity (Sm) is an echocardiographic measurement using tissue Doppler imaging to assess longitudinal left ventricular (LV) systolic function in children and adults. We determined growth related changes of Sm to establish reference values for the entire pediatric age group.
Methods and Results: A prospective study was conducted in a group of 690 healthy pediatric patients (age: 1 day to 18 years). We determined the effects of age, sex and body surface area (BSA) on the Sm values. Regression analysis was used to estimate Sm from age, BSA, and sex. Additionally a correlation of normal Sm with normal age-matched values of the M-mode parameter mitral annular plane systolic excursion (MAPSE) was measured. The Sm ranged from a mean of 5.8 cm/s (Z-score ± 2: 3.6–8.0 cm/s) in the newborn to 11.8 cm/s (Z-score ± 2: 8.5–15.1 cm/s) in the 18 year old adolescent. The Sm values showed a positive correlation with age and BSA with a non-linear course. There was no significant difference in Sm values between females or males. A significant correlation was found between Sm and MAPSE values.
Conclusion: Z-scores of Sm values were calculated and percentile charts were established to serve as reference data in patients with congenital heart diseases.

P-139

The value of echocardiographic exam for the diagnosis of cardiac involvement in non-Hodgkin lymphoma in child

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Background: Clinical signs and symptoms of cardiac involvement in non-Hodgkin lymphoma (NHL), is frequently undetected and diagnosis could be missed unless a routine echocardiographical investigation might be performed.
Objective: To reveal the value of echocardiographic investigation for diagnose and follow up the cardiac involvement in NHL in child, other than that determined by the specific treatment.
Methods: Patients: 38 children, 21 males and 17 females, aged between 3month and 17 years with NHL, hospitalised in in a 8 years period. In all cases were performed: clinical exam, ECG, chest X-ray and Doppler echocardiography(echo).
Results: Cardiac involvement was proved in 6 cases (15, 8%) of T-cell (5) and B-cell (1 case) NHL. Clinical signs on onset like astenia, dyspnea, cough, superior vena cava syndrome have been assigned to the base disease. ECG revealed: low voltage of QRS complexes and T waves. Chest X-ray: mediastinal involvement (5 cases), massive right-side pleural effusion (2 cases), cardiomegaly (5 cases). Ecocardiographic aspects of cardiac involvement were: pericardial effusion (4 cases) to cardiac tamponade (2cases); pericardial tumor (1 case). In 2 cases was revealed in first echo exam intracardiac masses, without the possibility of specify the initial starting point for the lymphoma: case 1 with a tumoral mass filling the right atrium with implantation base towards superior vena cava, case 2 – tumoral mass extending from the free wall of the right atrium to the septal tricuspid valve leaflet, associated with massive pericardial effusion. Cardiac modifications found in echocardiographycal exam in case 1 were confirmed later in the autopsy. Excluding patients who died, in all cases pericardial effusions and intracardiac masses decreased or disappeared by specific cytostatic treatment.
Conclusion: Because of high incidence of cardiac involvement in non Hodgkin malignant lymphoma in child and often difficulties of diagnosis in the early stages of evolution, echocardiographic exam is necessary in all cases at the first exam of patients, even if cardiac involvement signs are missing, in order to prevent the evolution to cardiac tamponade or other cardiac emergency. Echocardiography is the most important investigation to diagnosis and follow-up the cardiac involvement in NHL in child.

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P-140

The value of echocardiography for the diagnosis of anthracyclines induced cardiotoxicity in children with malignant diseases

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Objective: Assessment of the most important parameters of systolic and diastolic function of left ventricle (LV) and establishing who are most predictive for early dysfunction in children with anthracyclines treatment for malignancies.
Methods: Patients: 124 patients (aged 3 month -19 years) treated with anthracyclines for hematological malignant diseases. Mean cumulative dose of anthracyclines was 320 mg/m2 (240-820 mg/m2). Doppler echocardiography (echo) had evaluated the most important LV systolic and diastolic function parameters. The interval between examinations have been established depending on the cumulative anthracyclines doses (200,300, 400 and >500 mg/m2). The data were correlated with the dispersion of QT/QTc intervals in 40 patients. Control group: 40 healthy children without cardiovascular suffering.
Results: Echo exam revealed suggestive modifications for cardiotoxicity in 63 cases (51%) with/without clinical manifestations of cardiac suffering up to heart failure. The most important echo modifications have been found at the patients with a cumulative dose over 350 mg/m2: higher incidence (48%) and precocious onset of the diastolic dysfunction of the LV type relaxation impairment of LV (17%)or compliance impairment of LV (31%); systolic dysfunction of LV: decrease of ejection fraction (19%); increase of the deceleration time of E wave-DTE (35%) and of the isovolumic relaxation time- IVRT (28%). The Tei index: increased in 30 cases: 21 cases values beetwen 0.39-0.62, 9 cases >0.62. ECG:an increase in the dispersion of interval QT/QTc in 73% of patients correlated with the cumulative doses of the anthracyclines >400 mg/m2.
Conclusions: Diastolic dysfunction of LV occurs earlier and with a superior incidence (48%) versus systolic dysfunction (19%) of LV, proved by decline of the ejection fraction of LV. Diastolic echo parameters (E, E/A, DT, IVRT, IVCT) and Tei index, allows a precocious detection of antharcyclines-induced cardiotoxicity that are frequently missed or not suspected clinically, especially when the LVEF is normal. The echocardiographic modifications and increased of QT/QTc intervals, well correlated with cumulative doses of the anthracyclines, can put in evidence the signs of cardiac suffering in the stage infraclinic and it is necessary to be follow up booth during and after the cytostatic treatment

P-141

Evaluation of Left Atrial Function Using Two-Dimensional Speckle Tracking Echocardiography in Children with Type 1 Diabetes Mellitus

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Introduction: Left atrial (LA) deformation analysis by two-dimensional speckle tracking echocardiography (2D-STE) has recently been proposed as an alternative approach conventional echocardiography. Aim of this study is the evaluation of left atrial function with Speckle-tracking echocardiography in pediatric patients with Type 1 Diabetes Mellitus who did not yet develop micro and macro vascular complications, at least 3 years at last 10 years followed up.
Method: A total of 63 patients without diabetic complications (mean age: 13,81 years) and 36 healthy (mean age: 12,42 years) children were included in this study. Conventional transthoracic echocardiography and left atrial speckle tracking echocardiography were performed in all patients. Images of the LA were acquired from apical two- and four-chamber views. The LA strain (%) (LAS) parameters systolic [LAS-S], early diastolic [LAS-E], late diastolic [LAS-A] during atrial contraction were assessed. The LA volumes (LAV) were calculated using the biplane area-length method. The LA volume indices (LAVI) were calculated by dividing the LA volumes by the body surface area.
Results: Results of diabetic group and healthy children were compared. In diabetic group, LAS–S (39,91 ± 9,08/ 45,88 ± 8,85 $p < 0,003$) LAS-A (13,43 ± 4,68/15,82 ± 5,02) $p < 0,003$) and LAS-E 26,86 ± 7,54/29,77 ± 7,65) $p < 0,003$) were found decreased and LAVI (ml/ m2) (26,10 ± 2,81/23,53 ± 2,04 $p < 0,003$) was found increased. Additionally, Mitral valve A wave velocity (0,58 ± 0,14/0,50 ± 0,10 $p < 0,003$) increased, E/A wave ratio (1,76 ± 0,37/ 2,11 ± 0,34 $p < 0,003$) decreased. There were no statistical significant difference in systolic function with conventional echocardiographic examinations.
Conclusions: We found deterioration in atrial functions with speckle tracking echocardiography methods in Type 1 DM patients without diabetic complications. Early detection of effects on target organs may prevent the irreversible damage and taking preventive measures are important in terms of improving the quality of life.

P-142

Evaluation of cardiac functions in preschool children with bronchopulmonary dysplasia by using 2D strain–strain rate echocardiography

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Objective: The objective of this study was to determine the possibility of subclinic or asymptomatic myocardial dysfunction with strain echocardiography in the long-term follow-ups of the children with BPD.
Material and methods: The study population consisted of 32 healthy and 77 prematurely born children. Eighteen of the premature children included in the study had mild BPD, 17 had medium-severe BPD, 42 did not have BPD. The range of age was 3-6 years. Age and sex matched healthy children who were born at term and have no respiratory diseases are defined as the control group. Cardiac functions of all children were evaluated with conventional and strain echocardiographic methods.
Results: There were no differences between groups with conventional methods. The peak systolic strain values in the left ventricular (LV) lateral/mid-anterior obtained from apical 4-chamber view were found significantly lower in the group with BPD according to the control group ($p < 0.001$). The LV lateral apical strain values in the same view were significantly lower in the medium-severe BPD group ($p = 0.01$). However, the strain rate in the LV lateral-mid anterior segment was significantly lower obtained from apical 4-chamber view in the premature children independently from BPD. There was no significant difference between the groups in any segment of the right ventricle. The LV apical 4-chamber global longitudinal strain value was significantly lower in the medium-severe BPD group compared with the control group.

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Conclusion: In our study, Although it cannot be detected any alteration in cardiac function with conventional methods, strain echocardiography revealed that cardiac functions affected in the children who were born with very low birth weights with BPD. Myocardial 2D strain echocardiography is important in the assessment of cardiac function of children with BPD.

P-143

Assessment of left ventricular functions with strain and strain rate echocardiography in children after acute period of Kawasaki disease

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Introduction: Kawasaki disease is a vasculitis of childhood that is characterized by fever, cervical lymphadenopathy, rash, erythema of the lips and oral mucosa, changes in extremities and bilateral nonexudative conjunctivitis. Coronary aneurysms or ectasia develop in 15% to 25% of untreated children. Patients with coronary artery abnormalities may lead to cardiovascular morbidity and mortality after the disease. The aim of this study is to assess myocardial deformation leading to subclinical or asymptomatic myocardial dysfunction, by 2D strain echocardiography after acute phase of Kawasaki disease.
Methods: Our study included 30 patients after acute phase of Kawasaki disease and 30 age and sex matched healthy controls. In addition to conventional echocardiographic methods, tissue Doppler, 2D strain and strain rate imagings were performed to assess left ventricular functions. Statistical analysis was evaluated by using the T-Student's test, Chi-square test and and Pearson correlation.
Results: The mean follow-up time was 3.55 ± 2.20 (1-8.15) years. The mean age was 5.86 ± 2.46 years in patients with Kawasaki disease and 6.11 ± 2.57 years in the control group. There were no significant differences between two groups according to conventional echocardiography, tissue Doppler imaging and 2D strain-strain rate echocardiographic methods. The follow-up time had a positive correlation between global four, three chamber and global longitidunal left ventricular strain values. We found a significant positive correlation between global four chamber longitidunal strain values and erythrocyte sedimentation rate, but a negative correlation between global four chamber longitidunal strain values and hemoglobin values. Also, ejection fraction had a significant positive correlation between serum albumin values but had a negative correlation between erythrocyte sedimentation rate.
Conclusion: Strain values showed variability with the follow-up times which indicate that Kawasaki disease might cause left ventricular dysfunction after acute period of disease, either with coronary artery abnormalities or not. 2D strain echocardiography is a useful tool to define later phase cardiac evaluation of Kawasaki disease, especially in which that conventional methods can not present a more detailed analysis on global and regional myocardial function.

P-144

Abnormal Biophysical Properties of the Aorta in Post Surgical Patients with Congenital heart diseases: a Non-Invasive Study

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Introduction: Many congenital heart diseases (CHD) are associated with congenital and/or acquired structural alterations of the aortic wall, but little data are available on the functional status of the aorta. Using non invasive techniques, we sought to assess the aortic biophysical properties, total arterial compliance, hydraulic power and efficiency in post operative children with: [1] tetralogy of Fallot (TET); [2] coarctation of the aorta (COA) and [3] transposition of great arteries (TGA).
Methods: 55 children with CHD (TET = 24, 15.2y, COA = 20, 13.4y, and TGA = 11, 14.3y) were compared with 55 matched controls (CTRL, 14.1y). M-mode, and Doppler echocardiographic imaging, and carotid artery applanation tonometry were used to measure aortic flows and dimensions. Pulse-wave velocity (PWV), input (Zi) and characteristic (Zc) impedance, arterial stiffness (Ep) and β-index were calculated. Total arterial compliance (TAC), mean (Wm) and total (Wt) hydraulic power, and efficiency (HE) were calculated from carotid pulse tracings and flows using standard fluid dynamics equations. A Mann-Whitney U test was used to determine differences between groups. A p-value of 0.05 was considered statistically significant.
Results: All CHD subgroups had higher PWV (TET = 492, COA = 458, TGA = 527 vs CTRL = 360 cm/s) and TAC (TET = 1.71, COA = 2.32, TGA = 2.34 vs CTRL = 1.35 mL/torr/m^2) compared to CTRL. COA and TET had higher Zc (COA = 234, TET = 177 vs CTRL = 138 dyne sec/cm^5). Ep, β-index and Zi were similar. Wm and Wt were higher in TGA compared to TET and CTRL (TGA = 1010 vs TET = 730and CTRL = 680 mW/m^2) and (TGA 1273 vs TET = 936 and CTRL = 830 mW/m^2), respectively. HE was higher in TET compared to COA and TGA (TET 86% vs COA 83% and TGA 82%).
Conclusion: Children with post-operative TET, COA and TGA have stiffer aortas, increased work and higher total arterial compliance than CTRL. It is unclear if this is related to intrinsic lesions of the aorta, alterations of the aorta due to surgical repair or other factors. Further studies and follow up are needed to determine if these abnormalities predispose these patients to long-term cardiac dysfunction and cardiovascular risk.

P-145

Safety of Cardiopulmonary Exercise Testing in Children with Pulmonary Hypertension

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Background: Cardiopulmonary exercise testing (CPET) is a valuable tool to objectively measure exercise capacity. Recent evidence supports its role as a prognostic tool and to guide treatment in several conditions including pulmonary hypertension (PH). Evidence for its use in children with PH is extremely limited, partly because of concerns regarding its safety in this setting. The purpose of this study was to assess the safety profile of CPET in a large cohort of paediatric PH patients.
Methods: Retrospective data was obtained from all consecutive patients undergoing CPET at a single centre between March 2004 and November 2013. Exclusion criteria for CPET were: height <120 cm, WHO class IV, history of syncope or significant ischemia/arrhythmias during exercise. Significant events recorded were: symptoms reported by patients, arrhythmias, abnormalities detected on ECG and abnormal responses of arterial O2 saturation (SaO2).
Results: 113 patients (53 with PH associated with congenital heart disease and 36 with idiopathic PH) had 185 CPETs in the

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study period. The median patient age was 14 years (25th and 75th percentiles, 10.5 and 15 years). Peak oxygen uptake (VO2) (±SD) for all patients was 21.4 ± 8.0 mL/kg/min, which corresponded to 54.4 ± 20.0% of the predicted values. Peak respiratory quotient was 1.08 ± 0.16. All tests were maximal, except 2, requiring premature termination for clinical reasons. Baseline SaO2 was 93.8 ± 8.5% and 19% of children had a baseline SaO2 <90%. The average SaO2 during peak exercise was 82.7 ± 19.1%. A drop of SaO2 <5% was observed in 44.6% of patients, whereas 20.1% of patients had a decrease >20%. Seven patients (3.8%) experienced dizziness, associated with significant desaturation in 2, and requiring termination of CPET in 1. Five (2.7%) children experienced chest pain, which was associated with significant desaturation in 3 and early CPET termination in 1. No significant arrhythmias or ECG changes were observed.
Conclusion: Exercise testing in mild to moderately symptomatic children with PH in a controlled environment and with an experienced team is safe. Arterial O2 desaturation is common but asymptomatic in the majority of patients. No side effects of the test were serious and all resolved promptly when the test was terminated.

P-146
Microvascular Abnormalities in Coronary and Peripheral Circulation in Hypertrophic Cardiomyopathy of the Young
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Introduction: Sudden cardiac death (SCD) remains the most dreaded consequence of HCM, and may be the first presentation of the disease in the young. Initial assessment of young individuals with myocardial hypertrophy or asymptomatic individuals with HCM heredity is thus essential for further diagnostic and the therapeutic decision making.
Objective: To investigate peripheral vasomotor function and myocardial perfusion in young individuals with HCM and in those at risk for developing this disease.
Methods: Based on heredity or genetic predisposition, adolescents and young adults (median 20 years, range 12-30 years) with familial HCM (HCM; n = 10, mean IVS 19.6 mm, PW 11.5 mm), and individuals at risk for HCM (HCM-risk; n = 14, mean IVS 10.8 mm, PW 9.3 mm), were compared with healthy matched controls (n = 12, mean IVS 10.3 mm, PW 9.6 mm). All underwent assessment with echocardiography, cardiovascular magnetic resonance (CMR), and skin Laser Doppler with iontophoresis of acetylcholine (Ach) and sodium nitroprusside (SNP). CMR was performed at rest and during hyperemia with adenosine 140 mcg/kg/min. Myocardial perfusion (MP) was calculated as the ratio of coronary sinus flow and left ventricular mass (LVM) from CMR.
Results: Both echo and CMR demonstrated significantly greater left ventricular thickness and mass in HCM patients ($p < 0.05$) than in controls and HCM-risk. Compared to controls, cutaneous microvascular responses to Ach were enhanced in HCM and HCM-risk ($p < 0.05$). MP (ml/min/g) at rest was similar in controls, HCM-risk and HCM patients (0.8 ± 0.1, 1.0 ± 0.1, 0.9 ± 0.1; p = ns). During adenosine-induced hyperemia, HCM patients showed lower MP (2.5 ± 0.4, $p < 0.05$) than controls (3.9 ± 0.3) and HCM-risk (5.0 ± 0.5).
Conclusion: These results indicate that microvascular disease may be an early manifestation of HCM in the young.

P-147
Monocusp function in patients with Tetralogy of Fallot: comparison of echocardiography and cardiac magnetic resonance
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Background: In patients with Tetralogy of Fallot, one of the surgical methods that relieves right ventricular outflow tract obstruction is the transanular patch with insertion of a monocusp to prevent pulmonary regurgitation. Monitoring of monocusp regurgitation after surgery by echocardiography is a clinical practice. Its evaluation is however challenging, since the monocusp is placed in the right ventricular outflow tract and methods for evaluation of its function are neither described nor tested. We sought to test a method used for assessing regurgitation of the monocusp valve by echocardiography and compare with regurgitation measured by cardiac magnetic resonance (CMR).
Methods: Tetralogy of Fallot patients with inserted monocusp (n = 15, 3,5 ± 2 years) had an echocardiogram and CMR within one week of each other. On echocardiogram, the monocusp regurgitation was measured using parasternal and subcostal short axis views with a color Doppler. The regurgitation was graded on a scale of zero to four (0 = none, 1 = mild, 2 = mild to moderate, 3 = moderate to severe, 4 = severe), depending on the width of the regurgitation jet in the outflow tract in diastole. On cardiac magnetic resonance, monocusp regurgitation fraction (RF) was graded as mild (RF < 20%), moderate (RF = 20–40%), and severe (RF > 40%). Inter-observer variability of the echocardiographic data was calculated.
Results: On CMR, mean RF was 40 ± 11%. Echocardiography had good sensitivity identifying cases with RF > 40% (sensitivity 80%; 95% CI: 38–96%) but overestimated the amount of monocusp regurgitation when RF < 40% (false-positive rate 36%; 95% CI: 18–57%). The width of the regurgitation jet in the outflow tract on echocardiogram showed moderate correlation with RF on CMR ($R = 0.75$; $P < 0.01$). Interobserver variability of the echocardiographic measurements was moderate. 23% of observations differed in 1 grade, 9% observations differed in 2 grades.
Conclusions: This study suggests that width of the regurgitation jet in the outflow tract may make a modest contribution to the assessment of monocusp regurgitation in patients with repaired tetralogy of Fallot as compared with CMR.

P-148
Eosinophilic myocarditis in a 15 year-old: relationship between different modalities in assessment of the disease during the progress and resolution
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Background: Eosinophilic myocarditis is rare. We present a 15 year old boy, in whom, despite normalization of peripheral blood markers of myocarditis, there was an extensive inflammation on biopsy and MRI. Assessment of inflammation with ECHO-strain and laser Doppler of tissue endothelium were performed.

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Methods: Routine clinical, laboratory, ECHO, MRI and biopsy assessment were performed with findings of acute myocarditis. Inflammation assessment with laser doppler of the forearm and ECHO strain in the acute phase and after 6 weeks was done after informed consent and ethical approval.
Results: On initial presentation he had chest pain, raised levels of Troponin T, minimal ECG changes and normal routine echocardiography. Minimal ECG changes resolved within one week but chest pain recurred and Troponin T levels increased. MRI thorax revealed regional inflammation and a heart biopsy revealed eosinophilic infiltration and no viral agents were detected by nested PCR. No eosinophilia was found in peripheral blood. Laser Doppler imaging revealed signs of endothelial inflammation. Speckle tracking of the left ventricle revealed regional dyssynchrony coinciding with the same regions of inflammation and later scarring found on MRI.
A course of prednisolone was started at 1/2 mg/kg/day. CKMB levels declined to normal within three weeks but the Troponin T levels remained slightly elevated. A second MRI thorax after 8 weeks revealed progress of inflammation in extent and area and the dosage was increased to 1 mg/kg/day. Troponin levels decreased to normal. Biopsy after 3 month of therapy showed active inflammation but no eosinophilic infiltration. MRI revealed an unchanged area of inflammation and some scarring. The dosage of Prednisolone was reduced to 1/2 mg/kg/day and Azathioprine 1 mg/kg/day was added. MRI 6 months after presentation showed regress of inflammation and further scarring. Speckle tracking of the left ventricle revealed regional dyssynchrony coinciding with MRI. Laser Doppler imaging continued to show signs of mild inflammation.
Conclusion: In this case of eosinophilic myocarditis routine echocardiogram findings remained normal and ECG changes minimal. Laser Doppler and speckle tracking correlated better with MRI in revealing myocardial inflammation and damage.

P-149
Strain and Strain Echocardiography in Children with Wilson's disease

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Objective: The study aimed to evaluate strain and strain rate echocardiography in children with Wilson disease to detect early cardiac dysfunction.
Methods: In this study, consisted of 21 patients with Wilson's disease and a control group consisted of 20 ages and gender matched healthy children. All patients and control group was evaluated with 2D and colour coded conventional transthoracic echocardiography by using same echocardiography machine (Vivid E9, GE Healthcare), longitudinal, transverse radial strain and strain rate were assessed according to the recommended by American Society of Echocardiography. 2D strain and strain rate measurements were performed by using ECHOPAC software package.
Results: Global strain and strain rate: Wilson group have statistically significant lower peak 'A' longitudinal velocity of the left basal point and peak 'E' longitudinal velocity of the left basal point, and higher global peak 'A' longitudinal/circumferential strain rate according to the control group ($p < 0.05$).
Radial strain and strain rate
End systolic rotation was measured statistically lower values in Wilson group ($p < 0.05$). Segmental analysis showed that rotational strain measurement of anterior segment of Wilson group was measured statistically significant lower values in Wilson group and lateral segment of Wilson group.
Longitudinal strain and strain rate
Four chambers: End-systolic longitudinal strain and positive peak transverse strain was statistically low in Wilson group ($p < 0.05$). Segmental analysis showed statistically significance low values of end systolic longitudinal strain of basal lateral ($p < 0.05$) and statistically significance low values of end-systolic longitudinal strain of basal-septal segment ($p < 0.05$) in Wilson group.
Two chambers: End-systolic longitudinal strain and positive peak transverse strain was statistically low in Wilson group and segmental analysis showed statistically significance low values of end-systolic longitudinal strain of midanterior and basal-anterior segment ($p < 0.05$) in Wilson group.
Long axis: Segmental analysis showed that end systolic longitudinal strain measurements of basal posterior and mid-posterior segments statistically lower values in Wilson group ($p < 0.05$). End-systolic longitudinal displacement of basal posterior, mid-posterior, mid-antero-septal segments statistically lower values in Wilson group ($p < 0.05$).
Conclusion: In our study showed that, despite normal systolic functions, patient with Wilson disease shows diastolic dysfunction and regional deformation abnormalities especially rotational strain and strain rate abnormalities.

P-150
Testing two methods of quantitative flow measurement in the ascending aorta of patients with bicuspid and tricuspid aortic valves

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Introduction: Using 4D flow analysis was previously discussed, showing a great potential in visualizing and quantifying flow in cardiovascular diseases. In congenital heart diseases (CHD), 4D showed complex flow patterns; such as helical flow, in cases of aortic valve lesions. However intra-validation of flow measurements by 4D in CHD with complex flow pattern is lacking. The aim of this study is to test and validate 4D flow measurements using 2D phase contrast velocity encoding (PC-VENC) flow in the aorta in patients with bicuspid aortic valve with helical flow pattern.
Methods: Thirty-four participants underwent the 2D and 4D flow analysis in the ascending aorta. Eighteen of which were healthy volunteers (HV) and 16 were patients with bicuspid aortic valve disease (BAV). Velocity encoding was adjusted at 200 cm per second prospectively in both methods and in all patients. 2D Through Plane was planned perpendicular to the aortic segment from the localizer images. Field of view in the 4D was designed to include the aorta. Assessments of flow measurements from the 2D were done using the Argus flow tool by Siemens while the 4D flow measurements were done using the Fraunhofer Mevis flow tool. Region of Interest was determined at the level of the right pulmonary artery. Data from both methods were analyzed and compared. An interobserver agreement of the 4D data was

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performed using two blinded observers with the aforementioned 4D software to assess its reliability.
Results: The study showed good agreement between the 2D and 4D flow analysis in patients and normal subjects, as well as good agreements of the 4D interobserver assessment. Please refer to the Bland-Altman figures.
Conclusions: 4D flow analysis is a comprehensive method to assess flow measurements even in complex flow patterns in patients with CHD.

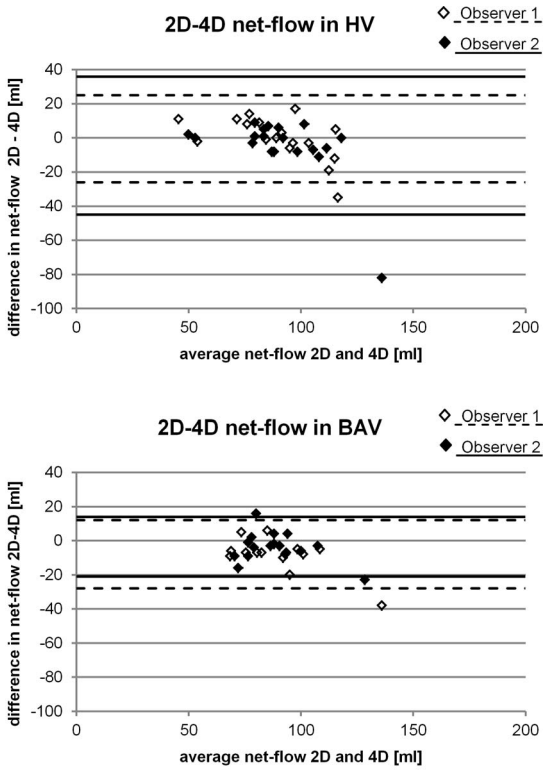

Figure 1 Bland-Altman-plot: Agreement of 2D- and 4D- net-flow-volumes in the ascending aorta of 34 participants (BAV n=16, HV n=18).

P-151
Three- Dimensional Echocardiographic Evaluation of Left Ventricular Systolic Dyssynchrony in Healthy Children

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Introduction: Three- dimensional echocardiography (3DE) is a most promising technology enabling pediatric cardiologists to evaluate left ventricular (LV) systolic function in children. One important determining factor of LV systolic function is the degree of dyssynchronous myocardial contraction among the segments of the left ventricle referred to as left ventricular systolic dyssynchrony (LVSD). Validated reference values of LVSD parameters are needed. Previous pediatric studies did not provide reference values relevant for children aged <5 years due to small sample sizes, underrepresentation of children aged <5 years or a distribution varying from normal.
Methods: We used 3DE and QLAB for defining reference values for the systolic dyssynchrony index (SDI) and other LVSD parameters in healthy children aged 0 to 14 years. 35% (n = 33) of the children included in our study were aged <5 years. The three- dimensional images were provided by one sonographer and were analyzed by one reader.
Results: LVSD parameters of 93 children (53 male, 40 female, mean age 6.8 +/- 4.1 years) were included in the calculation of reference values. The mean SDI was 2.00 +/- 0.97. It showed a normal distribution and was found to correlate with age.
Conclusions: Including 33 children aged <5 years in the study revealed an age dependency of the SDI. We were able to define an upper limit of 3.94% (Z+2) for the SDI. Further studies should define age specific upper limits for the SDI, a meaningful parameter of LVSD.

P-152
Cardiovascular Magnetic Resonance is Feasible in Many Patients Aged 3 to 8 Years Without General Anesthesia or Sedation

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Introduction: Cardiovascular Magnetic Resonance (CMR) of pediatric patients has become routine clinical practice. Patients under eight years are usually examined under general anesthesia (GA) or sedation without intubation. However, in our clinical experience, CMR may be feasible in patients aged 3 to 8 years without general anesthesia or sedation without intubation.
Methods: Retrospectively studied datasets of total number of 71 patients aged between 3 and 8 years. The total cohort was divided into two groups, first with no general anesthesia or sedation without intubation (no GA or sedation) and the second group patients with general anesthesia or sedation without intubation (GA or sedation). The patients' age groups and scan durations for each group, percentage of successfully answering the clinical question in each group, total number of scanned sequences in each group, and number of sequences per study were recorded and compared between both groups.
Results: Forty-four patients in the no GA or sedation group, 27 in the GA or sedation group. The scan duration was in the no GA or sedation group: 35 minutes ± 20.44 minutes, and in the GA or sedation group: 60 minutes ± 31 minutes ($p < 0.001$). The percentage of successful reports was 95% (42 of 44) in the no GA or sedation group and 89% (24 of 27) in the patients with GA or sedation group.
Conclusions: In scope of understanding the CHD hemodynamics, and clinical requests and questions; the decision of using neither general anesthesia with intubation nor sedation with without intubation was favored by our center's CMR unit over examining the patients under general anesthesia or sedation, to avoid the effect of general anesthesia and sedation on cardiac function during CMR, decrease the CMR scan duration, and increase the feasibility of the CMR in the young age group between three and eight years.

P-153
Intraindividual Validation of Ventricular Volume Measurement by Aortic and Pulmonary Arterial Flow Measurements in Routine Clinical Cardiovascular Magnetic Resonance of Congenital Heart Disease

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Introduction: Phase-contrast magnetic resonance imaging is an accurate quantitative tool for blood flow measurement in cardiovascular magnetic resonance (CMR). The aim of this study was to validate right and left ventricular stroke volume (RVSV & LVSV) measurement by forward flow stroke volume from aortic and pulmonary arterial measurements during CMR in routine clinical cases of congenital heart disease (CHD).
Methods: SV determined by ventricular volume assessment and arterial forward flow measurements during CMR of 147 consecutive routine patients (median age 22 years, range 0.5–64 years) with CHD, were retrospectively obtained from their clinical reports. Patients with ventricular septal defects, mitral valve regurgitation or severe tricuspid-valve-regurgitation were excluded. 126 LVSV were compared to the ascending aorta forward flow stroke volume (AoSV). 99 RVSV were compared to the main pulmonary forward flow stroke volume (MPASV). Ventricular SV was determined using a routine standard stack of cine axial slices. Arterial forward flow SV was determined using a routine standard phase-velocity quantitative flow sequence.
Results: AoSV correlated with LVSV by ($r^2 = 0.9$, $p < 0.0001$) and showed upper and lower limits of agreement in Bland Altman analysis of 11 ml and -12 ml, mean difference -1 ml. Similarly RVSV correlated with the accompanying MPASV ($r^2 = 0.8$, $p < 0.0001$) and showed upper and lower limits of agreement in Bland Altman analysis of 18 ml and -26 ml, and mean difference -4 ml.
Conclusion: Measured ventricular SV correlates closely with SV, assessed by CMR flow measurement in the originating great artery in routine clinical CMR of CHD. Validation of volume measurements in routine clinical CMR of CHD is very important, as this method can be used confidently in even complex and often distorted ventricular geometry in CHD.

P-154
Cardiac volumes can be quantified accurately during free-breathing in young patients with congenital heart disease by Cardiovascular Magnetic Resonance

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Introduction: Cardiovascular Magnetic Resonance (CMR) with respiratory commands is the gold standard technique to measure cardiac volumes. Although cardiac volumes can be measured during free breathing in patients with congenital heart disease (CHD), its accuracy is unknown. Therefore, the aim of this study was to compare cardiac volumes acquired during free breathing with volumes acquired during breath hold commands.
Methods: Cardiac volumes were measured in every patient using both free breathing and breath hold techniques, by a routine standard steady state free precession (SSFP) cine sequence in axial slice orientation. The sequence parameters were always identical besides the number of averages being three during free breathing and one during breath holds. Volumes were determined in a blinded fashion by endocardial contouring and then correlated using the coefficient of determination and compared by Bland Altman analysis. Eleven younger patients with CHD aged median 4 yrs, range 3 months-14 yrs, were examined under general anesthesia and intubation (intubated younger patient group). Twelve older patients with CHD aged median 20 yrs, range 11-61 yrs were examined consciously (conscious older patient group).
Results: The agreement of the end systolic volume (ESV) and end diastolic volumes (EDV) between scanning with and without respiratory commands in both ventricles was excellent in the intubated younger patients (LVEDV: $r^2 = 0.98$, LVESV: $r^2 = 0.99$, RVEDV: $r^2 = 0.99$, RVESV: $r^2 = 0.99$) and less acceptable in the consciously examined older patients (LVESV: $r^2 = 0.87$, LVEDV: $r^2 = 0.78$, RVESV: $r^2 = 0.84$ and RVEDV: $r^2 = 0.84$).
Conclusions: Cardiac volumes can be quantified very accurately during free-breathing in young patients with congenital heart disease by CMR using standard routine imaging techniques. If needed, even in older patients cardiac volumes can be quantified reasonably accurate during free-breathing. Therefore, free-breathing is an alternative technique for patients not able to hold their breath and thus obliterating the need of anesthesia for many patients undergoing CMR. In addition it will provide a chance for the CMR to assess the hemodynamics in a resting physiologic condition.

P-155
Physiscal exercise reduces aortic regurgitation - a study using a new magnetic resonance exercise test

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Background: Direct and quantitative assessment of aortic regurgitation (AR) during physical exercise has not been studied so far.
Methods and results: Twelve asymptomatic patients with isolated moderate aortic regurgitation (AR) and twelve healthy controls were studied by cardiovascular magnetic resonance (CMR) at rest and during submaximal physical exercise (25% of maximal exercise capacity in healthy controls) using a specially designed apparatus. AR was quantified as regurgitant fraction (in percent) of left ventricular stroke volume.
During submaximal exercise, heart rate, systolic and diastolic blood pressure, as well as cardiac index (CI) increased significantly and similarly in both groups.
In controls under submaximal exercise, median VO2 increased from 4.1 ml/kg/min (range: 3.7-4.9 ml/kg/min) to 8.6 ml/kg/min (range: 7.2-13.7 ml/kg/min, $p = 0.008$). Left ventricle (LV) volumes and ejection fraction (EF) did not change.
In patients, AR decreased from median 35% (range: 9-64%) of left ventricular stroke volume at rest to 16% (range: 7-42%) during submaximal exercise ($p = 0.003$). There was a linear correlation between AR at rest and increase in CI during submaximal exercise ($r^2 = 0.64$; $p = 0.001$); LV-EF increased ($p = 0.021$), LV end diastolic volume (LV-EDVI, $p = 0.004$) and LV end systolic volume (LV-ESVI, $p = 0.009$) decreased.
Conclusion: In asymptomatic patients with moderate isolated AR, regurgitant fraction decreases and LV function improves during submaximal exercise. This can be monitored precisely by submaximal exercise testing during CMR.

P-156
Lower activity value of antithorombin reflects comprehensive cardiac stress in infants before Glenn

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Background: Antithorombin (AT), which is produced in liver, is activated on vascular endothelial cells. Activity of AT (AT-activity)

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drops in serious conditions, such as hepatic disorder, myocardial infarction. However, there are no reports about relation between AT-activity and cardiac stress in congenital heart disease. High cardiac stress frequently subsists in infants before Glenn. We predicted lower AT-activity existed in pre-Glenn infants because of high cardiac stress. We attempted to identify clinical presentation in pre-Glenn infants with lower AT-activity.
Methods: Between 2004 and 2013, 56 pre-Glenn infants aged from 2 months to 6 months were studied. Cardiac catheterization was performed with Glenn in view. Venous blood samples for analysis of AT-activity were obtained before surgery. We defined lower AT-activity ($n = 21$) as AT-activity levels 80% or below, which is lower limit of normal in our hospital. Cardiac performances and clinical data which would influence lower AT-activity were determined.
Results: The ratio of patients with lower AT-activity in each first strategy classified by form of pulmonary artery (PA) was as below: 70% in bilateral-banded PA; 67% in ductus arteriosus-relaying PA; 35% in shunted PA; 22% in native-stenotic PA; 0% in trunk-banded PA. After multiple logistic regression analysis lower AT-activity was independently associated with odds ratio of 13.0 ($p = 0.022$) for early age ($\leqslant$0.30years), 9.9 ($p = 0.02$) for high levels of brain natriuretic peptide ($\geqslant$100 pg/dl), 8.9 ($p = 0.028$) for low levels of protein concentration ($\leqslant$5.4 g/dl), 6.2 ($p = 0.048$) for increased cardiothoracic ratio ($\geqslant$0.60). In univariate analysis lower AT-activity was related to pulmonary flow or systemic flow relaying on ductus arteriosus and high levels of gamma-glutamyl transpeptidase (R-square = 0.65). Lower AT-activity was no significant association with each parameter which was measured in cardiac catheterization.
Conclusion: Discrete cardiac performances were not related to lower AT-activity respectively in pre-Glenn infants. However, cardiomegaly, high concentration of brain natriuretic peptide and hepatic involvement were associated with lower AT-activity. These factors indicate cardiac stress is strong on the whole. Explanatory coefficient of these factors for lower AT-activity was relatively high. We could use lower AT-activity as method of picking up high cardiac stress comprehensively in pre-Glenn infants.

P-157
Speckle Tracking in small individuals – beware of the algorithm but not the insonation angle

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Objectives: In this study we aimed at validating the Speckle Tracking (ST) algorithm in small individuals by applying and analyzing ST (2Ch mode) from a single segment within the same animal with two different approaches for comparison of transverse strain and circumferential strain respectively.
Methods: Echocardiography (Vivid7, GE Vingmed) was performed in 14 pigs weighing less than 6 kg. First, the region of interest (ROI) was chosen parallel to the septum in parasternal long axis view for analysis of transverse strain (ParaTrans) with extraction of transverse strain from the longitudinal algorithm, and then transversely within the same segment analyzing the transverse strain with the longitudinal algorithm (LongTrans). Then we addressed circumferential strain in the basal septum in short axis viewed from two perspectives (insonation angles) perpendicular to each other (epigastric and parasternal) analyzing circumferential strain with the longitudinal algorithm (EpiCirc), then perpendicular to the same segment (ParaCirc), analyzing circumferential strain with the longitudinal algorithm for comparison. Paired t-test (mean $\pm$ SD), linear regression and Bland-Altman Plots (BAP) were performed.
Results: ParaTrans was significantly different from LongTrans ($28.2 \pm 5.4\%$ vs $32.6 \pm 9.9\%$, $p = 0.04$), however significantly correlated ($R = 0.7$, $p < 0.05$) but with a significant bias in the BAP ($R = 0.7$, $p < 0.05$). EpiCirc and ParaCirc was not different, significantly correlated ($R = 0.7$, $p < 0.05$) and with no bias in the BAP.
Conclusion: This study demonstrates a weakness within the 2Ch ST algorithm. Transverse strain measured with the longitudinal algorithm showed significant lower values with a larger variance biased relative to the magnitude of strain, therefore not capable of assessing reliable values compared to the longitudinal algorithm applied in the same segment. It also demonstrates the ST algorithms capability in assessing strain reliably regardless of insonation angle and direction within the 2D image also in small individuals.

P-158
MRI evaluation of coronary anatomy and myocardial perfusion after arterial switch for transposition of great arteries

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Background: Coronary arteries obstruction is the main cause of mortality and morbidity in patients who underwent arterial switch for transposition of the great arteries (TGA). Long-term outcome of coronary transfer and its consequences on myocardial perfusion is scarcely known.
Objective: To evaluate feasibility of cardiac MRI to describe coronary anatomy, myocardial perfusion and fibrosis after arterial switch for TGA.
Methods: 90 patients (mean age 13.5 y) were included. Twenty-five/90 had had previously documented coronary artery obstruction. cMRI procotol included cine SSFP in short axis, two-chamber, three and four chamber view, and perfusion analysis before and after dipyridamole infusion. Anatomy was evaluated by 3D heart sequence in 57 patients. Finally, late enhancement was evaluated ten minutes after injection of contrast agent in 50 patients.
Results: Perfusion could always be evaluated in this series of patients. No perfusion defect was identified but none of the 90 patient had a positive test for myocardial ischemia before cMRI. All patients with prior negative myocardial ischemia test had normal perfusion on MRI even those with mild to moderate coronary stenosis. Anatomical evaluation of proximal coronary arteries was possible in 50/57 patients. Finally, we found limited myocardial fibrosis in only two/50 patients who had left coronary artery obstruction that had been repaired.
Conclusion: cMRI is feasible and gives complementary informations on coronary artery anatomy and physiology after the arterial switch operation for TGA. Use of cMRI as a screening tool for late coronary artery obstruction in this condition has to be evaluated in large series of patients to obtain informations on its sensitivity and specificity.

P-159
New Pediatric Reference Values for myocardial strain and strain rate using 2D speckle Tracking Echocardiography

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Background: strain (ε) and strain rate (SR) measurements using speckle tracking echocardiography (STE) have been related to

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long-term outcome in congenital heart disease, but depend on the ultrasound system (US) and analysis software that is used. No normative data exists for the US iE 33 and QLAB software (Philips).
Study aim: to provide pediatric normal data for STE-derived ε and SR for an US that is widely used in pediatric and adult cardiology.
Methods: 72 healthy volunteers aged 4-18 years were studied with an iE33 US. Off-line analysis was performed using QLAB version 9.0. Circumferential ε (CS) of the left ventricle (LV) was measured from parasternal short axis views at the basal (6 segments), mid (6 segments) and apical (4 segments) levels. Longitudinal systolic ε (LS) of the LV (6 segments) and right ventricle (RV, 3 segments) and LV longitudinal systolic SR (LSSR) and early diastolic SR (LEDSR) were measured from the apical 4-chamber view. Global ε values were calculated from the average of the segmental values. Differences between age groups were tested using Anova.
Results: the measurement of ε and SR was highly feasible in this population (lowest for RV-LS; 94% and highest for LV-LS and LSSR; 100%).)

	4-9 years (n = 24) mean (SD)	10-13 years (n = 28) mean (SD)	14-18 years (n = 20) mean (SD)	Total (n = 72) mean (SD)	p-value
Global CS basal (%)	−23.9 (3.4)	−24.9 (4.0)	−24.1 (3.8)	−24.3 (3.8)	0.61
Global CS mid (%)	−26.0 (3.1)	−26.8 (4.1)	−24.9 (3.2)	−26.0 (3.6)	0.18
Global CS apical (%)	−30.7 (4.1)	−31.7 (6.5)	−31.3 (8.7)	−31.2 (6.4)	0.87
Global LV-LS (%)	−21.3 (3.1)	−19.9 (2.5%)	−19.7 (2.0)	−20.3 (2.7)	0.09
Global RV-LS (%)	−31.1 (4.1)	−28.3 (6.5)	−27.6 (3.6)	−29.0 (5.2)	0.07
Global LV-SSR (1/s)	−1.53 (0.35)	−1.31 (0.17)	−1.22 (0.16)	−1.36 (0.27)	<0.01
Global LV-EDSR (1/s)	2.29 (0.33)	1.92 (0.32)	1.89 (0.19)	2.04 (0.34)	<0.01

Conclusions: normal values for the pediatric population are presented for STE using the Philips platform. Higher SR values were found in young children versus older children, while myocardial ε values were not related to age.

P-160

Pathophysiological insights into cardiac involvement in patients with Duchenne or Becker muscular dystrophy

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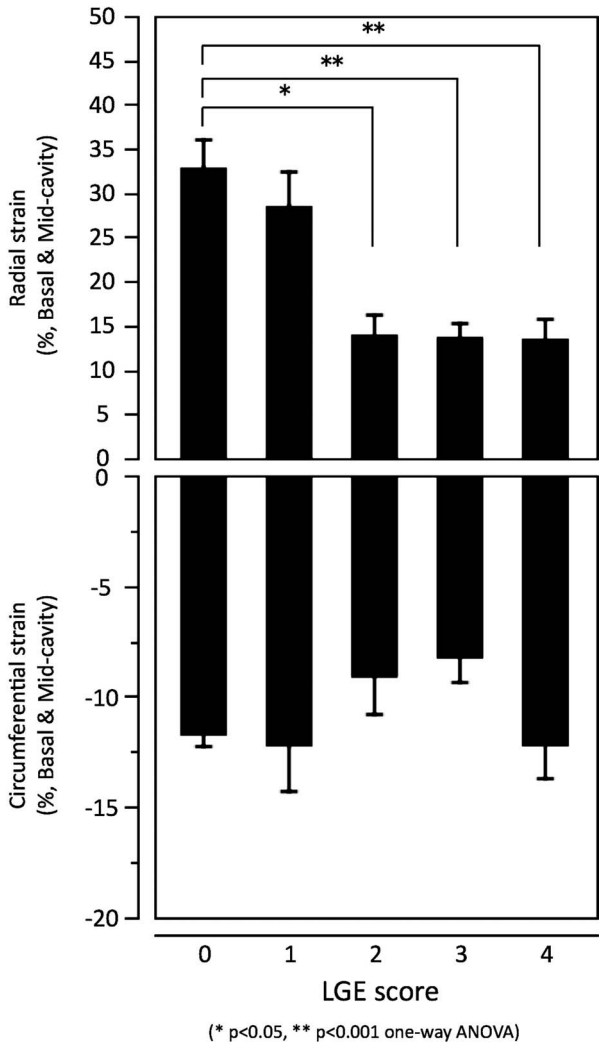
Introduction: Genetic dystrophin defects may cause myocardial fibrosis from the subepicardial layer due to intramural extension, resulting in potential regional myocardial dysfunction. For the early diagnosis and proper management of cardiac involvement in patients with Duchenne or Becker muscular dystrophy (DMD/BMD), the relationship between the extent of myocardial fibrosis and wall strain should be elucidated.
Methods: Cardiac magnetic resonance imaging (CMR) with late gadolinium enhancement (LGE) and two-dimensional speckle tracking echocardiography (STE) were performed in 7 patients with DMD/BMD (median age 11.7 years, 2.8–20.9 years) without any symptoms of heart failure. Each AHA segment except for apical segment was scored to determine the transmural extent of LGE (0=no LGE, 1 = 1-25%, 2 = 26-50%, 3 = 51-75%, and 4 = 76-100%). The relationship between the LGE score and STE strain (peak radial [RS] and peak circumferential strain [CS]) in a total of 84 myocardial segments was studied.
Results: Forty-three segments in four patients were LGE positive with predominant subepicardial involvement. The number of segments in each LGE score was n = 6 (LGE score of 1), n = 8 (2), n = 12 (3), and n = 17 (4). The RS was significantly decreased in the segments with LGE score >1 (Figure). The CS was not significantly different among the LGE score groups. ROC curve analysis revealed the cut-off RS value to predict an LGE score >2 was 22% (AUC 0.79), and the cut-off value for an LGE score of 4 was 12% (AUC 0.74).



Conclusions: Our data suggested that fibrosis of the subepicardial layer results in decreased RS, which can be detected by STE. Severely reduced RS may suggest intramural extension, which should be confirmed by CMR-LGE. Further investigation is required for detailed pathophysiological insights into cardiac involvement in patients with DMD/BMD.

P-161

Reference values for pulse doppler and tissue doppler velocities in pediatric echocardiography

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Introduction: Blood flow and tissue velocities measured by pulse Doppler are widely used tools for the evaluation of cardiac function. In healthy children, many of these parameters vary with growth, often in a non-linear fashion. Reference values for blood

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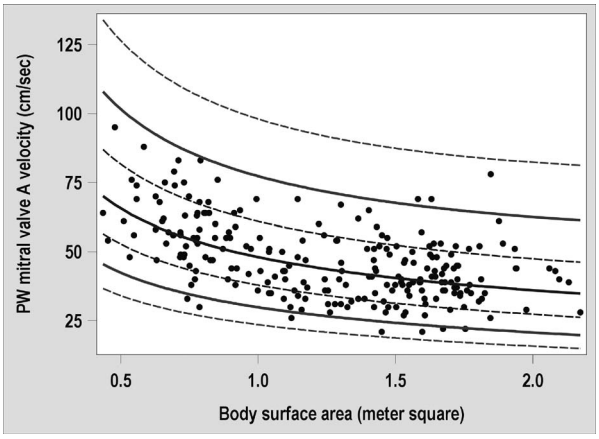
flow and tissue Doppler velocities are limited by inconsistent methodologies and small sample size. Furthermore, standardized approaches to perform normalization in pediatric echocardiography, especially in the presence of heteroscedasticity and non-linear relationships, are lacking. We aimed to determine reference values and Z scores equations for blood flow and tissue velocities in a large healthy pediatric population.



Methods: 233 healthy pediatric subjects 1-18 years of age were prospectively recruited. Fifteen pulse Doppler and 22 tissues Doppler measurements were recorded. Normalization for growth was done via a complete and standardized approach for parametric non-linear regression modeling. Several analyses were then performed to ensure adequate Z scores distribution and to detect potential residual associations with growth or residual heteroscedasticity.
Results: Most measurements adopted a non-linear relationship with growth and displayed significant heteroscedasticity. Compared to age, height and weight, normalization for body surface area was more efficient in removing the effect of growth. In general, polynomial models and allometric models yielded adequate goodness-of-fit. Residual values for several measurements had significant departure from the normal distribution. Logarithmic or reciprocal transformation was often sufficient to restore adequate distribution. Overall, weighed parametric non-linear models allowed us to compute Z score equations with adequate normal distribution and without residual association with growth. An example of Z scores boundaries for mitral A wave velocities is presented in the figure. Measurements known to be strongly dependent on heart rate often continued to have a residual association with heart rate despite adequate normalization for growth.
Conclusions: Here, we present Z scores for normalized blood flow and tissue velocities in pediatric echocardiography. Because Z scores describe the normal limit of a healthy population, further studies are needed to define the threshold beyond which health becomes disease by integrating other important factors such as ventricular morphology, loading conditions and heart rate.

P-162

Global and regional myocardial function in patients after arterial switch operation - speckle tracking study

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Introduction: Arterial switch operation/ASO/for transposition of the great arteries/TGA/was designed to restore the morphological left ventricle as the systemic ventricle, and thereby to improve cardiac function and longevity. Perfusion defects and ventricular wall motion abnormalities are late complications which affect the myocardial function and heart mechanics. Myocardial deformation imaging by 2D strain echocardiography is a novel method for assessing the ventricular myocardial function. This study aims to investigate the global myocardial function by standard and 2D strain echocardiography in children after ASO.
Methods: Medical files of patients divided in two groups were analyzed, first group - 58 children after neonatal ASO and second group - 17 healthy controls. Echocardiographic measurements by standard and Speckle Tracking Imaging/2DSE/in these groups, were compared. Apical two, three and four-chamber images (frame rate 74 ± 6 frames/s) were analyzed. Global and regional peak systolic strain (PPS) on the left ventricle (LV) was derived. The strain curves (εLL, εCC, εRR) were extracted and derived using a commercial software built on a 18-segment left ventricle. Data were presented as medians with range or means ± standard deviation. A parametric paired samples T-test integrated in the statistical software SPSS was used. A value of $p \leqslant 0,05$ was considered significant.
Results: Mean age was 5.7 ± 4 years in the ASO group and 7.6 ± 4 years in the control group, with no statistical difference. Global strain measures of the LV were significantly different between the groups (PSSLV -16,42 ± 3,08 vs. -19,29 ± 2,17, $p = 0,0001$). In the measurements of LV function, there was a clear tendency toward a decrease in the ejection fraction (63,6 ± 4,1 vs. 71,1 ± 3,2) with a concomitant increase in LV diastolic diameter (LVDD) (z-score 0.7 ± 1,0 vs. -0,3 ± 0,8). The systolic velocities were reduced significantly, especially of the medial LV segment, as well as the global longitudinal systolic strain and SR.
Conclusions: The reduced global peak systolic LVS/SR after ASO are associated with an unfavourable trend toward reduced pump function of LV. Reduced segmental LVS and SR values are a sign of regional hyperkinesia with a possible local segmental coronary ischemia during surgery.

P-163

Format for Reporting in Pediatric Echocardiography: State of the Art, Legal and Economical Implications, and Future Directions. A critical review

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Background: Various recommendations for reporting in pediatric echocardiography have been published, but a standardized format has not been established. Our aim is to evaluate strengths and limitations of published reporting formats in pediatric echocardiography.
Methods: A literature search was performed in October 2013 within the National Library of Medicine using the keywords "echocardiography", "pediatric", and "report". The research was then refined by adding the keywords "format" and "guidelines/recommendations".
Titles and abstracts of all articles identified by the search strategy were evaluated and rejected on initial screening if they did not consider reporting format (three works) or were written in languages other than English (two works).
Results: Twelve published studies that evaluated reporting for pediatric echocardiography were included in this review. Most

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Table 1. Principles of Structured, Balanced, and Practical Reporting

Consistency	Organized structure, fixed elements, and defined terminology
Flexibility	Ability to add new elements and free text
Completeness	Ability to include all potentially relevant information
Conciseness	Easily understood, quick read
Reproducibility	Independent of variable setting, disease, and operator skill
Practicality	Easily applied, pertinent to daily workflow
Ability to evolve over time	Ability to adapt to new knowledge and new techniques
Universality	Inter-operator compatibility despite variable electronic medical record systems

studies agree on the basic structure of a report in terms of organization based on the segmental approach as well as utility of common terminology and classification for various congenital heart diseases (CHD). However, the studies did not address issues such as disease severity, particularly in terms of shunt and valvar disorders. In addition, some studies recommended a unique format for all diseases, whereas others recommended different formats for different diseases. Similarly, the specific quantification components that should be part of a standard report have not yet been established. Lastly, the limitations of digital reporting as it replaced analogic technology have not been evaluated.
Conclusions: Guidelines and recommendations for reporting in pediatric echocardiography are limited. An established format for reporting may help to promote standardized care and optimize resources.

P-164
Serial assessment of the aortic conformation and distensibility in long-term Fontan survivors

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Background: Several studies have reported that aortic dilation and increased stiffness of the aorta in patients with congenital heart disease, including Fontan (F) patients, which may be an important predictor of the cardiovascular morbidity and mortality. However, the serial change of the aortic properties in F patients has not been clarified.
Methods: Ninety three postoperative F patients (F operation; 4.3 ± 3.4 years old, systemic ventricles; right ventricle 49, left ventricle 32, and biventricle 12, respectively) and 66 control subjects (C; 12.7 ± 7.1 years old) were included. All F patients underwent cardiac catheterization with aortography before and 1, 5, 10, 15 years after the operation (F0, F1, F5, F10, F15). We measured the diameters of the sinus of Valsalva (S), sinotubular junction (STJ), ascending aorta (AAo) and descending aorta (DAo) from the cine-angiogram, and calculated the ratios of the former three to DAo (S', STJ', AAo'). We also calculated the stiffness parameters of the ascending and descending aorta (β(AAo), β(DAo), respectively) with the corresponding systolic and diastolic pressures and changes in diameters.
Results: DAo were increased ($p < 0.001$) and S', STJ', and AAo' decreased over time ($p = 0.014$, 0.001, <0.001, respectively). However, S', STJ' and AAo' in Fs kept to be significantly larger than those in C (ex. AAo'; F0 2.2 ± 0.4, F1 2.1 ± 0.3, F5 2.1 ± 0.4, F10 2.0 ± 0.4, F15 1.9 ± 0.4, C 1.6 ± 0.2, $p < 0.001$). β(AAo) in Fs transitionally increased and continued to be significantly larger than those in C (F0 2.1 ± 1.7, F1 2.0 ± 1.3, F5 2.0 ± 1.2, F10 2.4 ± 1.5, F15 3.5 ± 2.7, C 1.3 ± 0.6, $p < 0.01$). On the other hand, β(DAo) didn't show any serial nor between-group variation across F0, F1, F5, F10, F15 and C.
Conclusions: Before and long after operation, Fontan patients have the stiffened dilated ascending aorta in contrast with well-distensible descending aorta. This long-standing conformational abnormality slowly improves, but the stiffness of ascending aorta may progress.

P-165
Speckle tracking echocardiography -based tissue motion annular displacement in assessing the longitudinal cardiac function among anthracycline-exposed survivors of childhood cancer

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Introduction: Our aim was to study left (LV) and right ventricular (RV) longitudinal systolic function by different echocardiographic methods and relate the results with volumetric data obtained with three-dimensional echocardiography (3DE) and cardiac magnetic resonance (CMR) among anthracycline-exposed long-term childhood cancer survivors (CCS).
Methods: Study group consisted of 75 (41 girls and 34 boys) long-term CCSs with a mean age of 14.3 ± 3.1 yrs. Median (interquartile range) cumulative anthracycline dose was 223 (163-301) mg/m^2, age at diagnosis 3.8 (2.0-6.7) yrs and follow-up time 7.1 (6.0-10.0) yrs. Control group consisted of 75 gender-, body surface area- and age-matched healthy children.
All subjects underwent conventional and speckle tracking echocardiography (STE), tissue Doppler imaging (TDI) (both apical 4-chamber view) and 3DE for LV. Sixty-one survivors also underwent CMR. Analysis on inter- and intra-observer variability for TMAD was included.
Results: CCSs had lower myocardial systolic velocities at LV basal septal (7.4 ± 1.0 vs. 7.8 ± 0.8 cm/sec, $P = 0.004$) and lateral walls (10.2 ± 1.7 vs. 10.9 ± 2.0 cm/sec, $P = 0.016$). RV lateral wall systolic velocities did not differ between the groups. STE–based tissue motion annular displacement (TMAD) values describing LV and RV systolic longitudinal fractional shortening (MAD mid% and TAD mid%, respectively) were lower among the survivors than controls (MAD mid% 15.4 ± 2.4 vs. 16.1 ± 2.2, $P = 0.049$; TAD mid% 22.5 ± 3.0 vs. 23.5 ± 3.0, $P = 0.035$), respectively. STE-derived mitral and tricuspidal annular mid-point displacements (mm, MAD mid and TAD mid, respectively) were measured. MAD mid correlated with 3DE-derived LV end-diastolic volume (EDV) ($r = 0.540$, $P < 0.001$), end-systolic volume (ESV) ($r = 0.419$, $P < 0.001$), as well as the CMR-derived LV EDV ($r = 0.463$, $P < 0.001$) and LV ESV ($r = 0.428$, $P = 0.001$). TAD mid showed correlation with the CMR-derived RV EDV ($r = 0.381$, $P = 0.002$) and RV ESV ($r = 0.334$, $P = 0.009$). Mean differences and limits of agreements for inter-observer MAD mid% were -1.3 (-5.5 to 2.9) and TAD mid% -0.9 (-6.8 to 5.0), and for intra-observer 0.1 (-2.4 to 2.5) and 0.5 (-1.7 to 2.7), respectively.
Conclusion: Anthracycline-exposed long-term CCSs have impaired LV and RV longitudinal systolic function compared with healthy controls. Assessment of the displacement of mitral

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and tricuspidal valve annulus using the STE-based TMAD is a useful and easy method for studying cardiac longitudinal function.

P-166

Dissipative Energy Loss within the Left Ventricle Detected by Vector Flow Mapping in Children: Normal Values and Effects of Age, Heart Rate, and Preload

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Introduction: The intraventricular blood flow pattern can reflect an alteration in the left ventricular (LV) function. Vector flow mapping is a novel echocardiographic technique that enables visualization of the intraventricular flow velocity vector using color Doppler cineloop images. Dissipative energy loss (EL), derived from the velocity vector field, is a flow dynamic parameter that quantifies spatial dispersion of the intraventricular blood flow. In the present study, we aimed to establish the reference value of EL within the LV in healthy children.
Methods: We reviewed the echocardiographic data of 48 children (29 males) with structurally normal hearts. Apical 5-chamber view images were used to obtain the velocity vector fields of intra-LV blood flow during 1 cardiac cycle, and the EL values during systole and diastole were estimated. The measurements were averaged over 3 cardiac cycles, and indexed to body surface area (BSA).
Results: The mean subject age was 7.1 ± 4.1 years and mean heart rate (HR) was 92 ± 17 beats/min. The mean EL was 3.30 ± 1.62 mW/m/m^{2}BSA during systole and 13.65 ± 7.96 mW/m/m^{2}BSA during diastole.
On multivariate analysis, age and HR were independent predictors of systolic EL, whereas age, HR, and E wave peak velocity were independent predictors of diastolic EL. The regression equations used to predict systolic EL and diastolic EL were as follows:
$\log_{10}$ (systolic EL) $= -0.277 - 0.00142 \times$ age (months) $+ 0.00927 \times$ HR (beats/min) (adjusted R^2 0.781; $p < 0.0001$)
$\log_{10}$ (diastolic EL) $= 0.317 - 0.00299 \times$ age (months) $+ 0.00603 \times$ HR (beats/min) $+ 0.00427 \times$ E wave peak velocity (cm/s) (adjusted R^2 0.793; $p < 0.0001$)
Conclusions: Systolic and diastolic EL were positively correlated with HR and negatively correlated with age. Moreover, diastolic EL was positively correlated with E wave peak velocity. Although the clinical implications of EL within the LV in the assessment of cardiac function are currently unclear, the present study provides reference values for systolic and diastolic EL that can be used by future studies examining patients with various heart diseases.

P-167

Right ventricular systolic and diastolic response to exercise in children after Tetralogy of Fallot repair- a bicycle exercise study

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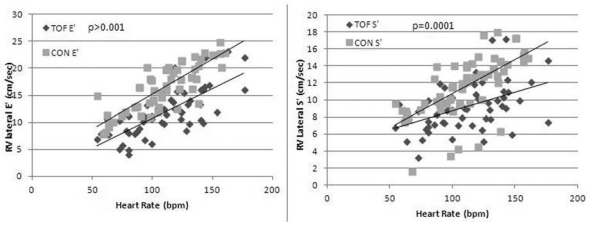
Background: Right ventricular (RV) systolic and/or diastolic dysfunction is an important clinical problem in children with repaired Tetralogy of Fallot (TOF). Tissue Doppler Imaging (TDI) represents a unique tool for measuring RV systolic and diastolic velocities of the tricuspid annulus in patients after TOF repair at rest and during exercise. The aim of the current study was to evaluate systolic and diastolic response to exercise in children with repaired TOF using semi-supine cycle ergometry stress echocardiography (SSCE).



Materials and Methods: A total of 12 children with repaired TOF and 12 age and gender matched controls were included. Median age at surgery was 6 months and median time from surgery was 11.7 years. A stepwise SSCE protocol was used. RV S' and E' were measured in all the subjects at rest and at incremental HR. Systolic and diastolic reserve was assessed by plotting RV S' and E' values against HR.
Results: Resting and peak exercise HR (mean $\pm$ SD) was not significantly different in the TOF group compared to controls (73 ± 17 vs 63 ± 13 bpm, $p = 0.33$; 141 ± 16 bpm vs 144 ± 21 bpm, $p = 0.87$). RV E' values were significantly lower at rest and peak in the TOF group compared to controls (rest: 7.8 ± 2.05 cm/s vs. 10.6 ± 2.1 cm/s, $p = 0.0005$; peak: 16 ± 4.3 cm/s vs. 20.9 ± 2.9 cm/s, $p = 0.003$). RV S' values were not significantly different at rest between the two groups (7.0 ± 1.7 cm/s vs. 8.7 ± 2.1 cm/s, $p = 0.1$) but were significantly lower at peak exercise in the TOF (9.6 ± 3.1 cm/s vs. 15.1 ± 3.2 cm/s, $p = 0.003$). RV systolic and diastolic response was blunted in TOF compared with controls (figures).
Conclusions: Our data suggest that RV systolic and diastolic response to exercise in children with repaired TOF is blunted compared to controls. The clinical implication of our finding needs further investigation but identifying early RV dysfunction could have important prognostic implications for the management of these patients.

P-168

Isovolumic Acceleration at Rest and During Exercise in Children with repaired Tetralogy of Fallot

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Background: Right ventricular (RV) systolic and/or diastolic dysfunction is an important clinical problem in children with repaired Tetralogy of Fallot (TOF). Tissue Doppler Imaging (TDI) is well suited to assess cardiac response to exercise for patients after TOF repair. RV myocardial acceleration during isovolumic contraction (IVA), a TDI derived parameter, has been proven to correlate well with indices of myocardial contractility. The force frequency relationship (FFR) reflects the increase in contractility with increasing heart rate (HR). The aim of the current study was to evaluate myocardial contractile response to exercise in children with repaired TOF using semi-supine cycle ergometry stress echocardiography (SSCE).
Materials and Methods: A total of 12 children with repaired TOF and 12 age and gender matched controls were included. Median age at surgery was 6 months and median time from surgery was

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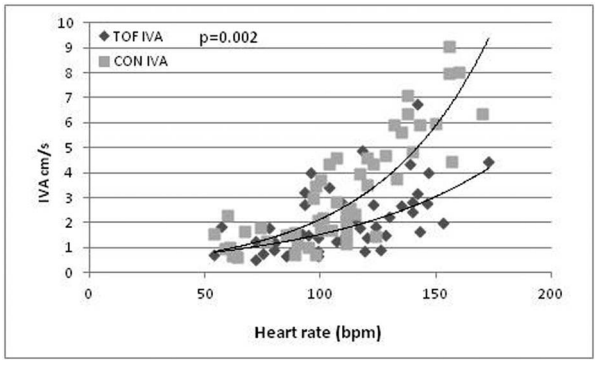
11.7 years. A stepwise SSCE protocol was used. RV IVA was measured in all the subjects at rest and at incremental HRs. FFR was constructed by plotting RV IVA against HR.



Results: Resting and peak exercise HR (mean ± SD) was not significantly different in the TOF group compared to controls (73 ± 17 vs 63 ± 13 bpm, p = 0.33; 141 ± 16 bpm vs 144 ± 21 bpm, p = 0.87). RV IVA values were not significantly different at rest in QJ;the TOF group compared to controls (0.82 ± 0.3 m/s^2 vs. 1.21 ± 0.8 m/s^2 p = 0.06) but were significantly lower at peak exercise in the TOF (2.7 ± 1.5 m/s^2 vs. 6.1 ± 2.71 m/s^2, p = 0.002). The contractile response as studied by the FFR, was blunted in TOF compared with controls (figure).
Conclusions: Our data suggest RV contractile response to exercise in children with repaired TOF is blunted compared to controls as shown by the FFR curve. The clinical implication of our finding needs further investigation but identifying subclinical RV dysfunction could have important prognostic implications for the management of these patients.

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Percutaneous closure of patent ductus arteriosus - Two decades of experience
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Introduction: Patent ductus arteriosus (PDA) corresponds to 5-10% of all congenital heart disease and is a risk factor for congestive heart failure, endocarditis and pulmonary hypertension. The percutaneous approach is now considered the method of choice for PDA closure.
Objective: Analyze the overall experience in PDA percutaneous closure for the last 23 years in a reference center of pediatric cardiology.
Methods: Retrospective analysis of intervention related covariates of patients undergoing cardiac catheterization with the intention of PDA percutaneous closure from June 1990 to May 2013. First and last five years were also compared.
Results: 623 patients underwent cardiac catheterization with PDA closure intention. Median age was 2 years, 62.1% female. Median weight was 13.3 kg. In 596 patients devices were used, 94% underwent a single intervention and in 87.6% was deployed a single device. In 4.3% of patients devices were not implanted, mostly because a non favorable anathomy. 6.4% of patients were catheterized in the first 5 years and 38.2% in the last 5 years. In the first 5 years the devices used in 89.7% of patients were Rashkind Umbrella® and in the last 5 years in 78.2% were used Coils®. Overall complication rate was 1.9% mostly due to embolization, with successful retrieval during the same procedure, except in one case. Mortality was null and there was no adverse events in the last 5 years. Fluoroscopy time was reduced (median of 17.5 minutes to 5.6 minutes) as well as the procedure duration (median 115 min to 40 min), both with statistical significance ($p < 0.01$). Residual shunt at 24h after procedure was reduced (46.2% to 10.2%, $p < 0.01$) as well as at 6 months after procedure (35.9% to 0.4%, $p = 0.1$).
Conclusions: Over the past 23 years there has been an overall improvement in the efficacy and safety of PDA percutaneous closure, with perfected technical skills and new devices available. The efficacy and low complication rate found in this study confirms that this should to be the method of choice in the treatment of PDA.

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Coarctation of the Aorta: predictors of reintervention and persistence of systemic hypertension
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Introduction: Effective correction of coarctation of the Aorta (CoA) can be associated with long term morbidity, such as recoarctation or persistence of systemic hypertension (HT). The aim of this study is to identify possible predictors for these long term complications.
Methods: Retrospective study in a reference center using patient-related and intervention type covariates such as age of intervention, weight, gender, age at follow-up, pressure gradients prior and post procedure and type of treatment (surgery vs dilation with or without stent implantation). Cox proportional hazards and logistic regression models were used for reintervention-free survival and persistence of hypertension, respectively.
Results: A total of 275 patients with CoA were analyzed: 66% male, with median age of 2 years and median weight of 12 kg; 44.7% underwent surgery (n = 123, median age 23days), 40% balloon dilatation (BD) (n = 110, median age 4y) and 15.3% underwent stent implantation (n = 42, median age 27y). 55 patients (20%) required reintervention, most frequently balloon dilatation.
The risk of reintervention was higher for patients with higher post-intervention gradients (even <20 mmHg) both by univariate (hazard ratio 1,06, $p < 0,01$) and adjusted analysis (HR 1,07, $p < 0,01$). Percutaneous methods did not differ from surgery concerning risk of reintervention in univariate analysis (hazard ratio 0,58 and 0,93, respectively for stent implantation and BD, p = NS) and multivariate analysis adjusted for age at time of treatment and initial gradient (HR 2,27 and 1,77 respectively for stent and BD, p = NS).
Persistence or late appearance of HT was predicted by higher initial gradients in both univariate and adjusted analysis (OR 1,03, $p < 0,01$ and $p < 0,05$ respectively). No statistically significant difference was found between surgical and percutaneous approach at first intervention in predicting HT at follow up on adjusted analysis.
Age at intervention or gender did not influence need for reintervention or presence of HT at long term follow up.
Conclusion: With careful selection of patients for treatment, the type of procedure does not seem to predict the need for reintervention. Higher gradients (even <20 mmHg) after surgery or intervention are predictive of need for reintervention. There is

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a correlation between higher initial gradients and presence of HT at follow up.

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Rapid left ventricular pacing during valvuloplasty

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Balloon aortic valvuloplasty is an established procedure in the treatment of aortic stenosis. During the procedure, cardiac contractions and pulsatile blood flow may cause instability of the inflated balloon, leading to failure of the procedure, suboptimal results, or damage to vessels and intraluminal structures. We report midterm results of rapid left ventricular (LV) stimulation.
Fast pacing method for valvuloplasty was performed in 56 patients. When we compare this method with standart method (74 patients), after the procedure left ventricle pressure declined from mean 160.3 ± 27.4 mmHg (96-238 mmHg) to a mean 123 ± 29.1 mmHg (73-238 mmHg). The systolic gradient of the aortic valve decreased from mean 67 ± 20.4 mmHg (26-120 mmHg) before the procedure to a mean 27.6 ± 17.8 mmHg (0-120 mmHg) after the procedure. There was no statistical difference compared to standart method ($p > 0.05$). Balloon valvuloplasty failed in two patients (3.6%) who had the procedure with a fast pacing; only 2 patients developed third grade aortic insufficiency. Among the patients who valvüloplasty performed by standard method, balloon valvuloplasty failed in 6 patients (8.3%), whereas third grade aortic insufficiency was seen in 15 (20.2%). Fast pacing balloon valvuloplasty decreased the incidence of post-procedure severe aortic insufficiency significantly ($p < 0.01$) but it had no effect on the success rate of the procedure. Fast pacing did not change the procedure duration, but decreased duration of flouroscopy significantly ($p < 0.01$).
Backup guidewires can be used effectively and safely for pacing during BAV procedures. Fast pacing can be used safely in all age groups; it decreases failure rate, eases the procedure and prevents the development of aortic insufficiency with stabilization of the balloon.

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Successful percutaneous closure of an aneurismal patent ductus arteriosus by aortic grafting and Amplatzer ASD closure device implantation

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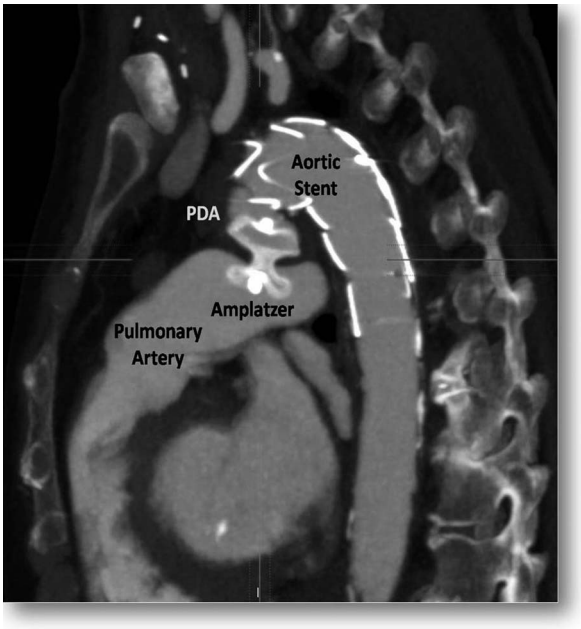
Closure of aneurismal patent ductus arteriosus (PDA) in the elderly is a high risk procedure due to the fragility of the aorta and the ductus.
We report a case of a 63 year old lady who is known to have chronic renal failure on hemodialysis, coronary artery disease and severe chronic obstructive pulmonary disease. She presented with refractory pulmonary edema and chest pain complicated by acute cardio-pulmonary arrest that needed resuscitation and support ventilation.
Her coronarography showed no significant stenosis and her echocardiography revealed a mildly dilated left ventricle with a good ejection fraction, normal valvular function and no pericardial effusion. Her chest Angio-CT uncovered a large PDA (35 × 30 mm) with peri-ductal hematoma suggesting acute fissuration.
Due to her frail physical condition, a percutaneous intervention was scheduled to close the PDA. An aortic stent graft was first placed by trans-femoral arterial approach in order to cover the aortic orifice. An Amplatzer ASD closure device was then successfully implanted on the pulmonary artery orifice of the ductus (figure: CT-Scan immediately after the intervention). The post-procedural evolution was uneventful and free of complications. A control CT-Scan that was done 6 months after the intervention showed stability of the grafts and complete isolation and involution of the PDA.



To our knowledge, this is the first time such a double grafting approach is successfully used to close percutaneously an aneurismal PDA, underlying both the feasibility and the safety of the technique.

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Early Experience in Use of the Advanta V12 Stent in Aortic Co-arctation; A Single Centre Case Series

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Introduction: In light of concerns regarding stent recoil and late failure of the Atrium Advanta V12 covered stent for severe aortic coarctation, this study examines experience with the stent design in a single center paediatric population.
Methods: All patients with Advanta V12 implanted at our center underwent serial non-invasive follow-up, retrospective case note and angiography review. P values were generated with two tailed students T-Test.
Results: Between 2005-2013, seven children underwent implantation of Advanta V12 stent for native (n = 6) or post surgical arch obstruction (n = 1). Median age at implantation was 13.5 (8-16) years. Median weight was 61.2 (26-72) kg. Time interval from implantation to latest follow-up was 15 (4.5-44) months.
Stent size ranged from 12 mm × 29 mm to 16 mm × 41 mm, according to patients' morphology and size.
Minimum aortic diameter at angiography was median 3.1 (0.8-9.1) mm, increasing to 14.1 (11.0-16.2) mm post stent placement, a mean increase of 9.4 mm ($p < 0.001$, 95% CI 24-30). Systolic

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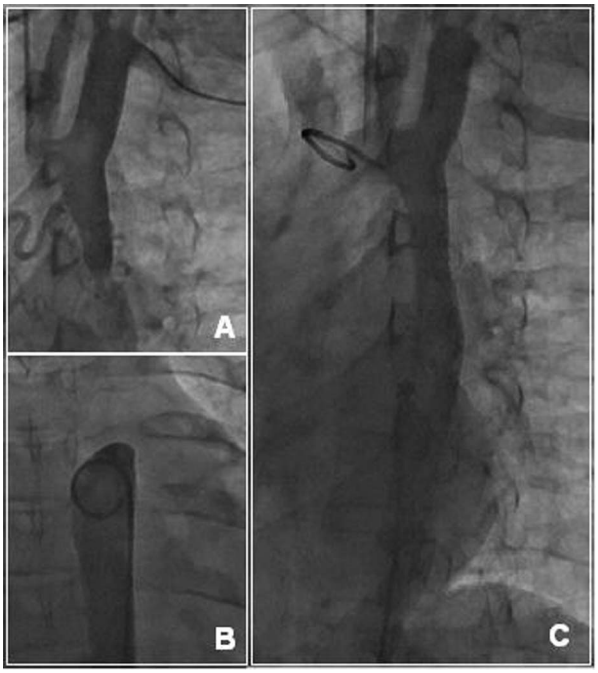
invasive pressure gradient decreased from 41(14-48) mmHg to 6(0-12) mmHg, a mean drop of 27 mmHg ($p < 0.001$ 95%CI 24-30). There were no complications. At primary inflation, stent recoil of 5-20% of diameter was observe; this was abolished in all by repeated high pressure balloon inflation or subsequent inflations with oversized balloons.



Median non-invasive SBP decreased from 150(120-165) mmHg to 127(112-148) mmHg at latest follow up, a mean decrease of 16 mmHg ($p = 0.02$ 95% CI 9-23). Serial CW Doppler evaluation over the described follow-up period did not show any increase in peak velocities, nor the presence of any notable diastolic decay.

Conclusions: In our limited experience with use of Advanta V12 covered stent in children with coarctation, no early or late complications or failures occurred. Use of the stent was effective, medium-term results were very satisfactory. It is important to contour the stent proximally to achieve complete adherence to the aortic wall, avoiding protrusion of the covered crowns into the transverse arch. Initial stent re-coil can be overcome with serial inflations or over-dilatation with a short balloon. The Advanta V12 provides a low profile, important stent design for treatment of childhood coarctation.

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Balloon valvuloplasty (BAV) as primary treatment for congenital aortic valve stenosis: a 20-year retrospective review

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Objective: The most appropriate treatment of congenital aortic valve stenosis is still under debate. We reviewed the outcomes of all 93 children 1 day to 18 years old (18 F- 75 M) treated with BAV as first line therapy from 1991 to 2012 in our institution.

Results: Mean age at BAV was 2.4 yrs, with 37 neonates (≤30 days), 29 infants (1-12 months), 27 children (≥1 year). Isolated aortic valve stenosis was present in 54 pts (58%), with Shone syndrome in 15 pts (16%) and small LV structures in 9 pts (10%). Mean f-up was 11 ± 7 years. Actuarial survival was 89% (73% for the neonates). The invasive aortic gdt (59 ± 22 mmHg) was reduced to 24 ± 12 mmHg, with a similar reduction in each age group. Four patients had significant AI post-BAV. At last echo, peak gdt was 37 ± 18 mmHg, mean gdt 23 ± 10 mmHg, 2 patients had significant AI. Most pts (58%) were free from any reintervention and 66% were free from surgery, with no correlation to age at BAV. A second BAV was performed in 6 neonates, 6 infants and 6 children, a repeat BAV (≥3) was necessary in 5 pts. Surgery was performed as 2nd intervention in 17 pts (12 Ross, 2 valvuloplasty, 3 subaortic membrane resection), as 3rd or 4th intervention in 12 pts (4 Ross, 6 valvuloplasty, 1 univentricular palliation, 2 heart transplant). Death occurred in 11 pts (10 neonates and 1 infant, 4 days to 7 yrs after initial BAV, median 25 days) in relation with inadequate LV in 7 pts, BAV complications in 4 pts (in 2 pts after 2nd BAV: 2 AR, 1 mitral tear, 1 cerebral hemorrhage during streptokinase treatment), or post-op complications in 2 pts (1 infant after coa+VSD repair, 1 valvuloplasty).

Conclusion: BAV is an efficient procedure which significantly decreased aortic gradient, with no mortality in the infants and children groups, an overall survival of 89%, 66% of pts free from surgery and 58% of pts free from any reintervention on long-term f-up. Deaths occurred mostly in the neonates group and were related to inadequate LV in 7/11, raising the difficult challenge of treating borderline LV.

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Pulmonary hemorrhage complicated with cardiac catheter examination and intervention in children

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Introduction: Pulmonary hemorrhage is the life-threatening complication of cardiac catheter examination and intervention. We investigate the frequency and risk factors of pulmonary hemorrhage, because there are a few reports about these.

Methods: We reviewed the clinical records retrospectively. From July 2003 to June 2013 (10 years), 1744 patients were done the cardiac catheter examination and intervention. From this group, we select the patients diagnosed with pulmonary hemorrhage. We defined the pulmonary hemorrhage as which was recognized by the hemoptysis or the bleeding from an intratracheal tube during or immediately after the cardiac catheter examination and intervention.

Results: 10cases (7 patients; mean age 13months) were diagnosed with pulmonary hemorrhage complicated with cardiac catheter examination and intervention. 3cases occurred in catheter examination, 2cases in pre-interventional examination, and 5cases in intervention (2 coil embolization, 2 balloon angioplasty, 1 stent placing). Pulmonary hemorrhage occurred 0.6% of all 1744 catheterized cases (examination or intervention), 0.8% of all interventions, 0.4% of all examinations. 7 cases were under 18 months old and the other 3 cases were 4 years old. Underlying diseases varied; AVSD 2, TOF 1, HLHS 1, MAPCA 1, PV stenosis 1, PA/IVS 1case. Cyanosis is recognized in 9 cases, systemic-to-pulmonary collateral vessels were recognized in the rest 1 case. Hemorrhagic spot wasn't detected in all cases. 9 cases of all 10 cases were examined under general anesthesia with intratracheal intubation. Therefore, we could handle quickly, and they could recover without sequel except one case. One case

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required resuscitation immediately after the hemorrhage due to the shock status, and it led mild dysfunction in legs.
Conclusions: Pulmonary hemorrhage occurred not only in catheter interventions but also in examinations. Children with cyanosis or under 18months old may carry the potential risk of pulmonary hemorrhage. When we treat these patients, we need tight management system at the prospect of pulmonary hemorrhage.

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Obstruction of the extracardiac conduit in children following the Fontan operation: Feasibility and results of percutaneous transcatheter stenting

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Objectives: An increasing number of Fontan patients have been reported with a significant obstruction of their extracardiac tunnel. No study has systematically examined the feasibility and the results of percutaneous transcatheter stenting of the extracardiac conduit in these patients.
Method: Our institutional database was searched to identify all Fontan patients with a significant obstruction of their extracardiac conduit who underwent percutaneous transcatheter stenting. Medical records, cardiac catheterization data, and echocardiographic investigations were reviewed.
Results: From 2011 to 2013, 10 transcatheter stenting procedures of the extracardiac conduit were performed. Patients presented with protein-losing enteropathy (n = 7), exercise intolerance (n = 2), and cyanosis (n = 1). An obstruction was suspected on echocardiography in only 4/10 patients. Median age at cardiac catheterization was 4.2 years (range 2.4–10.2 years). The median size of the stenosed area was 41% (range 28–85%). Collateral vessels were seen in 8/10 patients. We implanted 12 bare metal stents (Palmaz 4014 or 308) in 10 patients. The stents were balloon-dilated to the original size of the implanted extracardiac conduit (n = 16 mm in 7 patients, n = 18 mm in 3 patients). A unobstructed extracardiac conduit was established in all patients. No procedural complications occurred. A re-intervention was needed in 1 patient. The median follow-up after stenting was 8 months (range 1–12 months). Clinical improvement and reduction of edema was noted in all patients. However, relapses of protein-losing enteropathy occurred in all.
Conclusion: A significant obstruction of the extracardiac conduit can be encountered in a subset of patients early after Fontan completion. The feasibility and acute results of percutaneous transcatheter extracardiac conduit stenting are promising and should be recommended as the first line of treatment.

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Paediatric interventional cardiology and radiation-induced cancer risk: the Coccinelle study

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Introduction: Children with congenital heart disease frequently undergo interventional cardiology (IC) procedures for diagnostic or therapeutic 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, named Coccinelle (Cohorte sur le risque de cancer après cardiologie interventionnelle pédiatrique), is carried out in France to evaluate the risks of leukaemia and solid cancers in this population. A total number of 8000 included children is expected.
Methods: All children who have undergone at least one IC procedure since 2000 and were under 10 years old and permanent residents of France at the time of the procedure will be included. 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 cohort will be followed up through linkage with French paediatric cancer registries. Radiation exposure will be estimated retrospectively for each child included in the cohort.
Results: Up to now, 4500 children have been already included in the cohort but recruitment is still ongoing. On average, each child has undergone 1.3 IC procedures, for a total of over 5,000 procedures. Nearly half of these were performed during the first year of life. Dosimetric data were analysed for 801 IC procedures performed between 2010 and 2011. Preliminary results showed that, for diagnostic procedures, effective dose varied from 0.3 mSv to 23 mSv with a mean value of 4.8 mSv. For therapeutic procedures, effective dose varied from 0.1 mSv to 48.4 mSv with a mean value of 7.3 mSv.
Conclusions: These preliminary results revealed that therapeutic IC procedures can lead to important exposure levels. This reinforces the need to conduct epidemiologic studies such as the Coccinelle study.

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Angiographic long-term follow-up after arterial switch operation: experience of our Institution

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Introduction: The arterial switch operation (ASO) has become the surgical treatment of choice for complete Transposition of the great arteries (cTGA). However there is an increasing evidence of complications in long-term follow-up.
Objectives: Aim of the study was to detect late complications of ASO.
Methods: Between 2000 and 2013, among 150 patients (pts) treated with ASO at our Institution, cardiac catheterization was performed in 79 pts (M 54, F 25, mean age 11 y). Indication to catheterization was evidence of complication by echocardiography, by pulmonary perfusion scintigraphy or routine screening in patients aged more than 8 years old. We assesed the following features: Cardiac Index (CI), Pulmonary Vascular Resistence (PVR), pulmonary branch stenosis, aortic regurgitation (AR), aortic root dilation (ARD), coronary arteries patency.
Results: All patients were in NYHA class I, on sinus rhythm and without cardiac medication except 4. Coronary anomalies before surgery were present in 24 pts. The echocardiogram, the EKG-Holter and the ergometric stress test did not raise the suspicion of myocardial perfusion defect. CI and PVR were normal in all patients. Residual trivial to mild AR was present in 14/79 pts (17%)

and moderate to severe in 3/79 (4%); mild to moderate ARD was present in 22/79 (27%); moderate to severe ARD in 2/79 (2%). Coronary angiography showed coronary complications in 8 of 79 pts (10%): total occlusion of the main LCA in 2 asymtomatic patients and occlusion of the LDA in 1 pt. In 3 subjects there were stenosis of the LCA and in 2 cases there were a stenosis of the RCA. One subject without evidence of coronary anomalies died from ventricular fibrillation. Pulmonary arteries angiography showed stenosis in 24/79 pts (30%). Those lesions required percutaneous angioplasty in 23 pts and stent implantation in 1 with good result. *Conclusions:* Our patients treated with ASO are asymptomatic despite the relative high incidence of coronary anomalies or pulmonary branches stenosis. No evidence-based treatment is yet available for asymptomatic coronary complications, however the risk of sudden death is real. This issue should be discussed with parents and patients and strong sport and stress activities should be discouraged.

P-179

Interventional closure of perimembranous ventricular septal defects with left ventricular – right atrial communication

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Introduction: In perimembranous ventricular septal defects (pVSD) located below the attachment of the septal tricuspid valve (TV) leaflet, TV leaflets may affected by the pVSD resulting in a communication between the left ventricle (LV) and the right atrium (RA) mimicking tricuspid regurgitation. To date it has not been established if interventional closure of these VSDs can be performed with a favourable outcome.
Methods: In 4 consecutive patients (aged 6-12 years) with a hemodynamically relevant pVSD associated with moderate LV-RA shunting, the pVSD was closed under fluoroscopic guidance by establishing an arteriovenous wire-loop via a femoral artery and advancing the delivery sheath from a femoral vein. Before device release (or if necessary withdrawal), residual shunting across the device and TV valve function was investigated by transthoracic echocardiography.
Results: In two patients, the anterior TV leaflet, in two patients, the anterior and the septal TV leaflet were affected by the pVSD. pVSD sizes of 4;5.5;8;8.5 mm were closed with a 4/4;6/6 Amplatzer duct occluder II;8;10 mm Amplatzer muscular VSD occluder device, respectively. There were no or only minor residual postinterventional LV/RV and LV/RA shunts. Diastolic TV function remained normal. No conduction disturbances were observed.
Conclusions: In 4 consecutive patients, interventional closure of pVSDs associated with LV-RA communication was successful. We conclude that in this type of defects, interventional closure should be attempted.

P-180

Excluding an iliacal artery aneurysm with the new Cardiatis Peripheral Multilayer Stent

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Introduction: A 16 year old girl with an aortic coarctation was successfully treated by stent implantation. Two weeks later, signs of claudication of the right leg appeared due to an obturated iliacal artery. A balloon dilation of this vessel was performed with a good result. Four weeks later in follow up, an MRI was arranged, showing a small sized aneurysm of the iliacal artery. Further three month later, the MRI was repeated, detecting a growing aneurysm. *Method/Result:* Finally, the patient went back in the cath lab for the third time: a six french sheath was inserted in the right femoral artery, a four french sheath in the left femoral artery. After cross over angiography from the left side, the right iliacal artery with an almost seven cm long aneurysm was visualized. Knowing the diameters from the MRI, the left iliacal artery had a diameter of six mm. According to the implantation table, an eight cm long peripheral multilayer stent CPMS, Cardiatis (Isnes, Belgium) with a diameter of seven mm was successfully implanted via the six french sheath and excluded the aneurysm entirely.
Conclusion: This new Multilayer Stent is a self-expandable device with a tridimensional mesh of cobalt alloy wires interconnected in multiple layers. This new stent generation is very flexible and reduces flow velocity within the aneurysm vortex while improving laminar flow in the main artery. These favourable characteristics makes this stent a good option for excluding peripheral aneurysms, even in adolescents. Follow up data (MRI) are not available so far, but will be demonstrated in Helsinki.

P-181

Improved weight development and decrease of left ventricular enlargement after interventional closure of small and moderate sized ventricular sepal defects

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Introduction: Ventricular septal defects (VSD) are the most prevalent congenital heart diseases. Large VSDs cause heart failure, failure to thrive and pulmonary hypertension. Surgical closure is indicated early in life. Benefit from closure of small and moderate sized VSD is still unclear. The aim of this study is to evaluate growth and left heart development after interventional closure of small and moderate sized VSD.
Methods: Analysis of growth and left heart development before and after interventional VSD closure using NitOcclud Le VSD Coil.
Results: After successful and safe interventional closure of small and moderate sized VSDs in 33 patients at mean age of 6.4 ± 3.8 years significant improvement of weight development (improvement of weight for age z-score from -0.88 ± 1.42 to -0.46 ± 1.31; $p < 0.01$) and decrease of left ventricular enlargement (decrease of left ventricular enddiastolic diameter z-score from 0.93 ± 1.12 to 0.14 ± 0.89; $p < 0.01$) could be found in the mid-term follow up after 2.22 ± 1.42 years.
Conclusion: Interventional VSD closure of small and moderate sized VSD by NitOcclud Le VSD Coil in paediatric patients is safe, reduces left ventricular dilation and improves weight development in the mid-term follow up.

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Transcatheter Closure Of Coronary Artery Fistulas; Single Center Experience In 6 Years Period

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Introduction: Recently, transcatheter closure of CAFs has emerged as an alternative to surgery. We present our experience with CAF's between 2007 and 2013.

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Method: 21 patients aged between 2 months–67 years (median 7 years) underwent to cardiac catheterization. The procedure was performed preferably with retrograde approaches if it is feasible, otherwise antegrade approach was used by establishment an AV loop.
Results: In 21 patients, coronary angiograms demonstrated 23 fistulas arising from right coronary artery (RCA) (10), left anterior descending artery (7), left main coronary artery (3), circumflex artery (CX) (3). They were opening to right ventricle (11), right atrium (7), pulmonary artery (3), superior vena cava (1) and bronchial artery (1) with single (15) and multiple orifices (8). There were 4 fistulas with multiple feeding arteries. In 5 patients, the fistula was no closed since they were very small. In one pt the catheter couldn't be advanced to the distal then the procedure was abandoned. Intervention was successful in 15 of 16 patients and realized with retrograde and antegrade way in 12 and 3, respectively. Complete occlusion was achieved in cath lab in 12 immediate after, in 14 after 24 hours. Only residual shunt was another very small fistula that was left untreated in the same patient. 6 coils, 9 vascular plugs (1, 2 and 4) and 4 duct occluders were used. A patient suddenly died four days later probably due to thrombosis in huge and slow filling coronary arteries since the reciprocal competitive flow after closure. There were huge dilated RCA and CX artery communicating with each other acting as multiple feeding arteries and opening into the RV with an acute angle after short course from the RCA origin. It was closed by a vascular plug just distal to the orifice. Other patients are well without recanalization during the median 25 months of follow-up.
Conclusion: CAF's may present in a great variety in morphology. It is not uncommon to see multiple feeding arteries and especially multiple distal openings. Effective and safe percutaneous transcatheter closure is possible in majority of cases but it is not free of complications.

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Transcatheter Closure of Perimembranous and Muscular VSD with Cardiofix Muscular VSD Occluder

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Background: The difference of Cardiofix muscular VSD occluder (mVSDO) from Amplatzer mVSDO having a shorter (7 mm vs 5 mm) connecting waist choices. Taking into consideration that the muscular septum is thinner in children and also rims of perimembranous VSD (pmVSD) is slim; we preferred this device for closure of all muscular VSD and some selected pmVSDs.
Material and Method: 37 Patients underwent transcatheter VSD closure with Cardiofix mVSDO in our clinic between April 2007 and December 2013 were analyzed. During the study period Cardiofix mVSDO was used in all muscular VSD and in some selected pmVSDs in the existence of septal aneurysm by inserting the left disc of the device into the aneurysm or if there is sufficient ($\geq$4 mm) subaortic rim by leaving the left disc at the LV side.
Results: The procedure was successful 35 of 37 (94%) patients. The age of patient ranged from 1 to 34 years (median 7 years). Mean defect diameter was 8,4 mm $\pm$ 3,1 (4,3-18 mm) and mean Qp/Qs ratio was 1,86 $\pm$ 0.60. The defect types were perimembranous in 17 and muscular in 18 patients. 13 patients those pmVSDs were closed by left side, the mean distance between defect side and aortic valve (aortic rim) was 5,4 mm $\pm$ 1,1 (4-7 mm). In four patients with pmVSD the device was placed into the aneurysm. The only significantly complicated patient who developed moderate tricuspid regurgitation and significant residual shunt even after device releasing was referred for elective surgery. Full occlusion rate was 94% on follow up. There was trivial non-progressive new onset aortic regurgitation in one patient. There was no permanent complete AV block during early and mid-term follow up.
Conclusion: VSD closure with Cardiofix mVSDO having a short connecting waist (5 mm) is safe and efficacious. Shorter connecting waist than Amplatzer mVSDO is more convenient for muscular VSDs in children when regarding to septal thickness. On the other hand, it may be preferred in pmVSD closure with sufficient subaortic rim since the larger connecting waist than the eccentric pmVSD occluder may cause lesser pressure to conducting pathway to reduce the risk of AV block.

P-184

Transcatheter Occlusion of Abnormal Vascular Connections with Different Devices in Children with Congenital Heart Disease: A Single-Center Experience from Turkey

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Introduction: Transcatheter occlusion of abnormal vascular connections can be applied with a high success rate by pediatric cardiologists and interventional radiologists. The main types of abnormal vascular connections are between the aorta or its branches and the systemic veins or heart chambers, major aorto-pulmonary collateral arteries (MAPCAs), pulmonary arteriovenous fistulas, coronary fistulas, veno-venous communications, which may lead to desaturation in patients with a single ventricle in the period after Glenn or Fontan procedures, scimitar artery, and the aorto-pulmonary window. In this report, we discuss the experiences of patients who underwent transcatheter occlusion of abnormal vascular connections.

Table 1. Major indications of transcatheter occlusion of abnormal vascular connections and the devices used.

Indications	n	AVP-I	AVP-II	GC	DC	ADO-I	ADO-II	ASO
MAPCA	24	8	4	18	2			
PAVM	3	7	1					
VVC	3	2						1
CCF	3				4		1	
Ao-SVF	2						1	
SA	2			5				
APW	1					1		
Total	38	17	5	23	6	1	2	1

MAPCA: majör aorto-pulmonary collateral artery, PAVM: pulmonary arteriovenous malformation, VVC: veno-venous connection, CCF: coronary-cameral fistula, Ao-SVF: aort-systemic venous fistula, SA: scimitar artery, APW: aorto-pulmonary window.n: number of patients, AVP-I,II: Amplatzer Vasküler Plug I,II, GC: Gianturco coil, DC: Detechable coil, ADO-I,II: Amplatzer Duct Occluder I,II, ASO: Amplatzer Septal Occluder.

Methods: The charts of patients, who underwent transcatheter occlusion for abnormal vascular connections between March 2010 and December 2013, were retrospectively reviewed. A total of 38 patients (25 males) were studied.
Results: The mean age of the patients was 5.1 $\pm$ 9.0 years (range, 3 months–43 years). The mean weight of the patients was 16.1 $\pm$ 17.3 kg (range, 3.3–89 kg). The major indications of the

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transcatheter occlusion of abnormal vascular connections and the devices used for the occlusion are summarized in Table 1. Fifty-five devices were used for the occlusion of 44 abnormal vascular connections in 38 patients. In 24 patients, 32 MAPCAs were occluded. Tetralogy of Fallot was present in 13 cases, pulmonary atresia with ventricular septal defect was present in 7 cases, congenital heart disease with single ventricle physiology was present in 7 cases, and the primary pathology was closed vascular structures in the remaining 12 patients. In all the patients, the process was successfully completed. Major complications associated with occlusion were not observed in any of the patients either during and after the procedure or throughout the follow-up period The mean follow-up period for the subjects of this investigation was 17.5 ± 12.7 months (range, 1–45 months).
Conclusions: Transcatheter occlusion of abnormal vascular connections can be applied with very high success rates and low complication rates, depending on the type of pathology that is being treated. Transcatheter occlusion can be used to treat abnormal pathologies, to act as a complement to surgical treatment, or to mitigate post-operative complications.

P-185
Mid-Term Outcomes of Transcatheter Intervention for Pulmonary Atresia with Intact Ventricular Septum
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Objective: We report aİ single-institution experience and mid-term results for pulmonary atresia with intact ventricular septum (PA-IVS).
Method: Enrolled in the study were 34 PA-IVS patients who underwent transcatheter procedures in the neonatal period between April 2010 and October 2013. Six of the patients were lost in follow-up before the second stage. We retrospectively evaluated the mid-term results of the remaining 28.
Results: Out of 28 patients, 6 had a monopartite right ventricle (RV), 19–bipartite and 3–tripartite. Three patients dropped out of follow-up (2 bipartite, 1 monopartite). The remaining 25 had a mean age of 10.8 ± 6.1 months and mean weight of 8.8 ± 4.1 kg. Mean post-procedural follow-up period was 414 ± 362 days. Out of 3 tripartite patients, 2 had RF and balloon valvuloplasty performed and required no further intervention, while 1 underwent 1.5-ventricular repair. In the bipartite group, 6 patients underwent the Glenn procedure, 1 had biventricular repair and 4 had surgery and biventricular repair. Of these 4, 3 also had right ventricular outflow tract (RVOT) reconstruction, infundibular muscle resection, pulmonary valve commissurotomy and a combination of the two, respectively; only 1 patient had RVOT reconstruction due to stent obstruction. Out of the 6 patients that had the Glenn procedure, 2 were lost in the early post-operative period, and 2 also underwent RVOT reconstruction with pulmonary valve commissurotomy and pulmonary valve reconstruction, respectively. Shunt surgery was only performed in 1 patient. Two monopartite patients underwent the Glenn procedure. Three monopartite and 3 bipartite patients have very recently undergone procedures and are continuing follow-up.
Conclusion: In evaluation of PA-IVS patients, many factors influence the choice of univentricular, biventricular or 1.5-ventricular repair. We had patients with a tripartite RV that needed 1.5-ventricular repair as well as those with a bipartite RV that progressed to biventricular repair. For this reason, repair options must be considered based on RV development. RVOT reconstruction and pulmonary valve surgery may be needed for biventricular repair in bipartite patients. The survival rate in monopartite patients appears to be low.

P-186
Transcathater Closure of Atrial Septal Defects Improves Right Ventricular Function
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Introduction: Atrial septal defect (ASD) is one of the most frequently encountered congenital heart conditions. Ostium secundum ASD is the most common type, accounting for 50-70% of all cases. Transcatheter closure of ASD, has been introduced into practice 30 years ago and because of new developed devices it gained speed in the last ten years.
Right ventricular volume overload is a well-known cardiac consequence of ASD shunt. Thus cardiac volumetric unloading is a major aim of transcathater ASD closure. Echocardiographic measurement of right ventricular functios is challenging due to its complex geometrical shape. The tissue Doppler myocardial performance index (MPI) and tricuspid valve annular plane systolic excursion (TAPSE) allow assesment of right ventricular function in children.
Aim of our study is to determine the effects of atrial septal defects and their closure closure on systolic and diastolic right ventricular functions.
Material and Methods: We enrolled of 25 patients with secundum ASD that was suitable for transcathater closure and underwent succesful transcathater closure. 20 healthy, age and body mass index matched children were enrolled too. Echocardiography was performed before, 24 hours and 1 month after transcatheter closure and compared with control group.
Results: For study group mean age was 98,92 ± 41,06 months, mean BMI was 17,2 ± 4,05 kg/m^2, 56% male, 44% female mean ASD diameter was 14,56 ± 1,84 mm. For control group mean age was 101,2 ± 31,92 months, mean BMI was 16,58 ± 2,74 kg/m^2, 50% male, 50% female. Compared with control group TAPSE measurements were statistically significantly low in study group before closure (17,42 ± 2,88 mm, p = 0,001), there were statistically significant increase after the first day and first month of closure (19,27 ± 2,9, 19,59 ± 2,35, p = 0,008, p = 0,015, respectively). Compared with control group MPI measurements were statistically high in study group before closure (19,59 ± 2,35, p = 0,0001). There were a statistically decrease after the first day and first month of closure (0,48 ± 0,09, 0,33 ± 0,05, p = 0,0001, p = 0,0001 respectively).
Conclusions: Transcatheter ASD closure led to increase in TAPSE and decrease in right ventricular MPI in early period. When compared with control group; high MPI and low TAPSE measurements before closure suggests decreased right ventricular function. After transcatheter closure markedly decreased MPI and increased TAPSE show that right ventricular function improve after transcatheter ASD closure in early period.

P-187
Multiple interventional procedures in patients after multi-stage treatment of single ventricle - one center experience
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Introduction: The patients after multi-stage treatment of single ventricle often have various complications associated with

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changed physiology of circulation. Often, this situation requires a number of corrective procedures. In this paper, we present a single center experience in percutaneous performing several procedures during one session.
Methods: We analyzed patients after staged palliation of single ventricle treated in our hospital in last 10 years. The majority of them were catheterized in intention of some kind of percutaneous treatment. Multi-procedural treatment was performed in 14/86 (16%) patients, weighted 12,1 ± 2 kg (range: 10,5-18). The mean age was 34 ± 7,8 (range: 24-50) months. Ten of them were diagnosed as HLHS previously and were operated in accordance with Norwood strategy. The completion of Fontan physiology was performed in 3/14 patients. The percutaneous treatment was made 20,2 ± 7,7 months (range: 1-35) after last surgical operation.
Results: There were performed 33 interventional procedures during 14 treatment sessions (2,35 procedures/1 patient): 2 procedures in 19 patient, 3 in 5 patients during one anesthesia. Stents to pulmonary artery (PA) were implanted in 9 patients, PA-angioplasty was performed in 5, angioplasty of neoaorta-aorta connection in 8, vascular fistula was closed in 7 (including 2 recanalized additional left superior caval vein), inferior caval vein plasty was performed in 2, angioplasty of connection of superior caval vein with PA in 1 and dilation of fenestration using cutting balloon in 1 patient. The most common combination of treatment was: stent implantation to PA with angioplasty of neoaorta-aorta (8 patients) and closure of fistula with angioplasty or stent implantation to PA (6 patients). In 1 patient the arterial dissection appeared after PA-plasty and stent implantation was performed consequently. During the follow-up (20.6 ± 6.2 months) in 1 patient after angioplasty of neoaorta-aorta and stent implantation to PA, the aortic restenosis and the stent fracture occurred after 10 months. Therefore second aortic angioplasty, second stent implantation and closure of the vascular fistula were performed.
Conclusion: Methods of interventional cardiology may eliminate the adverse effects of Fontan-type correction, even in the presence of multiple clinical problems. They often allow to avoid extensive cardiac surgery, and the complication rate is very low.

P-188
Ten Years Experience of a Single Center in Percutaneous Atrial Septal Defect Closure
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Background: Nowadays percutaneous closure is prefered as treatment in pediatric ASD patients. The purposes of the study were to (a) share ten years experience of our center in percutaneous ASD closure, (b) compare the septal closure procedure using transtoracic echocardiography with the transesophageal used ones. (c) show whether transtoracic echocardiography can be sufficient in ASD closure in certain circumstances. As far as we know our study is the first study that huge number of only children patients were included in a single center.
Material and Method: 340 patients whose ASD were closed between dates December 2003-August 2013 were included in this study. Physical examinations, electrocardiograms, transtoracic echocardiography, were done before the procedure and 24 hours, 1st, 3rd, 6th months after the procedure. The patients whose ASD were closed by TEE guidance compared with ones done by TTE.
Results: There was not a statistically significance in the epidemiologic features of patients between two groups. Size of device, procedure time a was significantly higher in TEE group. But there wasn't a significant difference between the complication and residual shunt rates between two groups.
Conclusion: TEE procedure has certain risks in pediatric population. Therefore it shouldn't be done routinely in ASD closure; but only in selected cases. The usage of TTE must be increased and if the conditions are suitable TTE should be done priorily because no difference was shown in success and complication rates in ASD closure between TTE and TEE guided groups.

P-189
Postoperative outcomes and type of re-interventions after correction of truncus arteriosus (TA) in children. Long term follow-up
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Background: TA is an uncommon heart defect with frequent postoperative re-interventions-requiring anomalies.
Objective: A retrospective analysis of post-operative abnormalities and type of implemented therapy in post-TA correction pts.
Material/methods: Among 52 TA infants (nine with IAA) operated on in 1989-2012, with the median age of 1.4 months and median body weight of 3.5 kg, there were 11 (21%) early deaths, all in the years 1989-2003. Four other pts died on average 4.8 +/- 4.3 years post-correction from non-cardiac problems (n = 3) and after re-operation (n = 1). Follow-up of the remaining 37 pts was 10 ± 6.0 years before discharging to adult cardiologists at 18 years of age.
Results: Significant postoperative problems were found in 29 post-cardiac catheterization pts x = 3.2 ± 3.1 years after correction. Fourteen pts had interventional procedures, such as balloon aortic angioplasty for isthmus stenosis in six (TA and IAA pts) and angioplasty of right/left or both pulmonary arteries stenosis in 12. One patient had obturative pulmonary hypertension. Fourteen further pts underwent re-operations (a hybrid procedure with stent implantation into the stenotic pulmonary arteries-2) on the average 4.1 +/- 3.8 years post-correction due to conduit failure (stenosis-8, aneurysm-1) in nine subjects, coexisting with left/right or both pulmonary branch stenosis in five, truncal valve dysfunction in two (replacement-1, valvuloplasty-1) and aortic arch stenosis in another two pts.
The 10-year survival in our study was 71.2%. Freedom from reoperation was 70%.
Conclusions: During the last decade, we succeeded in achieving low early mortality; however, in follow-up, we noted a risk of late deaths and the necessity for relatively common re-interventions and re-operations.

P-190
Pericardial effusion related to transcatheter device closure of atrial septal defects
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Introduction: Cardiac perforation is an uncommon but important complication following transcatheter ASD occlusion. Device related erosion normally leads to a pericardial effusion. However

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the incidence of small pericardial effusions unrelated to perforation in this patient group is unreported. Aim of this study is to establish the incidence and behaviour of pericardial effusion both before and after device related ASD occlusion.
Methods: Retrospective review of echocardiograms (both pre and post closure up to 1 year follow-up) and clinical information on all patients >16 years (102 cases) undergoing trans-catheter ASD occlusion in a single institution from 2010-2014.
Results: 5/102 (4.9%, mean age 62.2 years) were found to have a rim of pericardial effusion post procedure that resolved in further follow up. 6/102 (5.9%, mean age 50.2 years) patients developed a small pericardial effusion following a device where there was none before: 2 of these resolved, 3 remained unchanged and one was smaller a year after the procedure. 9/102 (8.8%, mean age 41.7 years) patients had small pre-existing effusions prior to ASD occlusion detected on departmental echocardiograms. 2 of these 9 effusions resolved spontaneously following device closure within a year from the procedure, 4 remained unchanged and 2 got progressively smaller. In 1 patient with a pre-existing effusion, the effusion looked larger in the first three weeks after the device closure and afterwards decreased in size. In all of these cases these was no clinical or echocardiographic evidence of compromise. All cases were followed very carefully (weekly for the first month). In all patients there was significant and rapid reduction in right heart size. All patients were entirely asymptomatic except 2 who had non-specific symptoms that responded well to additional anti-inflammatories. All patients remain well until present.
Conclusions: Not all post device pericardial effusions are caused by cardiac perforation. Small pericardial effusions are a common finding prior to device closure of an ASD. In a proportion of cases these effusions can appear larger after device closure; this is presumably related to rapid reduction in right heart size. High quality echocardiography both prior to and after device closure is essential for monitoring these patients.

P-191
Follow-up after Melody revalvulation: "aggressive" prestenting has nearly abolished stent fractures; endocarditis is a concern

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Introduction: Long term function of the Melody valve depends on stent integrity and leaflet function. Follow-up data beyond 1 year are rare. Prestenting has been put forward to reduce stent fractures.
Patients and methods: Prospective ongoing interim analysis of PPVI; single center; Melody implants since 2006; systematic follow-up with dedicated database. Leaflet function analyzed by Doppler velocity across the valve or regurgitation. Chest X-ray at 6 and 12 months and thereafter annually to look for stent fractures. The registry was screened for the event of endocarditis.
Results: 109 Melody valves were implanted in 108 patients in 2006-2013; mean age 18,4 years (4,5–81,6); follow-up 2,4 years (31 days–6,9 years). In the first 8 patients no prestenting of the RVOT was performed (label recommendation). In the next 95 patients prestenting was always performed prior to or at the time of PPVI. 125 prestents were implanted until the outflow tract became a rigid tube without relative motion nor wringing; 78 pts had 1 prestent,16 pts had 2, 5 pts received 3 stents; Covered CP stents (n = 56), Andrastent XXL (n = 48), Max LD Intrastent (n = 17) and Genesis (n = 4). The PPVI was dilated to 22 mm in 71pts, to 20 mm in 28pts and in some younger kids to 18 mm (n = 6). During follow-up stent fractures were observed in 4/8 non-prestented and 2/100 prestented group ($p < 0.05$); no recompression. There was no relevant increase in peak RVOT velocity (+0,2 m/sec at 3y, 0,5 m/s at 5y); pulmonary regurgitation showed minimal change (at implantation 0,5/4; at 3y: 0,5/4, at 5y: 1/4). In 7 patients endocarditis occurred; freedom from endocarditis was 77% at 5 years. All were sterilized with antibiotic treatment, 2 patients had residual increased gradient, which in 1 pt required restenting and delayed re- PPVI.
Discussion: Aggressive prestenting of the RVOT before revalvulation offers good stent support which nearly abolishes stent recompression or fracture. Maximal dilation leads good leaflet survival. The Melody valve is vulnerable for endocarditis which is a major threat for conduit longevity.

P-192
Valve-in-valve implantation for tricuspid bioprosthetic valve failure. A multicentric experience

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Tricuspid valve-in-valve (VIV) implantation has recently emerged as a possible therapeutic option when a bioprosthetic valve degenerates. We report here a multicentric experience and a series of 5 patients.
From 2012 to end 2013, 5 patients underwent tricuspid VIV implantation. All patients had a combination of tricuspid valve stenosis and regurgitation with right heart failure requiring high dose of diuretics. In addition, patients 2 and 3 suffered from protein losing enteropathy (PLE).
Patient and catheterization data are listed in Table.

	Pt 1	Pt 2	Pt 3	Pt 4	Pt 5
Pathology	Ebstein	VSD closure	Ebstein	Ebstein	Tri. dysplasia
N° of previous surgery	3	1	3	2	1
Bioprothesis	unknown	27 mm Mitroflow	27 mm Mosaic	33 mm CE	31 mm CE
Sex	F	F	M	M	F
Age (years)	60	44	16	15	27
Prestenting	Yes	yes	No	yes	Yes
Pacing	no	no	yes	no	no
Valve	22 mm Melody	22 mm Melody	23 mm Ed. Sapien	23 mm ED Ed. Sapien	29 mm Ed. Sapien
Follow-up (months)	11	6	12	12	/

CE: Carpentier Edwards; Ed.: Edwards, Pt: patient; Tri.: tricuspid.

In pt 1, tricuspid annulus reduction was performed by 4 overlapping stents before implantation. In patients 3 and 5, valved stent embolized just after release respectively in the right ventricle and right atrium. Over the extrastiff exchange wire, Sapien valve was stabilized close to the tricuspid annulus using a self-expandable stent (Optimed) before further valve implantation with success. During follow-up, control echocardiography showed a minimal mean transvalvular gradient and no significant regurgitation. The 2 pts with PLE improved significantly the albumin level.
To conclude, tricuspid VIV implantation is an effective procedure using the Melody or the Edwards Sapien valves with

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good short and mid-term results. Knowledge of the true minimal diameter is essential to choose the appropriate valved stent. However, further studies with longer follow-up are clearly mandatory.

P-193

Complications following transcatheter ASD closure with the Amplatzer septal occluder

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The aim of this study is to report complications of transcatheter ASD closure using the Amplatzer Septal Occluder (ASO) (St Jude Medical).
From December 1999 to October 2013, 750 patients underwent ASD closure with the ASO. Closure was mostly realized under general anaesthesia and transoesophageal echocardiography control. Choice of the device diameter was established after balloon sizing and calculation of the stretched diameter.
Mean age of the patients was 31.9 ± 22 years (0.5 month–84 years). The stretched diameter was 22.5 ± 6.6 mm (5–40 mm) and device dimension 22 ± 6.7 mm (4–40 mm). Duration of the procedure was 41 ± 15 minutes (10–120 minutes) and fluoroscopic time 7.63 ± 6.65 minutes (1–92 minutes). Dose of radiation was 18.7 ± 22 Gy.cm^2 (median 12 Gy.cm^2).
Implantation succeeded in 96.3% of pts and failure was mainly related to deficient rim. No device related death was noticed. Embolization occurred in 4 pts (0.5%): 1 in the aorta, 1 in the left ventricle, and 2 in the pulmonary artery. All but one underwent surgical extraction and ASD closure. The patient with aortic embolization had percutaneous device extraction and underwent subsequently successful implantation with a larger device. No patient required blood transfusion for any groin hematoma. One patient without aortic rim had hemopericardium one month after implantation; this was corrected by drainage without any recurrence and ASD full occlusion was noticed on Doppler control. No late complication was observed. The rate of full occlusion on Doppler control is more than 90%, and the remainings have trivial shunt.
Transcatheter ASD occlusion with the Amplatzer Septal Occluder is a safe and effective procedure. The rate of immediate complication is very low with a need of surgery in less than 0.4% of patients. No device related late complications were reported up to 14 years after implantation. The risk of aortic perforation in absence of anterior rim (noticed in about 20% of pts) is trivial and not a real limitation in clinical practice.

P-194

Balloon Atrial septostomy in very low birth weight infants <1500 g. Tips and tricks From NICU to the catheterization laboratory

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The interventional management of d-TGA in VLBW infants (<1500 g) is more required with the advances in neonatal intensive care unit (NICU) and Pediatric Interventional Cardiology.
Objective: To describe our experience in balloon atrial septostomy (BAS) in VLBW patients with d-TGA.
Methods: we reported retrospectively all BAS in the infants weighting <1500 g from January 2002 to June 2013. We collected information about the vital parameters before and after BAS. We detailed BAS techniques.
Results: We reported 3 patients with a mean weight of 1333 g (1200-1500 g). d-TGA was diagnosed at a mean age of 3 days. 4 BAS attempts were performed at a mean age of 10.3 days (2-25 day). Procedures were performed in catheterization laboratory (n = 3) and in NICU (n = 1). Patients were ventilated, sedated and transported by the NICU team. Venous access was obtained by femoral vein puncture (n = 2), by femoral vein cutdown (n = 1) and by the umbilical vein (n = 1). In patient1, BAS was done under fluoroscopic control, procedure was aborted due to atrial wall perforation by the guide-wire without tamponnade (patient died few days later for non-related causes). Patient2 had 2 attempts: under echocardiographic control, atrial septal balloon dilatation was performed first because of very restrictive FO, procedure was complicated by bradycardia. Because of persistent restriction, a second attempt was done 24 h later under echocardiographic and fluoroscopic control, BAS was performed using 5 Fr. Z-Med septostomy catheter without complication. Patient3 procedure was done under echocardiographic and fluoroscopic control. BAS was performed using 4Fr. end-hole Swan Ganz catheter and it was complicated by balloon rupture. Duration of Procedures was shorter in catheterization laboratory (45 to 60 min) versus 120 min in NICU. When achieved, BAS was effective with sustained clinical improvement.
Conclusion: BAS is mandatory in VLBW patients with d-TGA and restrictive FO, as soon as possible to use the umbilical vein. Cautious manipulation of catheters and guide-wires under control by fluoroscopy and echocardiography is necessary because of frequent complications. Atrial septal balloon dilatation constitutes another therapeutic option. Procedures require close collaboration with the NICU team to avoid transport and hypothermia complications.

P-195

Balloon valvuloplasty of critical aortic valve stenosis in the first day of life – an option or treatment of choice?

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Background: Critical aortic valve stenosis carries high risk of mortality and morbidity, therefore it needs urgent diagnosis and treatment. Balloon and surgical valvuloplasty are the optional methods of treatment. The aim of this paper is to report our experience in aortic balloon valvuloplasty in critically ill newborns on the first day of life.
Methods: Between 01/1999 and 08/2013 109 patients underwent BP of critical aortic valve stenosis. Thirty seven newborns, all with prenatal diagnosis, weighting 1.7 to 4.3 kg received interventional treatment on the first day of life. Mean pressure gradient and ejection fraction were 40,4 mmHg (10-90 mmHg) and 33,8%(10-75%) respectively. 27 patients (73%) had decreased left ventricle contractility. The diameter of the aortic valve annulus ranged from 5 mm to 8,5 mm (average-7,2 mm) and the balloon to annulus ratio ranged 0,95-1,15.

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Results: All interventions were successful with reduction of pressure gradient (average 22 mmHg, $p < 0{,}001$) and increase in contractility of the left ventricle. 8 patients died early after the intervention. In 1 patient BP was performed during resuscitation, further 2 premature patients were born in 34hbd with low birth weight, 1 of them had prior fetal valvuloplasty. In 1 patient rupture of aortic cusp resulted in significant aortic insufficiency. Due to recurrence of the stenosis and/or insufficiency 8 patients required further surgical intervention 4 to 60 months after initial BP. 3 patients had Ross operation, 5 required Konno-Ross operation. One patient had concomitant plasty of stenosed mitral valve. Because of the small left ventricle 2 patients underwent modified Norwood operation. One patient had ductal stenting with bilateral banding of pulmonary arteries and at the age of 12 months was qualified to biventricular correction.
In the follow-up (4 to 84 months) 18 patients had residual pressure gradient of 18 to 44 mmHg, 9 patients have moderate and 9 mild aortic insufficiency.
Conclusions: Balloon valvuloplasty of critical aortic valve stenosis on the first day of life is effective and allows for patient's survival and eventual future surgical treatment. In our department it is a method of choice especially in patients in bad general condition or low birth weight neonates when surgical treatment is associated with high risk of complications.

P-196
Transcatheter interventions in post-Fontan patients - a single center experiance

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Introduction: Fontan palliation for univentricular hearts carries significant early and late morbidity. Most of these residual lesions or complications can be successfully managed with transcatheter interventional procedures. We analyzed our institutional experience in transcatheter treatment of post-Fontan patients.
Methods: Retrospective review of all transcatheter interventions in Fontan patients since January 2001 at Polish Mothers Memorial Hospital.
Results: A total of 145 interventions were performed in 97 patients (1-3 procedures per patient, in 29 catheterizations more than 1 intervention was performed). Before 2006 only 30 interventions were preformed compared to 115 in the most recent years ($P < 0.05$) Patients with RV dependent circulation 58 (60%) required interventions more commonly compared to patients with LV dependent circulation 37 (38%) ($P < 0.05$). Sixteen interventions were undertaken in the early postoperative period, 28 by the end of the first year after Fontan completion and 53 in the second year after surgery. Median age at intervention was 6.5 (range 2.45–24.1) years and median weight was 20 (range 9.4–114) kg. Median time interval between Fontan surgery and intervention was 15 (0–241) months. The most commonly performed intervention was fenestration closure 78 patients (37 – ASO, 31 – ADO II, 10 – covered stent). In 27 procedures pulmonary branch stenosis was treated (19 – stent implantation, 7 – stent redilation, 1 – balloon angioplasty). In 21 procedures tunnel stenosis was treated (18 – isolated balloon dilatation, 3 – stent implantation). Further 12 interventions were performed to close collaterals (11 veno-venous, 1 – arterio-venous), 5 interventions to dilate Glenn shunt and single intervention to dilate fenestration and recoarctation. Complications related to transcatheter treatment occurred during 6 interventions (4.1%): arrhythmia – 3, death due to thromboembolic complication – 1, vascular trauma – 1, device embolization – 1.
Conclusions: Transcatheter interventions belong to integral management of patients after Fontan completion. Performed interventional treatment was highly successful with low incidence of complications and resulted in significant improvement of venous pressure in the Fontan circuit, diameters of pulmonary arteries and extracardiac tunnel and increase in oxygen saturation. All of the above led to improvement in patients' hemodynamics.

P-197
Long-term follow up is not indicated after routine interventional closure of persistent arterial ducts

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Introduction and Aims: Interventional closure of persistent arterial duct (PDA) is routinely performed. Potential side effects and complications include residual shunts, haemolysis, device embolisation, and obstruction to flow in the adjoining vessels. Follow up of these patients is frequently for many years after the procedure. It is unclear whether long term follow up is necessary.
The aims of the study was to determine if (1) immediate complications after the procedure resolve during follow up (2) to determine if any new complications develop during follow up and (3) the need for long term follow in patients after interventional PDA closure.
Methods: Single centre retrospective study of all patients undergoing interventional PDA occlusion between Nov 2002 and Nov 2011. Patient's demographics and post procedural follow up echocardiographic data were collected. Patients older than 16-years, those with additional cardiac disease requiring long term follow up, pre-existing flow disturbances in the aorta or branch pulmonary arteries, those that required surgical ligation of the PDA following the intervention, incomplete records and death not related to the procedure were excluded. The primary end point was discharge from outpatient follow up. Descriptive statistics were used.
Results: 201 patients, who underwent interventional occlusion of a PDA between November 2002 and December 2011, were included. Haemolysis did not occur in any of the patients. Six patients needed re-intervention (3 for device embolisation, 3 for residual shunt). Six had mild obstruction to flow in the adjoining vessels (1 in descending aorta and 5 in LPA), all, except one of which resolved by the first follow up (within three months). None of these required any intervention. All sequelae were found prior to discharge, whilst none developed later during follow up of up to three years. 111 patients were still under follow up.
Conclusions: Complications of interventional closure of PDA are observed immediately and do no develop after the first follow up (within three months). Obstructions to adjoining vessels tend to resolve spontaneously after the procedure by the time of the first follow up. Long term follow up is not indicated in cases when no complications are seen early after the procedure.

P-198
The Bioresorbable ABSORBTM stent for coronary stenosis in children

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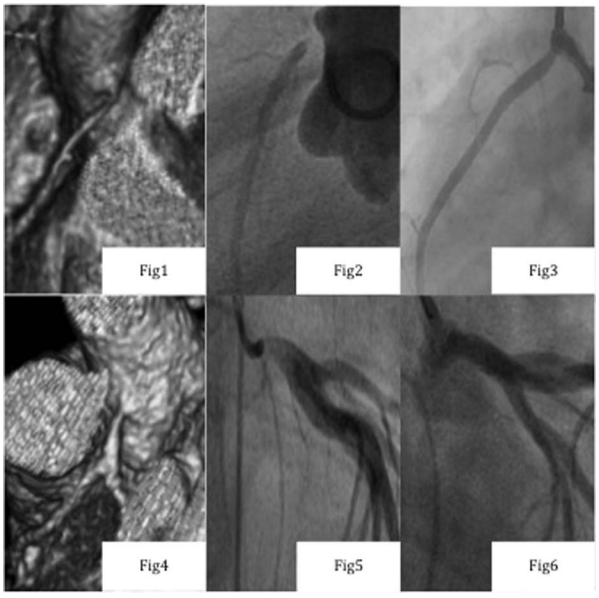
Introduction: Coronary stenosis is rare in children. Most lesions are acquired following surgical re-implantation or after Kawasaki disease. Revascularization has to take into account the small diameter of the coronary arteries and the need for growth of the child and the coronary artery.



Case report 1: a 4 year old (23 kg) boy was referred for management of asymptomatic abnormal origin of the right coronary artery arising from the left sinus. We elected for surgical reimplantation. The right coronary artery was reconnected in the anatomical position using a pericardial patch to enlarge the anastomosis. Six months later CT scan showed significant ostial stenosis (fig 1), confirmed by angiography (fig 2). Balloon angioplasty (non-compliant balloon) was unsuccessful. Implantation of a bioresorbable, everolimus coated stent (ABSORB^{TM} bioresorbable vascular scaffold- Abbott vascular) was considered. Informed consent was obtained. A 2.5 mm × 18 mm ABSORB^{TM} stent was implanted to match the distal coronary artery size, with good angiographic result (fig 3). Aspirin and clopridogel were given. Control CT scan 6 months later showed mild central restenosis that was successfully dilated with a 3 mm, non-compliant balloon.

Case report 2: a 10 years old boy (37 kg) was referred for management of stress syncope with abnormal origin of the left coronary artery arising from the right coronary artery (single coronary), with stenosed inter-arterial course. Direct left coronary trunk reimplantation was performed into the anatomical position using a pericardial patch to enlarge the anastomosis. Acute restenosis was seen on echocardiography and confirmed by CT scan (fig 4). A 3.5 mm × 12 mm ABSORB^{TM} stent was implanted (fig 5, fig 6) after informed consent was obtained. Aspirin and clopridogel were administered. CT scan performed at 6 months showed good patency and exercise test was normal.

Conclusion: Data regarding pediatric coronary artery revascularization are scarce and disappointing. Bioresorbable stent implantation to treat post-surgical stenosis is a simple procedure that allows conservation of the artery's growth capital. However, the stents have not been approved yet for use in children. Radial strength is still a matter of debate. Close medium and long-term monitoring remain essential.

P-199

Left subclavian artery to left superior vena cava fistula in a neonate: from pre-natal diagnosis to percutaneous closure

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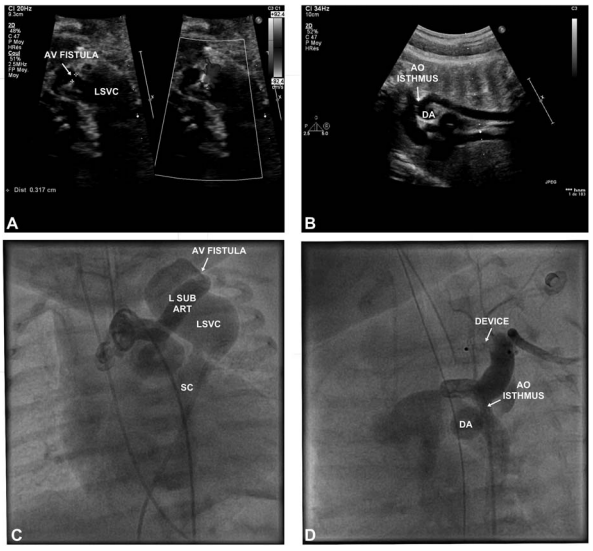
Introduction: Congenital arterio-venous fistulae involving the systemic thoracic vessels and in particular the subclavian artery and vein, innominate veins or superior vena cava are rare. Neonatal congestive heart failure is frequent. To our knowledge, prenatal diagnosis has not been reported so far. Surgical ligation is the usual treatment but the broad spectrum of devices that are currently available for percutaneous closure offer a concrete alternative.



Case report: A 40-y-old woman was referred at 19 weeks of pregnancy for management of a foetal intra-thoracic mass. Foetal echocardiography revealed a fistula between the left subclavian artery and an extremely dilated left superior vena cava (LSVC) (1A), a large and tortuous ductus arteriosus (DA) and marked aortic isthmal hypoplasia (1B). Prenatal MRI confirmed the diagnosis. The baby was born at term. After postnatal echocardiographic confirmation, intravenous prostaglandins were started to maintain the DA patent. Cardiac catheterization was performed on day 6 of life from femoral arterial and venous accesses. Aortographies showed a very dilated proximal left subclavian artery, a short and restrictive fistula (3 mm) and a very dilated LSVC (1C); isthmal hypoplasia was confirmed but without significant peak-to-peak gradient (5 mmHg). The fistula was easily crossed retrogradely and closed with an Amplatzer® ADO II 4 × 4 mm device with no residual shunt on angiography (1D) and echocardiography. Prostaglandin infusion could be weaned. Clinical evolution was uneventful without significant blood pressure gradient. The baby was discharged 4 days after the procedure. Clinical follow-up at 2 months is favorable with only mild isthmal hypoplasia and obstruction, but no arterial hypertension.

Conclusion: Precise prenatal assessment of congenital arterio-venous fistulae involving systemic thoracic vessels is possible. It allows rapid postnatal treatment before development of

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congestive heart failure. Transcatheter occlusion is a safe and effective alternative to surgery. Aortic coarctation resulting from foetal diminished flow through the isthmus may be associated.
Legend: Fig. 1 At the top: foetal echocardiography showing the fistula between left subclavian artery and LSVC (A), hypoplasia of aortic isthmus and the large ductus arteriosus (B); at the bottom: aortic angiography of the fistula before (C) and after (D) deployment of the device.

P-200
Late exercise desaturation following percutaneous atrial septal defect closure

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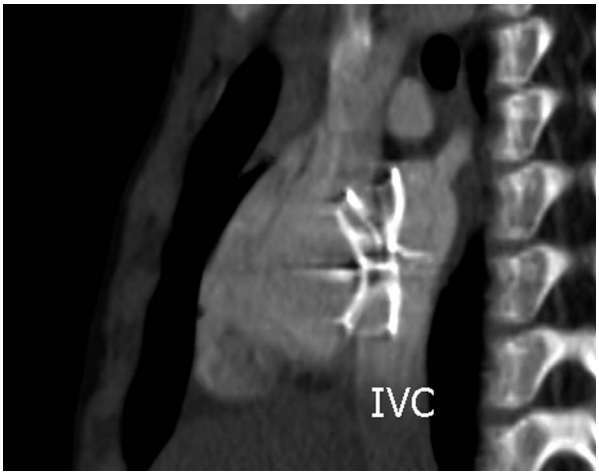
Background: Percutaneous device insertion is the preferred option for closure of secundum atrial defects (ASD). Closure rate is high and complication rate low in experienced hands. Positioning of the device may be challenging in patients with small, especially inferior rims.



Case report: a 4 year old, 14 kg girl was referred for percutaneous closure of a large ASD. Transoesophageal echocardiography described a 21 mm secundum ASD with small inferior and posterior rims. A 22 mm Amplatzer® ASO was inserted using the balloon-assisted implantation technique. Position was considered satisfactory with no residual shunt at final transoesophageal echocardiography. Aspirin was given for 6 months. Clinical evolution was uneventful over the next 3 years but echocardiograms showed abnormal retrograde flow in the inferior venae cavae (IVC) and abnormal protrusion of the device above the IVC. At closer inquiry, the child presented persistent exercise intolerance. Exercise test revealed marked desaturation (68%). CT scan showed the abnormal position of the device (figure). The child underwent surgery under cardiopulmonary bypass, through a posterior "aesthetic" right thoracotomy with femoral venous canulation. The device was well positioned and endothelialized for most of its circumference. However, at the lower part, the device was unattached and completely on the right side, protruding above the IVC lumen. A small communication between the IVC and the right atrium and a large communication between the IVC and left atrium (residual "ASD") were noted. No inferior rim was seen suggesting that the defect was not a secundum but an inferior sinus venosus type. The device was gradually dissected out and patch closure (Gore-Tex®) of the defect was performed. Post-operative evolution was uncomplicated.
Discussion: Immediate cyanosis following surgical closure of ASDs is a well-known complication, resulting from inappropriate positioning of the patch. To our knowledge this has never been described after transcatheter closure. Differential diagnosis between a secundum defect with minimal inferior rim and an inferior sinus venosus defect is difficult but important. Exercise saturation was in our patient very helpful to demonstrate abnormal right to left shunt resulting from the device malposition.
Figure: CT scan showing the device protrusion above the IVC.

P-201
Late prosthetic endocarditis 3 years after ASD transcatheter closure on an 8 year old child: case report

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Introduction: Prosthetic infective endocarditis (IE) after transcatheter closure of atrial septal defect (ASD) is extremely rare, especially beyond the first months after procedure. We report one recent late IE case which occurred on an 8 year old boy.
Case report: Our patient was born in 2005 and underwent in 2010 transcatheter closure of ostium secundum ASD with Amplatzer device. Regular follow-up was uncomplicated. During an extended stay in Morocco in summer 2013 he suffered from dental caries and episodes of fever. Several antibiotics were prescribed but blood cultures and dental car were not made. In October 2013 when returning to France he presented in our center with prolonged fever, weight loss, typical palms and soles Janeway lesions. Echocardiography showed very thick "pannus" lining the left wing of the prosthesis, with some mobile elements. MRI showed several cerebral microemboli. Blood cultures found Oxacillin-susceptible Staphylococcus Aureus. Patient underwent classical antibiotic treatment with Gentamicin (2 weeks, dose of 3 mg/kg/day i.v.) and Oxacillin (6 weeks, dose of 200 mg/kg/day i.v.). Dental care was performed. We rapidly obtained negative blood cultures. At the present time we are at the end of medical treatment. Although this technique has not been validated in this situation, we just performed a PET scan which showed no activity. Cerebral lesions on MRI remained stable. Surgical removal of the prosthesis is planned in a few days.
Conclusion: This exceptional case of severe IE on an Amplatzer ASD device 3 years after procedure should not question the recent European guidelines on prevention, diagnosis, and treatment of infective endocarditis (European Society of Cardiology, 2009). Indeed the Task Force proposes limitation of antibiotic prophylaxis to patients with the highest risk of IE undergoing the highest risk dental procedures. But in the mean time the guidelines highlight good oral hygiene and regular dental review as a very important role in reducing the risk of IE. Once ASD in successfully closed by any device, it is our pediatric cardiologist's responsibility to ensure that dental care is truly and correctly made.

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P-202

Long Term Results of Percutaneous Balloon Valvuloplasty in Infancy

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Introduction: Percutaneous balloon valvuloplasty (BV) is the treatment of choice in the management of pulmonary stenosis (PS) in infancy. Despite the excellent short and medium-term results, data on long-term outcomes with respect to exercise capacity and right ventricular (RV) function is limited. The purpose of the study was to assess the long-term functional outcomes of BV performed in infancy.
Methods: We conducted a retrospective study of 44 patients with isolated valvular PS successfully treated with BV. All children had cardiac magnetic resonance and cardiopulmonary exercise testing. Exercise data was matched using Kernel scoring to 218 healthy controls who had an exercise test at our centre. Data are presented as median (inter-quartile range).
Results: Median age at exercise test was 15.8 years (13.3-16.9). Maximum exercise tolerance (144.0 watts; 117.0-187.0; $p < 0.001$), and peak oxygen uptake (34.2 mL/kg/min; 29.6-42.9; $p < 0.05$) were significantly lower than values in healthy controls matched for age, sex, height and weight. Peak VO2 was 86.8% (71.9-104.0%) of predicted value and 43.2% of children had an abnormal peak VO2 (<84% of predicted).
Indexed RV end-diastolic volume (EDV) was 88.9 mL/m2 (78.7-112.8), which corresponded to a Z score of 2.37 (0.88-4.92), significantly deviating from the general population ($p < 0.05$). 24 patients (54.5%) had a RV EDV z-score ⩾2 suggestive of RV dilation. RV EDV z-scores correlated negatively with RV ejection fraction (EF; $R = -0.455$; $p < 0.05$). Left ventricular EDV z-score was -1.550 (-2.603—0.035), significantly ($p < 0.001$) less than the general population.
PR fraction was 18.6% (10.9-28.4). A PR fraction >15% was present in 27 patients (60%), and 8 patients (17.8%) had a PR fraction >30%. Worsening PR fraction correlated with RV EDV z-scores ($R = 0.392$; $p = 0.009$). There was no difference in peak VO2% between patients with PR fractions <15% and >15% (80.4 vs. 86.5%, $p > 0.05$).
Conclusions: The majority of children who have undergone BV for PS in infancy have at least mild PR and some degree of RV dilation as adolescents and a large proportion of them has reduced exercise tolerance. In this setting, a larger PR fraction is not associated with reduced exercise capacity.

P-203

Results of balloon aortic valvuloplasty for severe and critical aortic stenosis in neonates and children - predictors of success

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Introduction: In most centers, balloon aortic valvuloplasty (BAV) is a first line treatment, especially for neonates and infants with a congenital aortic stenosis (AS). The aim of this study was to evaluate results of BAV and identify variables associated with good immediate and midterm results of BAV in newborns and neonates.
Methods: data from cardiac catheterization (including vascular access) and echocardiography before BAV, immediately after BAV and the last available result of echocardiography study in further observation were reviewed.
Results: BAV was performed in 77 patients. The mean age at the time of the procedure was 27 ± 42,9 days. There were 59 (77%) newborns. The mean body weight was 3,64 ± 1,18 kg.58 patiens were followed, mean follow up time was 980 ± 882 days.
Significant reduction of the pressure gradient across aortic valve (PG) after BAV was found both in cardiac catheterization (15 ± 11 mmHg after BAV vs. 46 ± 26 mmHg before BAV) and in echocardiography (before BAV 74,5 ± 33,3 mmHg vs 35,5 ± 12,7 mmHg after BAV). Patients with a good result of BAV (78%) in cardiac catheterization (decrease of PG of more than 50%), had a higher PG, better left ventricular systolic function and smaller mitral regurgitation before procedure. Factors indicating good result of BAV in echocardiography (maximal PG less than 50 mmHg) immediately after procedure were a reduction of PG after BAV in cardiac catheterization and the morphology of the aortic valve. Good results of BAV in the followup was associated with the age and weight at the time of BAV and the diameter of the aortic valve. Moderate and severe aortic insufficiency (AI) was found in 24% of patients immidiatelly after BAV. Significant AI in followup was present in 62% of patients and was associated with function and size of the left ventricle pre-BAV.
Conclusions:

1. BAV is a valuable method of treatment of severe and critical AS in neonates and infants.
2. Immidiate result of BAV is associated with lef ventricle function, mitral regurgitation and morphology of the aortic valve. Result of BAV in followup is associated with the body weight and age at the time of BAV and diameter of the aortic valve.

P-204

Therapeutic strategies in patients with pulmonary atresia

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Introduction: Newborns with pulmonary atresia (PA) with or without ventricular septal defects have a severe prognosis. Their future outcome depends on the growth of the right ventricle (RV) and the pulmonary arteries (PAs). Recent observations and new interventional and hybrid procedures may improve their perspective.
Methods: We report on 12 newborns with PA and ventricular septal defect (VSD) as well as on 12 patients with PA and intact ventricular septum (IVS). Mean age at first intervention was 158 days (1d – 6.7y), bodyweight 4.3 kg (2.5-19 kg). In patients with PAVSD lung perfusion was secured by either surgery (3 Sano shunts, 3 transanular patches) or RVOT stent implantation in 5 and stenting of an atypical arterial duct (PDA) from the ascending aorta in one patient. In all patients with PAIVS the PDA was stented. High frequency perforation with balloon dilation of the pulmonary valve (PV) was necessary in 4, PV dilation only in 5 patients to increase the RV volume load. Interventional closure of RV to coronary sinusoids (RVCS) was possible in one patient.
Results: Follow up period ranges from 3 to 90 months (mean 20m). All patients with PAVSD have forward flow through the RV to the native PAs and await Fallot-repair, which has been

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already performed in two patients. Seven patients with PAIVS after PA opening and stenting of the PDA already achieved biventricular circulation. One of these 7 patients had additional RVCS which were suited for interventional closure as was done. Three patients with RVCS have univentricular palliation. Two patients died after their initial procedure, one on a pulmonary infection followed by ECMO and the other one on acute heart failure.
Conclusions: In patients with PAVSD staged interventional and/or surgical procedures can promote the growth of hypoplastic pulmonary arteries. Collaterals can therefore be closed and surgery will completely change from unifocalisation to Fallot repair. In patients with PAIVS staged interventional procedures can promote the growth of a hypoplastic RV, converting a single ventricle physiology to a biventricular scenario. The presence of RVCS remains critical, but interventional closure may become an option in some patients.

P-205
Pulmonary atresia with intact septum – Impact of right ventricular outflow tract morphology on Radiofrequency assisted pulmonary valvotomy
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Objective: Radiofrequency assisted pulmonary valvotomy is well established intervention in membranous pulmonary atresia. Successful valvotomy in these patients depends on many factors including well formed right ventricular outflow tract (RVOT). There is limited literature in relation to RVOT morphology and its impact with regards to successful immediate outcome. This study sought to assess morphology of RVOT in relation to procedural outcome.
Methods: A single-institution, retrospective review of all the patients undergoing radio frequency assisted pulmonary valvotomy between Jan. 2004-May 2013 was performed. Data was collected by reviewing the angiograms and medical notes. Procedural outcome and complications in relation to RVOT morphology were primary outcome variables.
Results: There were 88 patients found from database. Median age at intervention was 22 days (1-4015), median weight was 3.2 kg (2.3-12.4) and male: female ratio of 1:1. Mean fluoroscopy time was 28.5 minutes (9-100 min) while mean procedure time was 93.8 min. (range 20-200 min). Mean Valve plate thickness was 0.8 mm (0.47-1.3 mm). Mean valve annulus was 5.8 mm (3.6-13.6 mm). Mean RVOT-PA angle was 19.7 degrees (range 5-50). Mean energy used was 4.8 watts (range 2-6 watts). Good interface between the valve plate and MPA lumen (cupping) was found in 59% of the cases. Infundibular stenosis was present in 29% of the cases.
Eleven cases had RVOT-PA angle of more than 25 degrees. Six cases with RVOT-PA angle >25 had missed perforation as compared to none where angle was less than 25 ($p < 0.001$). Valve plate was considered thin if this was less than 0.7 mm. The thicker valve plate (>0.7 mm, n = 11/56) needed higher energy than thinner valve plate (<0.7 mm, n = 2/32) but this was not statistically significant ($p = 0.055$).
Conclusion: RVOT morphology has an important impact on successful outcome of the procedure. Well developed RVOT with thin valve plate had favorable outcome while RVOT – PA angle of more than 25 is a risk factor for misperforation. A thorough assessment of RVOT is important for favorable outcome.

P-206
Effective Radiation Dosages in 3D Rotational Angiography in Children
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Background: Three-dimensional rotational angiography (3DRA) is a relatively new but promising imaging technique in the pediatric catheterization laboratory. However, data on effective dose (ED) of this technique in children are lacking. The purpose of this study is to provide ED of 3DRA and to correlate this to parameters readily available in daily practice. Furthermore the effect of dose reducing techniques is evaluated.
Methods and Results: Effective doses were calculated with Monte Carlo PCXMC 2.0 in 14 patients who underwent a total of 17 3DRA's at our pediatric catheterization laboratory. Median age was 5.7 years (range 1 day–16.6 years). Median ED was 1.6 milliSievert (mSv) (range 0.7–4.9). ED did not correlate to age and Body Surface Area (BSA) but did correlate to Dose Area Product (DAP) and milliGray (mGy) with a r-square of 0,75 and 0,83 respectively. Reduction of the total amount of frames from 248 to 133 per rotation resulted in further dose reduction of over 50% with preserved image quality.
Conclusion: The median ED of 3DRA in children is 1.6 mSv and seems to be age and BSA independent. ED correlated to DAP and mGy in our population. By applying frame reduction a further 50% ED reduction can be achieved without loss of image quality.

P-207
Unpredictable ducts – experience in transcatheter closure of Patent Ductus Arteriosus in children born prematurely
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Background: Transcatheter closure of large tubular Patent Ductus Arteriosus (PDA) is technically demanding procedure, with significant risk of device embolization, and possibility of ductal spasm occurrence.
Material and methods: Retrospective analysis of 971 transcatheter PDA closures performed in single Institution was done to select patients with large, tortuous tubular ducts. This type of duct was found in five children born prematurely (28-30 hbd). Age at the procedure was 18-52 mths (med. 37), body weight was 12-18 kg (med. 16). Diameters of the ducts ranged from 2.5 to 7.4 mm (med. 6.4) at the pulmonary end, and from 4 to 9 mm (med. 8.4) at the aortic end, length of the ducts ranged from 12 to 16 mm (med. 14.5). Procedures were performed from the femoral artery approach. Amplatzer Duct Occluder II (ADOII, 9-PDA2-04-06, AGAMedical, Plymouth, MN, USA) was used in 4 children, detachable coil (MReye Flipper Detchable Embolization Coil, IMWCE-5-PDA5, CookMedical) in one.
Results: In two children spasm of the duct occurred during the procedure. In one of them initial aortography revealed no shunt, and it was not possible to cross the duct with the wire. Due to significant flow through the duct in control echo next day after the procedure, child was readmitted for PDA closure three months later. Twelve mm long, 2.5-4 mm wide duct was closed during second procedure with Flipper coil (IMWCE-5-PDA5, CookMedical). In second child aortography showed PDA type A (diameter at the pulmonary end 0.5 mm, diameter of ductal ampulla 3.5 mm). Flipper coil (IMWCE-3PDA4, CookMedical) was chosen. Due to unexpected change of duct morphology coil

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could not be positioned properly. Control aortography revealed 15 mm long, 9 mm wide duct, which was closed successfully with ADOII. In reminder three children duct was uneventfully closed with ADOII. Complete occlusion was achieved in all cases.
Conclusions:

1. Large tortuous tubular duct in children born prematurely is associated with high incidence of ductal spasm during the procedure (two of five cases).
2. Spasm of the duct may result in procedural failure, or improper occluder selection.
3. ADOII is an useful device for closure of large tubular ducts.

P-208

Factors influencing activated clotting time after single intravenous bolus administration of 100 IU per kilogram of unfractionated heparin during cardiac catheterization in children

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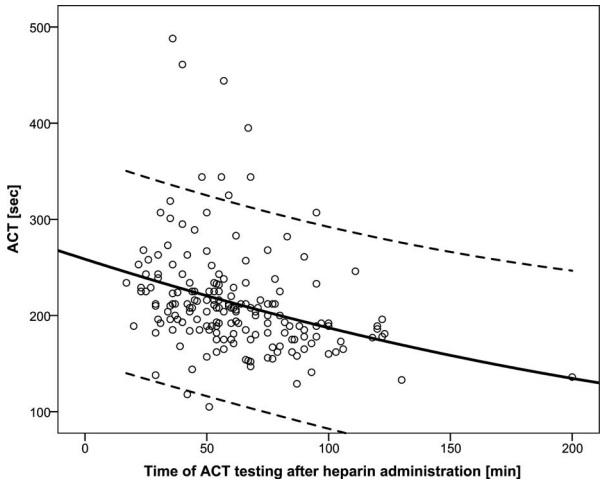
Background: Anticoagulation using single intravenous bolus administration of unfractionated heparin (UFH) as primary prevention of thrombotic events in children undergoing cardiac catheterization (CC) is standard of care. Potential side effects of UFH include severe bleeding, therefore dosage must be calculated carefully. Aim of the study was to determine the effect of UFH monitored by activated clotting time (ACT) during diagnostic and interventional CC in children.
Methods: In a retrospective single centre case study all consecutive patients undergoing diagnostic or interventional CC between April 2012 and October 2013 were evaluated. By routine, they were treated by single intravenous bolus administration of 100 IU per kilogram UFH. ACT values were measured at the end of CC.



Results: We included 183 patients (90 female) aged median 2.8 years (range 0 – 18.9 years). CCs were diagnostic in 39 and interventional in 144. ACT values were obtained at (mean ± SD) 61 ± 26 min after UFH bolus. ACT levels ranged between 105 seconds up to 488 sec, with a 25-percentile of 182 sec, 50-percentile of 204 sec and 75-percentile of 229 sec (see figure, solid line mean ACT, dotted line 95% confidence interval).
Age dependent difference of ACT values comparing different age groups from neonates to adolescents were not obtained ($p > 0.05$). The amount of total fluid volume during CC was 17.7 ml/kg (13.1–26.5) correlating with ACT values (Spearman-Rho -0.24, $p < 0.001$). Factors influencing ACT values were medications before CC such as acetylsalicylic acid (ASA) ($p = 0.002$), enoxaparine ($p < 0.05$), but not phenprocoumon ($p > 0.05$). Eight patients (4.4%) had an arterial (7 of 8) or venous (1 of 8) iliac vessel thrombosis after CC, 2 patients (1.1%) had bleeding complications, both complications after CC did not correlate with ACT values ($p > 0.05$).
Conclusions: Single intravenous bolus administration of 100 IU per kg UFH during CC in children obtains a therapeutic range of ACT values within 182 and 229 sec. ACT values are not influenced by age, but by hemodilution, antiaggregation with ASA and anticoagulation using enoxaparine. Complications after CC such as vessel thrombosis or bleeding are not influenced by this UFH regimen.

P-209

Surgical repair of atrioventricular septal defect. 10 years experience

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Introduction: The alterations in endocardial cushion and atrioventricular septation are defects characterized by varying degrees of incomplete development of the septal tissue surrounding the atrioventricular valves. Since the first successful repair of complete atrioventricular septal defect (AVSD) the treatment of this malformation has improved. Nowadays, AVSD repair can be performed with excellent midterm outcomes but late morbidity and the need for reoperation complicate the long-term results. The purpose of this study was to evaluate surgical outcome in a single center during the last 10 years.
Methods: Data of 97 consecutive patients who underwent surgical correction of AVSD between May 2003 and October 2013 at our institution were collected retrospectively. The primary endpoints in the study were in-hospital and late mortality and early and late reoperation. Other endpoints analyzed simultaneously included complications after repair: left atrioventricular valve (LAVV) regurgitation, subvalvular aortic stenosis (SAS), residual VSD, LAVV replacement, permanent pacemaker implantation and infectious complications. The association of these data with the presence of Down syndrome, preceding pulmonary artery banding (PAB), weight less than 5 kg, age less than 6 months and type complete AVSD was simultaneously analyzed.
Results: The in-hospital mortality was 3.1% (3 patients) compared with late mortality of 7.1% (7 patients). Three patients (3.1%) required an early reoperation due to severe AVV regurgitation (2) and severe residual VSD (1). Six patients (6.2%) required a late reoperation due to severe AVV regurgitation (5) and SAS (1). The estimated overall survival for all patients was 89.7% at 10 years. The estimated overall survival for patients with previous PAB was 65% at 10 years ($p = 0.001$). The estimated freedom from late reoperation for all hospital survivors without an early reoperation was 93.8% at 10 years.
Conclusions: AVSD can be carried out with good long-term results. Correction in patients weighing less than 5 kg and younger than 6 months is safe and beneficial. Palliative procedures previous to a definitive repair are no longer recommended unless other associated abnormalities make primary repair extremely a higher risk

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operation. Despite significative improvement in operative mortality, postoperative mitral regurgitation remained a concern during long-term follow-up.

P-210
Complete repair of tetralogy of Fallot in first three month period

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Objective: Complete repair of tetralogy of Fallot in first three month period has confirmed the attainability of excellent results with elective neonatal surgery avoiding palliative procedures and their morbidity and mortality.
Methods: This is a retrospective review of the results of elective three month old infants who has underwent a complete correction of tetralogy of Fallot between 2000 and 2013. Sixteen neonates and infants with either symptomatic tetralogy of Fallot, symptomatic ventricular septal defect with pulmonary stenosis or symptomatic tetralogy of Fallot with valvar pulmonary atresia underwent complete repair. Mean age at repair was 35 +/- 24 days (7-90 days) and mean weight was 3.2 +/- 0.4 kg. Palliative shunts had previously been placed elsewhere in one patient. Twelve transannular patches were used for reconstruction of the right ventricular outflow tract. In seven cases patch was not necessary. One patient need a palliative fistula before the complete repair.
Results: There were no deaths in the hospital. One patient died during the follow up due to no cardiac cause. Actuarial survival at 12 years was 95%. Actuarial freedom from need for reoperation was 100% at 5 years. All patients are symptomatically well.
Conclusions: Complete repair of congenital heart disease in neonatal period, such us "arterial switch" or "Ross op." has demonstrated excellent results. This experience with neonates suggests that, elective repair of tetralogy of Fallot could be reasonably undertaken during the first months of life. Palliative procedures are restricted to symptomatic patients with hypodevelopment of pulmonary artery.

P-211
Elevated plasma B-type natriuretic peptide and C-reactive protein levels in children with restrictive right ventricular physiology following tetralogy of Fallot repair

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Objectives: This study aimed to compare the plasma B-type natriuretic peptide (BNP) and C-reactive protein (CRP) levels in relation with oxygen transport between patients with restrictive right ventricle group (rRVG) and those without restrictive right ventricle (non-rRVG) early after tetralogy of Fallot (TOF) repair.
Methods: Eighty patients (30 in rRVG) underwent TOF repair in 2011-2012. BNP and CRP were repeatedly measured within postoperative day (POD) 7, with oxygen transport variables including arterial and superior venous oxygen saturation (SaO2 and SvO2), oxygen extraction ration (ERO2) and lactate. Demographic data included age, durations of cardiopulmonary bypass (CPB), aortic cross clamp (ACC), mechanical ventilation, ICU and hospital stays.
Results: Within POD7, BNP did not change in either of the two groups, but was consistently higher in rRVG ($p < 0.0001$). CRP increased in POD1-2, and decreased thereafter. The decrease was slower in rRVG ($p = 0.04$). The increase of SvO2 and decrease of ERO2 were slower in rRVG ($p < 0.05$). Lactate decreased in both groups ($p < 0.05$), but was consistently higher in rRVG ($p = 0.03$). BNP negatively correlated with SvO2 and preoperative SaO2, and positively correlated with ERO2 and lactate ($p < 0.05$). No correlation was found between CRP and oxygen transport variables. rRVG was older with longer CPB, ACC, mechanical ventilation, ICU and stay hospital compared with non-rRVG ($p < 0.05$ for all).
Conclusions: Restrictive right ventricular physiology is associated with significantly higher BNP and CRP levels with poorer balance of systemic oxygen transport. The information about the pathophysiological changes may help to identify the appropriate treatment strategies in this difficult group of patients.

P-212
Impact of the ventricular morphology on the early postoperative outcome after extracardiac Fontan operation

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Introduction: We sought to assess the impact of right (RV) or left (LV) systemic ventricular morphology on early postoperative morbidity after extracardiac Fontan operation (ECFO).
Methods: A total of 173 consecutive patients (median age 3.9 years, median weight 13.9 kg) underwent ECFO between 1995 and 2013. Pre- and intraoperative data and early postoperative outcomes during the first and second postoperative day were compared between the two groups (LV $n = 109$ vs. RV $n = 64$).
Results: There were no differences in any pre- and perioperative parameters between two groups, except for heterotaxy (LV 9% vs. RV 16%, $p = 0.001$) and intraoperative fenestration (LV 21% vs. RV 40%, $p = 0.06$). Early postoperative mortality (total 6.4%) and need for postoperative mechanical circulatory support were higher in the RV group ($n = 8$, 12.6% vs. LV $n = 4$, 3.6%, $p = 0.025$). Comparing the early postoperative hemodynamics (all data medians), there were no differences in mean PAP between the two groups 24 and 48 h postoperatively. On the other hand, RV patients had significantly higher LA pressures and lower mean arterial pressures (MAP24h: LV 65 mmHg vs. RV 55 mmHg, $p = 0.036$; MAP48h: LV 68 mmHg vs. RV 54 mmHg, $p = 0.002$) as well as a greater requirement for inotropic support (i.e. longer than 72 h: LV $n = 14$ of 107 (13%) vs. RV $n = 21$ of 59 (36%), $p = 0.001$). Ventilation with nitric oxide inhalation was more often necessary in RV (54% vs. 31%, $p = 0.004$). Early extubation <24 h (LV $n = 80$, 74% vs. RV $n = 31$, 50%, $p < 0.001$) was more common in LV patients but led in the total group to rapid increase of the MAP and decrease of the PAP ($p < 0.001$). Signs of acute renal failure with ascites (LV 29% vs. RV 57%, $p = 0.001$), oliguria and/or need for dialysis (LV 5% vs. RV 31%, $p < 0.001$) were observed more frequently in RV patients. Overall, patients with RV had a longer ICU stay (6 vs. 3 days in LV, $p = 0.006$).
Conclusion: RV morphology remains a risk factor for early post-Fontan morbidity despite rigorous patient selection. The optimal strategy to improve outcome in these patients is the use of fenestration and aggressive and timely reduction of the pulmonary vascular resistance.

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P-213

Recent experience and 12 years follow-up after surgical closure of atrial septal defect type II in 120 children

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Objectives: Catheter closure of secundum ASD is an effective treatment and compared favorably with surgical closure. The surgical approach is still mandatory for a significant number of patients.
Methods: This retrospective study included all 120 children (85 girls, 35 boys) operated for isolated ASD closure from 1999 until 2011 in our institution. Mean age was 4.6 ± 0.7 yrs (4 months-16 years), mean weight 17 ± 2 kg (3.6-63 kg). Perioperative course, hospitalization, and late-follow-up were analyzed.
Results: Surgical closure was effective on the first attempt in 118 patients (98.3%). Mean time of extracorporeal circulation was 38 ± 2 minutes; mean time aortic cross clamping 19 ± 2 minutes. There was 4% direct suture and 96% patch closure, 84% of the patients operated by sternotomy and 16% by thoracotomy. ICU stay was 2.4 ± 0.2 days (2-9 days), hospital stay 11.2 ± 0.9 days (4-43 days). No complication was observed in 60/120 patients (50%). Eight children (6.7%) presented major complications: 1 patient (0.8%) died from sepsis and respiratory infection, 2 (1.7%) needed a redo procedure for residual shunt, 3 (2.5%) underwent invasive treatment (2 pericardial drainage for tamponade, 1 resuscitation for cardiac arrest), 2 (1.6%) presented thromboembolic complications (1 cardiac thrombus, 1 cerebral embolism). Fifty patients (42%) had mild in hospital complications: 33 (27%) pericardial effusion requiring medical treatment (non-steroid anti-inflammatory drugs n = 25, steroids n = 8, pericardiocentesis n = 2), 15 (12.5%) infections requiring treatment (respiratory n = 9, gastroenteritis n = 5, fever without clear origin n = 3), 1 sternal instability, 4 anemia requiring transfusion, 7 (6%) pulmonary atelectasia, 2 post extubation laryngospasm (requiring steroids n = 2, reintubation n = 1). During follow-up after hospital discharge (mean 3.6 ± 0.6 yrs, 2months-13 yrs), there were 4 (3.3%) complications (2 respiratory infections, 1 wound infection, 1 fever without clear origin).
Conclusions: Our recent surgical experience of isolated ASD closure is similar to reports in the literature. Mortality is rare but not absent (0.8%). Major complications are rare (6.7%) but more frequent than the 2% complications after ASD transcatheter closure in 214 children in our institution during the same period (reported in AEPC 2013). Minor complications are frequent but did not result in sequela.

P-214

Changing Trends in Tetralogy of Fallot: Impact of Improved Antenatal Detection Rate, Earlier Catheter and Surgical Intervention

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Introduction: The management of tetralogy of Fallot (TOF) is evolving with better antenatal detection rates, and earlier catheter and surgical interventions over the years. Whether these changed have made any impact on the overall outcome in terms of morbidity and mortality is not clear. We aimed to provide a contemporary insight into the diagnostic trends, morbidity, and mortality of patients in South Wales over the past 12 years.
Method: We retrospectively reviewed preoperative and post-operative status of 92 patients diagnosed with TOF over the past 12 years.
Results: There were 92 patients with equal male to female ratio. 17% had genetic and 31% had other system abnormalities. Antenatal diagnosis rate improved from 0% to over 50% in the study period with the implementation of outflow tract screening method. Frequency of arterial shunt procedures have decreased over time from 67% in 2000 to 0% in 2012 and age at total correction changed from 15 months to less than 7 months. 12% underwent catheter palliation (including two outflow tract stent) as interim procedure. 73% of patients had transannular patch with majority being less than 6 moths old. Although the patients younger than 3 months had a longer stay in hospital (24 days) and a higher morbidity score the post-operative survival was 100% in all age groups. 4 patients died prior to total correction of TOF.
Conclusion: Majority of TOF is now detected antenatally owing to inclusion of outflow tract views as part of fetal cardiac screening programs. In parallel to this, the clinical use of propranolol for hypoxic spells was reduced substantially and the need for palliation with arterial shunt became obsolete, as alternative option of catheter interventions became readily available. Furthermore, reduction in age at complete repair to 6 months did not have adverse effect on survival, but led to an increase in the use of transannular patch.

P-215

Chest Ultrasound vs. Chest X-ray in Pediatric Patients after Congenital Cardiac Surgery - A Comparative Study

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Introduction: Postoperative management of patients with congenital heart disease requires imaging techniques to recognize complications like pleural effusions, pneumothoraces, pneumonia, atelectases or pulmonary venous congestion. Imaging techniques are as well used for verification of correct positions of chest tubes, central venous lines and endotracheal tubes. The most common technique to identify intrathoracic pathologies is chest X-ray, but particularly in pediatric patients radiation exposure is detrimental. Chest ultrasound is fast, repeatable and harmless, and has been increasingly used in pediatric patients in the last years. Yet it is unknown if ultrasound, for selected indications, is equivalent to chest X-ray. Prospective studies concerning this topic on pediatric patients following congenital heart surgery are still missing.
Methods: Prospective, blinded study on 50 pediatric patients following congenital cardiac surgery since May 2013. Evaluation of the chest ultrasound examination on first postoperative morning not knowing the corresponding, routinely performed, chest X-ray. Analysis of the findings of both imaging techniques according to the question whether the pathologies diagnosed by X-ray can adequately be seen by ultrasound. Purpose of our study is to find out if the number of X-ray images and thus radiation exposure to the patients may be reduced by the use of ultrasound.
Results: Yet 42 of 50 patients are completed. Expected end of our study will be in February 2014, final results will be presented on the AEPC meeting.
Preliminary results: 19 pts. with atelectases (2 not diagnosed by ultrasound, 13 not diagnosed by X-ray), 16 pts. with pleural

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effusions (all diagnosed by ultrasound, 14 not diagnosed by X-ray), 2 pts. with minor pneumothoraces (none diagnosed by ultrasound, all by X-ray), 24 pts. with mild pulmonary venous congestion and 2 pts. with pulmonary infiltrations.
Conclusions: Preliminary findings show that major postoperative problems as atelectases and pleural effusions were more frequently diagnosed by ultrasound than by X-ray. Diagnosis of pulmonary venous congestion and pneumonia require chest X-ray, but correspond to clinical symptoms if relevant. Chest ultrasound should be daily routine after congenital cardiac surgery. Chest X-ray should be considered for special indications, if clinical symptoms appear.

P-216

Current outcomes of the bidirectional cavopulmonary anastomosis in single ventricle patients: analysis of risk factors for morbidity and mortality, and suitability for Fontan completion

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Objectives: The bidirectional cavopulmonary anastomosis (BCPA) is considered an essential step in staged palliation for univentricular heart. This single center review aims to identify risk factors for morbidity and mortality, the role of antegrade pulmonary flow (APF), and suitability for later Fontan.
Methods: The records of 114 consecutive patients who underwent BCPA between 1992-2012, were reviewed to investigate risk factors for hospital mortality, reintubation, new drain insertion, prolonged intensive care (ICU) stay, and mortality before Fontan completion. Median age and weight at BCPA were 8 months (interquartile range (IQR) 5,7-14) and 6,9 kg (IQR 5,8-8,1). Ventricular dominance was left in 58%, 11% had bilateral caval veins. In 83% of patients, 1 to 3 interventions preceded the BCPA, with progressive increase of Norwood-type procedures over time.
Results: Extubation occurred after a median of 4 hours (IQR 3-6), mean pleural drainage was 2,2 ± 1,3 days, and median ICU stay 2 days (IQR 2-3). Ten patients needed reintubation (8,8%), 15,8% received a new drain. Hospital mortality was 11,4%, mortality awaiting Fontan 5,3%; 71% of survivors underwent Fontan, 15% are alive waiting for completion, 4% evolved to 1,5 ventricle repair. Operative mortality was independently affected by lower age ($p = 0,02$), longer bypass time ($p = 0,04$), and particularly need for reintubation ($p = 0,004$). Lower post-operative saturation, higher central venous pressure and trans-pulmonary gradient ($p = 0,01$) were risk factors for new drain insertion. Higher preoperative pulmonary pressure correlated with increased need for inotropic support, prolonged drainage and longer intubation ($p = 0,01$). Interstage mortality was mainly influenced by the ventricular function ($p = 0,028$). Additional APF resulted in a lower lactate ($p = 0,01$), higher saturations postoperatively and at discharge ($p = 0,001$), however without influencing late outcomes.
Conclusion: The interstage BCPA remains associated with adverse outcomes, mainly related to an increasing frequency of more complex UVH variations, leading to the need for BCPA at younger age. Considering the important effect of reintubation on operative mortality, perioperative surgical and medical management should focus on optimizing cardio-respiratory status. Once this selection step is taken, a successful Fontan completion can be expected if univentricular function can be maintained.

P-217

Results after surgery for vascular rings; a study of 40 consecutive cases operated 1994-2012 in a single institution

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Objective: Vascular rings are embryological malformations of the aorta and the great vessels that can give rise to complications from the encircled organs, mainly the trachea and the esophagus. Diagnosis is commonly delayed after onset of symptoms. Treatment is surgical.
Methods: Our institution serves about half of Sweden's 9 million inhabitants. From 1994-2012, 40 patients (mean age 5,7 years, range 1month to 16 years, 24 girls) underwent surgery for right aortic arch (16 cases), double aortic arch (13), aberrant right subclavian artery (9) and pulmonary sling (2). Symptoms were breathing problems, feeding/swallowing difficulties, or both. Three patients showed acute respiratory failure prior to operation. Patients had no other congenital heart defect except for PDA (2 cases). Co-morbidities were Down-syndrome (2), Turner-syndrome (1), diabetes type I (1), Behcet (1) and aberrant bronchal anatomy (2).
Treatment consisted of division of one of the double aortic arches, division of the ligamentum arteriosum, division and re-implantation of the aberrant pulmonary artery and division with or without re-implantation of the aberrant subclavian artery. A prominent or aneurysmal Kommerell diverticulum was resected or oversewn when found necessary. Aortopexy was performed as indicated.
Results: No short or long-term mortality. Complications were seen in four patients (10%) consisting of incomplete Horner syndrome (2 cases) and transient post-thoracotomy neuralgia (1). One patient required three operations for complications: ligature of lymphatic vessel for chylothorax, aortopexy for continued tracheal impression and puncture of pericardial effusions. This patient also suffered paresis of the recurrent nerve. One patient suffered tracheal fibrosis as a consequence of long-standing impression, leading to resection and end-to-end anastomosis.
Conclusions: The long time to diagnosis, severity of symptoms as well as the good short- and long-term prognosis after surgery show that these congenital malformations require a high index of suspicion in pediatricians for children with wheezing, stridor, respiratory distress and dysphagia. Surgical treatment almost completely relieving symptoms is performed without mortality and with a low complication rate.

P-218

Surgical Management of Chronic Rheumatic Mitral Valve Disease in Children: Single-Center Experience

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Introduction: The lack of consistency in approaches to surgical care of pediatric patients with chronic rheumatic mitral valve disease is of significant concern. International reports are based on small numbers of children with mixed morphology, different age at surgical correction, patients with or without previous heart surgery and various operative techniques and materials. The purpose of this study was to assess the immediate (short-term) outcome of this subgroup at one tertiary center. The primary outcome was in-hospital mortality. Secondary outcomes included need for mitral valve reoperation (secondary repair or replacement) during the same admission, utilization of postoperative mechanical circulatory support, duration of postoperative mechanical ventilation period and length of intensive care unit stay.
Methods: Prospectively collected data on preoperative, operative and postoperative outcomes of pediatric patients undergoing surgical repair of their rheumatic valve(s) between 2007 and 2012 was retrospectively analyzed.
Results: Forty-four mainly African patients, screened by international medical teams, were referred for mitral valve surgery, of who 41 underwent consecutive primary rheumatic mitral valve repair. No patient had prior surgery. Only two patients (4.9%) were scheduled for primary mitral valve replacement. However, two patients needed mitral valve replacement at initial surgery. Sixteen patients underwent concurrent tricuspid valve repair and another five aortic valve repair at the primary mitral valve operation. Early death occurred in two patients with mitral valve replacement after failure to wean from heart lung bypass despite immediate use of Extra Corporal Membrane Oxygenator. Secondary outcomes included four patients who needed mitral valve reoperation (repair n = 1, replacement n = 3) during the same admission. Only one patient underwent aortic valve replacement at the same time as the mitral valve re-operation. The median postoperative mechanical ventilation period and intensive care stay was one day (range 1-10) and 3.5 days (range 2-25), respectively. No other death occurred before the patients returned to their referring countries.
Conclusions: In young patients with severe rheumatic heart disease of the mitral valve, valve repair is the preferred approach. The present short term results regarding mortality and morbidity are encouraging.

P-219
Use of inhaled nitric oxide in paediatric cardiac intensive care unit

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Objectives: Inhaled nitric oxide (iNO), a selective pulmonary vasodilator, has been in use for the therapy of pulmonary hypertension (PH) in pediatric cardiac intensive care unit. The main concerns for its use are production of NO2 and methemoglobin, decreased platelet aggregation and increased risk of bleeding.
Patients and Method: We collected the data of 32 pediatric patients who were treated with inhaled nitric oxide at our pediatric cardiac intensive care unit between 2011 and 2012. Patients were divided into three groups.
Group I; postoperative
Group II; newborns with persistent PH
Group III; primary PH or Eisenmenger syndrome.
Age, sex, weight, primary diagnosis, arterial blood sample, pulmonary arterial pressure, systemic arterial pressure, and oxygen saturation levels were analysed.
Results: There were 25, 3 and 4 patients in group I, II and III, respectively. Eighteen of the patients were male. The median weight was 8 kilograms (3-40 kilograms), median age was 7 months (2 days-10 years). iNO was started 12 hours (1-48 hours) after admitting to the unit and was continued at a median time of 24 hours (12-168 hours). Three patients received iNO under ECMO support. In group-I, the diagnosis were atrioventricular septal defect, total anomalous pulmonary venous connection, ventricular septal defect, and truncus arteriosus.
The systolic pulmonary arterial pressure (PAP) was 40 ± 15 mmHg, systemic arterial pressure (SAP) was 57 ± 18 mmHg, PAP/SAP ratio was 0.69 and oxygen saturation levels were 88% before iNO treatment. After iNO treatment PAP decreased to 24 ± 9 mmHg ($p < 0,05$), and PAP/SAP ratio decreased to 0,4 ($p < 0.05$), SAP did not change (60 ± 12 mmHg), and saturation levels increased to 98% ($p < 0.05$). Methemoglobinemia occurred in all patients but only 3 of them needed treatment adjustment or modification. Thrombocytopenia developed in 2 patients.
Conclusion: iNO seem to be effective drug in reducing pulmonary arterial pressure. During pulmonary hypertensive crisis iNO can be used effectively and safely in pediatric cardiac intensive care units. Clinicians should be careful about methemoglobinemia and thrombocytopenia.

P-220
Left Pulmonary Artery Obstruction Due to a Large Congenital Thymic Cyst: A Rare Pulmonary Stenosis Cause

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Objective: Thymic cysts occur relatively rarely and account for only about 3% of all anterior mediastinal masses. Mediastinal thymic cysts are usually asymptomatic and are incidentally found on routine chest roentgenograms. They may rarely cause symptoms of vascular obstruction. This study presents a case of thymic cyst that caused pulmonary artery obstruction and respiratory symptoms.
Methods: A 9-year-old boy was admitted to our hospital with shortness of breath on exercise that had been ongoing for one month. On admission, his physical examination and routine blood tests were within normal limits. The chest X-ray showed a large round-shaped opacity of left perihilar localization. Transthoracic echocardiography revealed a solid-cystic mass obstructing the left pulmonary artery with a 20 mmHg systolic pressure gradient. Computed tomography of the thorax revealed a round shaped, smooth boundary cystic tumor, in close proximity to the main and left pulmonary artery in the anterior mediastinum. Surgical exploration by median sternotomy revealed an encapsulated multilocular cyst arising from the left lobe of the thymus. The cyst was localized posterior of the left phrenic nerve and in close proximity to the main and left pulmonary artery. It was completely removed by a resection of the thymus.
Results: After an uneventful recovery, the patient was discharged on postoperative day 3. The gross macroscopic examination revealed a unilocular cyst, measuring $80 \times 65 \times 15$ mm with a cyst wall thickness of approximately 3 mm. The posterior wall of the cyst was thickened with granulation. The histological and cytological examination revealed cuboidal epithelium and lymphocytes, which were on benign pattern. The pathological diagnosis was congenital thymic cyst.

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Conclusions: These cysts are usually asymptomatic, they may cause large variety of symptoms. Although thymic cysts are benign lesions, there are more malign lesions with cystic changes including thymoma, teratoma, lymphoma or seminoma. For that reason, most authors agree that surgical resection remains the curative treatment of choice, and histological examination is the only definitive means of diagnosis. In summary, this is the first report of a congenial thymic cyst in which pulmonary artery compression related symptoms and relieved of symptoms after surgical treatment were shown.

P-221

Risk factors for thrombosis, overshunting and death in infants after Modified Blalock-Taussig-Thomas Shunt

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Objective: We evaluated 44 subjects under 2 years old who had Modified Blalock Taussig shunt (MBTS) since 2009 to 2013 to investigate risk factors for thrombosis, overshunting and death.
Methods: Fourty four patients under 2 years of age, undergone MBTS procedure in our hospital from 2009 to 2013 were included in this observational study. Severe cyanotic newborns with pulmonary stenosis or atresia and duct dependent circulation, infants having TOF with small pulmonary arteries were the indications for surgery. Pre operative characteristics such as hemoglobin, hematocrite, mean platelet volume, protrombin time, partial tromboplastin time and post operative medication were noted. Risk factors for post-operative overcirculation, thrombosis and death were investigated.
Results: Patients' age and weight at the time of procedure were ranged from 1 day to 20 months (median 12 days) and 2,4 kg to 12 kg (mean 4,6 kg), respectively. 8 patients died after surgery at a median of 8 days (1-31 days) of which 4 patients were diagnosed with Tetralogy of Fallot (TOF), 2 patients with Pulmonary atresia (PA) and intact ventricular septum, 1 patient with PA and ventricular septal defect, 1 patient with tricuspid atresia and pulmonary stenosis. The mortality rate was %18,2. 4 patients (9,1%), had shunt thrombosis. 3 of 4 did not receive heparin infusion while one had shunt thrombosis on post operative sixth day despite immediate therapeutic heparin infusion administered for 24 hours. Partial thromboplastin time (aPTT) was 28.7 seconds in patients with thrombosis, 35 in rest. ($p = 0,04$). Overcirculation was detected in 5 patients by echocardiography. Shunt size/body weight ratio was 1,25 in patients who had overcirculation, 1,06 in rest. But the difference was not statistically significant.
Conclusion: Modified Blalock Taussig Shunt operation stands out as a good option when total correction is impossible in infants with cyanotic heart disease especially in developing countries. So it is important to assess risk factors associated with the procedure. A preoperative low aPTT value may be an indicator for thrombosis in infants undergone MBTS surgery.

P-222

Pulmonary aspergilloma – a very rare complication after delayed sternal closure in a neonate

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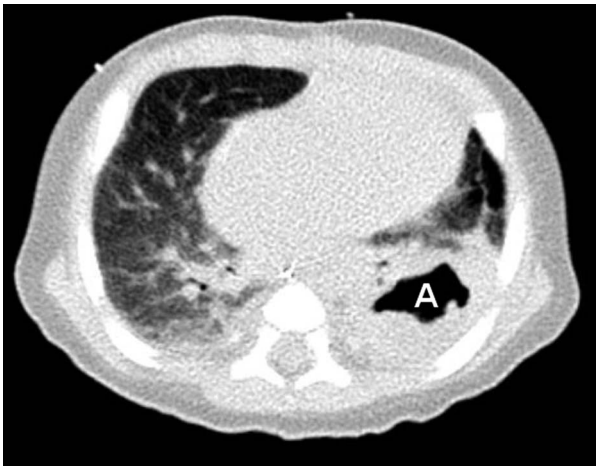
We report of a child with hypoplastic left heart syndrome (HLHS) after Norwood I procedure and invasive aspergillosis progressing to cavitary pulmonary aspergillosis.
Case report: The child was born at term after prenatal detection of HLHS with mitral and aortic atresia. The foramen ovale was severely restrictive with additional communication via an atypical interatrial venous network. Despite facial dysmorphism standard genetic tests were normal. The child underwent Norwood I procedure on day 5 of life with postoperative transthoracic venoarterial ECMO therapy for 3 days. Sternal closure was on day 6 post surgery. Due to rising inflammation markers and clinical signs of infection broad spectrum antibiotic treatment was initiated. Tracheal aspirate on day 11 after surgery grew aspergillus fumigatus with simultaneous positive galactomannan antigen. Treatment with voriconazole was initiated. 20 days after surgery a hyperlucent lesion on chest X-ray evolved that was further characterized by CT as an aspergilloma of 2×3 cm of the left lower lobe (figure; A – aspergilloma). Follow-up imaging showed decreasing size of the lesion under longterm oral therapy with voriconazole. The child successfully underwent bidirectional Glenn at the age of 6 months and antifungal therapy was stopped one month thereafter. Eight months after discontinuation the child remains in stable clinical condition.



Discussion: Nosocomial infections are an increasing problem, including fungal infections. Although antifungal treatment was initiated in time, pneumonia evolved to pulmonary aspergilloma, which is very rare in infants. The contribution of transthoracic ECMO therapy with open chest for 6 days remains unclear. Usually single aspergillomas are treated surgically. In patients undergoing Fontan pathway an optimal pulmonary status is a prerexquisite. Since total or subtotal resection of a lung lobe was thought to put the child at a higher risk in the long run, we opted for longterm antifungal therapy. Optimal duration of antifungal therapy remains unclear and was stopped in our patient after six months of therapy after successful Glenn procedure.

P-223

Surgical Age and Morbidity after Arterial Switch of Transposition of the Great Arteries

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Background: Although transposition of the great arteries (TGA) accounts for less than 5% of congenital heart disease, the clinical

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course is often dramatic with need for early diagnosis and careful preoperative care as well as advanced surgical correction and postoperative support. Since 1993, Lund is one of the two tertiary referral centers for pediatric cardiac surgery in Sweden, with nearly 400 surgical procedures each year.
Methods: A single-institution 12-year retrospective survey of 127 neonates and infants (median for birth weight, gestational week, and age at surgery: 3.5 kg, 39 weeks, and 4 days, respectively) with TGA corrected via arterial switch operation (ASO). Postoperative morbidity and mortality during the hospital stay were reviewed. Patients with double outlet right ventricle and chromosome abnormalities were excluded. "Major postoperative morbidity" (MPM) was defined as presence of 1 or more of the following: prolonged mechanical ventilation (MV), delayed sternum closure, reoperation, CPAP/NIV after extubation, and ECMO. Patients were grouped based on distance between Lund and referral clinic as follows: "local"- within 200 km radius (n = 67), and "external" >200 km (n = 60).
Results: There was only 1 death, born preterm (gestational week 34) with a body weight <2500 g. Prematurity (<36 weeks of gestation, n = 5) was associated with significant increase in postoperative morbidity (p = 0.01). Among those born full term, neither early (ie, ≤2 days, n = 19) nor late (>7 days, n = 25) surgical age had impact on MPM (p > 0.4). Among those without fetal diagnosis of TGA, neither age at surgery (p = 0.8) nor MPO (p = 0.5) differed between "local" and "external" groups.
Conclusion: ASO can be performed safely in full term neonates and in infants with TGA regardless of surgical age. This finding, along with the similar postoperative outcome regardless the distance between Lund and the referral clinic lend further support to the concept of centralization of pediatric cardiac surgery.

P-224
Sildenafil in the postoperative course after surgery in children with CHD
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Pulmonary hypertension (PHT) is one main cause of post-operative morbidity and mortality after repair of CHD. Although efficacy of iNO is well-established, the utilization of sildenafil is not yet clearly defined. The aim of this study was to assess efficacy of sildenafil after CHD repair in children.
Methods: Retrospective analysis of patients who received sildenafil in postoperative course after CHD repair, from 2005 to 2012. Duration of mechanical ventilation, CICU and in-hospital stay, pulmonary pressure and overall outcome were asssessed. Patients were divided into group I (sildenafil onset <H24) and group II (>H24).
Results: 50 patients were included: 30 in group I and 20 in II. Mean age was 11.5mos, (med3.7mos), mean weight 4.4 kg. CHD included: 20VSD, 2VSD+coarctation, 9AVSD, 3truncus arteriosus, 2TGV, 3TOF, 9abnormal pulmonary venous return, 3miscellaneous. Mechanical ventilation duration was 8.4d, CICU stay 12.9d, hospital stay 21.6d. Bypass time was 100.8 ± 45mn, aortic clamp time 60.2 ± 22mn. InhaledNO was administered within 5mn post-bypass, at 11.3 ± 5.5ppm and duration of 4 ± 3 days. Sildenafil dose was 0.25-2 mg/kg/4 h, 1stdose was 0.67 mg/kg/4 h. Mean sildenafil dose was 1.16 mg/kg/4 h at the time of iNO withdrawal. Duration of sildenafil was 22.4 ± 14.6 days. Preoperative systolic pulmonary pressure (PAPs) to systolic aortic pressure (PAs) ratio was 0.94 ± 0.1. Preoperative PAPs decreased from 72 to 36.5 mmhg at iNO cessation, PAPs/PAs from 0.94 to 0.43 and PAPmean/PAmean ratio from 0.8 to 0.46. PaO2/FiO2 ratio increased from 121.4 at end of bypass to 269 at iNO cessation. Duration of mechanical ventilation was 6.2d vs 11.6d (p = 0.04), CICU stay 9.4d vs 18d (p = 0.005), in-hospital stay 16d vs 30d (p = 0.001), respectively in groups I and II. Duration of iNO administration was shorter in group I (3.3d) than in II (5d), p = 0.05. Preoperative PAPs did not differ between the 2 groups. No significant side effect occurred. Overall mortality was 4% and 4 patients needed long term sildenafil therapy. At latest evaluation, 38% were in NYHA class I, 40% in NYHA II and 22 in NYHA III.
Conclusion: Sildenafil is safe and reliable in the postoperative course after surgery for CHD in children and efficacy is optimal if administered <H24 after bypass.

P-225
ALCAPA long term follow-up and prognosis
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The aim of the study was to assess the long term outcome of patients with ALCAPA
Methods: This study is a retrospective single-center analysis of patients who underwent surgery for ALCAPA from 1980 to 2012. Surgical techniques, demographics, echocardiographic parameters and outcomes were collected. Patients were divided into group I < 2 years at diagnosis, group II: >2 years).
Results: 48 patients (28 females) were included, median age 6months (min 4mos, max 65y), median weight 6 kg (min 1.9 kg): 36 in group I and 12 in II. Symptoms of HF were patent in 39 patients (mean age 4y), 9 were asymptomatic (mean age 10y) and Qwave on ECG was present in 87% of cases. Mean LVSF = 24.2% (group I = 26.6% vs II = 37.7%, and 16.6% in patients <6 mos of age), LVEDD and LVESD Z-scores were respectively +2 and +5 in groups I and II; 73% had MR: severe in 6%, moderate in 46%, mild in 21%. Left coronary artery ostium located in the left posterior sinus in 31cases, right posterior sinus in 12 and in right pulmonary branch in 4. Direct coronary artery reimplantation was performed in 71%, Takeuchi technique in 6%, Meyer technique in 20% and LCA ligation in 3%. Mean age at surgery was 29 mos, mean weight 9 kg. Postoperative mechanical circulatory support was required in 3 cases, who had more severe HF, lower LVSF and longer bypass duration. Mean FU was 81mos (6 to 312 mos). Freedom from reoperation was 100% at 1y, 91% at 10y and 88% at 20y. LVSF increased by 20% in the early postoperative course and 36% at late FU. MR improved significantly in most of the cases. Overall mortality was 33% (15 in group I died before post-operative Day-30, none in group II), decreasing over time from 55% to 11%, and was lower in patients who underwent direct reimplantation. Q wave disappeared in 82% of the cases; 93% of the survivors were asymptomatic at latest evaluation.

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Conclusion: ALCAPA patients have overall good long-term survival and outcome. Age >2y at diagnosis and direct implantation are factors of favourable prognosis.

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Haemostasis in newborns with congenital cyanotic heart defects

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Basics: to study haemostasis in newborns with critical cyanotic heart defects
Methods: 21 newborn with critical cyanotic heart defects and mean blood oxygen saturation pre-operatively 51.8%. The subclavian-pulmonary artery anastomosis surgery was carried out urgently on 2 ± 1 day of hospital stay on average. All the patients were clinically investigated, including clotting, full blood count and biochemistry.
Results: pre-operatively we noted that cyanotic newborns have: higher platelets count 12% on average, Prolonged prothrombin index by an average of 19%, prolonged activity of the prothrombin complex by Quick's method by 26.2% on average, Increased INR by 25% on average, Increased fibrinogen level by 194%, Decreased activity of factor X in intrinsic clotting system by 30% in first blood clotting phase, Antithrombin level decreased by 5%, Increased fibrinogen degradation products D-dimer levels by 94.6%. Post-operatively we evaluate: Decreased haemoglobin level by 12% on average, normal range of Erythrocytes, haemoglobin, Platelet levels were incersed by 42% on average, Prothrombin time was normal, Activity of the prothrombin complex by Quick's method was increased by 10.2% on average, Fibrinogen level was 12% higher than normal level for age, INR prolonged by 9. 5% on average, AT level decreased by 5% on average, D-dimer level increased 141.4% of the normal level. These findings allowed to institute appropriate antithrombotic therapy post-operatively. Standard anti-thrombotic therapy with heparin followed by aspirin was carried out. Human antithrombin and fraxiparine were used alongside with standard anti-thrombotic therapy when high D-dimer and low antithrombin levels.
Conclusion: Haemostasis in newborns with critical congenital heart defects can be characterised as immature. Pre-operatively newborns with critical congenital heart defects have: moderate increase in platelet levels, prolonged INR, increased fibrinogen and D-dimer levels. There have been an increase in platelet count, decreased fibrinogen, increased D-dimer and moderately decreased antithrombin level post-operatively. Given high D-dimer level and low antithrombin level it is wise to combine standard antithrombotic therapy with fraxiparine and human antithrombin respectively.

P-227

Influence of bypass time and systemic inflammatory response after cardiopulmonary bypass on the increase of neutrophil gelatinase-associated lipocalin in infants after cardiac surgery

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Introduction: Acute kidney injury (AKI) after cardiopulmonary bypass (CPB) surgery in infants is a frequent complication. Currently the diagnosis of AKI in children is defined by the pediatric RIFLE classification. In the last decade neutrophil gelatinase-associated lipocalin (NGAL) becomes a promising early biomarker for the development of AKI. The aim of this study was to investigate the influence of bypass time and systemic inflammatory response after CPB on the increase of NGAL in infants after cardiac surgery.
Methods: We enrolled consecutively infants undergoing surgery on cardiopulmonary bypass because of congenital heart disease in this prospective study. NGAL in serum and urine and interleukin6 (IL6) were checked before CPB, 2–4 h after CPB and on postoperative day 1 (POD 1). AKI was defined by the pRIFLE classification. We compared NGAL, IL 6 and bypass time between infants with and without AKI.
Results: In the 59 infants the median NGAL in urine and plasma and IL 6 increased significantly from before CPB to 2-4 h after CPB (NGAL in plasma: 35 vs. 41 ng/ml, $p = 0.05$; NGAL in urine: 2.4 vs. 25 ng/ml, $p > 0{,}0001$; IL 6: 2.2 vs. 89 pg/ml, $p < 0.0001$). After the pRIFLE classification 27 infants developed AKI, 32 don't. The median bypass time between infants with and without AKI was comparable (75 min vs. 77 min, $p = 0.9$). There was no significant difference in NGAL in urine or plasma or IL 6 between infants with and without AKI to any time point. 27 infants have bypass times below 75 min and 32 infants ⩾75 min. The infants with a bypass time more than 75 min showed significant higher NGAL values in urine 2–4 h after CPB and on POD 1 as infants with bypass times below 75 min (42 vs.9 ng/ml, $p = 0.01$ and 17 vs. 2 ng/ml, $p = 0.01$).
Conclusion: In our study NGAL is depending on bypass time and systemic inflammatory response after CPB and couldn't be used as reliable early biomarker for the development of AKI after surgery on CPB in infants.

P-228

Pediatric Open Heart Surgery in Emerging Countries: A 10-year Experience at the Maputo Heart Institute, Mozambique

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Background: The Chain of hope acts since 1988 to free operate African, Asian and Middle East children with heart diseases. This study aimed to assess the feasibility and results of a 10-year open-heart surgery program at the Maputo Heart Institute, Mozambique.
Methods and Results: Data of all consecutive patients with history of at least one heart surgery performed at the Maputo Heart Institute between Jan. 2001 and Dec. 2011 were opening were analyzed, in regard peri-operative (in-hospital) as well as long-term follow-up. Overall, 891 operations were performed in 776 patients, including 93% performed exclusively by one of the

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7 international teams and 7% by Mozambican surgeons alone. The mean follow-up was 2.7 +/- 2.8 years (1 month–11 years). The mean age of patients was 15 +/- 16 years, with almost one third of patients living at more than 1000 km from the heart institute. In 86%, the Maputo Heart Institute offered surgery for free, whereas 14% paid specific price (adults with comfortable incomes). Principal indications of surgery was congenital heart disease in 47% (14.1% of ventricular septal defect, 13.4% of tetralogies of Fallot, 5.8% of atrial septal defect, 5.3% of ductus arteriosus and 7.4% of more complex congenital heart diseases), rheumatic heart disease in 33%, endomycardial fibrosis in 8%. Mean hospital stay was 8 days +/- 12 days. Peri-operative mortality rate was 5.9%, with significant difference between indications: 6.6% for congenital heart diseases, 2.3% for RHD and 6.8% for EMF (P = 0.02). After hospital discharge (do not consider here those who died in hospital), the mortality rate at one year was 2.1%. Among patients with mechanical prostheses, the mortality associated with the prosthesis was high (18.3%). More than half of patients were lost of follow-up at 3-year, and prevention of rheumatic heart disease non optimal for many children…

Conclusion: Our findings suggest the feasibility of such a program, with development of local competences and finally the set-up of an independent surgical team with effective intensive care unit. However, our results also emphasize some weaknesses, especially the issue of follow-up of patients who would benefited the most of long-term preventive measures.

P-229

ASO for TBA: Are the results any different from TGA & Large VSD

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Objective: An arterial switch operation (ASO) is considered the treatment of choice for double-outlet right ventricle with subpulmonic ventricular septal defect (VSD) and also TGA & VSD. The clinical results of an ASO with VSD closure for this Taussig—Bing anomaly (TBA) were retrospectively studied and compared with the group of patients with TGA & large VSD undergoing ASO in the same time period.

Methods: From January 2008 through June 2013, 30 patients with TBA and 54 of TGA with large VSD underwent ASO at the Escorts Heart Institute New Delhi.

Median age at surgery for TBA 3 months; and for TGA with VSD it was 2 months. Weight of the patients for TBA was 4.1 ± 2.6 kg and TGA VSD group was 4.4 ± 2.5 kg.Great arteries relationship was side by side in 30%, 20%; Non type1 coronary arteries arrangement was present in 23%, 13%; associate aortic arch obstruction (AAO) was33%, 11%; cleft in AMLwas17%, 3% respectively. 46.7% patients in TBA and 30% patients in TGA presented with sepsis secondary to pneumonitis. 13% of TBA and 5.5% of TGA patients were on intermittent positive pressure ventilation (IPPV) which continued up to surgery.

Results: Anatomical & surgical complexity was higher in the TBA group due to statistical significant higher incidence of side by side great arteries (p0.0001); AAO (p0.01); MR (p0.03). Despite this difference in complexity the mortality was not different between the groups. The only factors in both groups contribute to mortality was presence of preoperative sepsis (p0.019) and preoperative mechanical ventilation (p0.001). Freedom from reoperation was 100% at 5 years. Hospital mortality was 23.3% (7 of 30) for TBA and 11.1% (6 of 54) for TGA with Large VSD. Among the patient who had no preoperative sepsis and IPPV the mortality was 0%. Actuarial survival was 85% at 5 years. Follow-up was 92% completed from July to November 2013 with a mean follow-up 4.1years (range 6 months to 5.9 years).

Conclusions: In our experience preoperative conditions (recent sepsis/IPPV) of this patient is the only determinant factor for mortality and morbidity but not the underlying disease per se.

P-230

Surgical management after proceeding balloon aortic valvotomy for neonates with critical aortic stenosis

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Objectives:: The optimal management strategy for neonates with congenital aortic stenosis and duct dependent systemic circulation (critical aortic stenosis) is still questionable. Since 1996, we have treated all patients with balloon aortic valvotomy (BAV) at first, then subsequent surgical management was planned.

Methods: Thirteen patients were enrolled between 1996 and 2013. BAV was performed at the median age of 1 day old (range, 0-28). For patients with maintained left ventricular (LV) systolic function (n = 7), the definitive Ross or Konno-aortic valve replacement (AVR) was scheduled after infantile period. For patients with reduced LV ejection fraction of less than 30% and/or endocardial fibroelastosis (n = 6), palliative surgery aiming at biventricular repair was subsequently planned (n = 4), or since 2010, bilateral pulmonary artery banding (bPAB) with ductal stenting was conducted as bridge to decision for further treatment (n = 2).

Results: Follow-up was completed on all patients and median follow-up period was 3.5 years (max, 16.0). The overall survival rate at 15 years was 66.1%. Six of 7 patients with maintained LV systolic function could reach the definitive Ross or Konno-aortic valve replacement (AVR) at the median duration of 311 days after initial BAV without any mortality. Of patients with reduced LV systolic function, one died before subsequent surgical palliation by progressed LV dysfunction. Three patients underwent palliative surgery, which resulted in only 1 survivor. After 2010, one patient ultimately underwent Fontan completion at 38 months of age following bidirectional Glenn concomitantly with Norwood type arch reconstruction at 22 months of age. The other successfully underwent Ross-Konno operation at 9 months of age after the recovery of LV systolic function.

Conclusions: The proceeding BAV could provide elective Ross or Konno-AVR for patients with maintained LV function. Although the statistically significant improvement has not been observed yet, the application of bPAB and ductal stenting following BAV would be a favorable alternative for patients with reduced LV systolic function, to avoid high risk neonatal Ross or Norwood type operation, and also to determine further treatment carefully.

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Perioperative assessment of left ventricular function by 2D strain (speckle tracking) in pediatric cardiac surgery

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Introduction: Cardiopulmonary bypass can be responsible for post-operative left ventricular dysfunction. Conventional echocardiography

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parameters, such as left ventricular ejection fraction measured by Teicholz or Simpson methods, are neither accurate nor reproducible enough during this critical period. 2D strain by speckle tracking could be interesting in this context. The main objective of the study was to estimate the feasibility and reproducibility of 2D strain throughout the peri-operative period. The second objective was to assess the post-operative evolution of 2D strain values after surgical closure of atrial septal defect.
Methods: 34 pediatric patients (<18 years) with congenital heart disease undergoing cardiac surgery with cardiopulmonary bypass were included in this prospective single center study. Daily echocardiography was performed from the day before the surgery until the fifth post-operative day. Left ventricular ejection fraction (Teicholz and Simpson methods) and 2D strain values (longitudinal, circumferential and radial) were measured.
Results: The mean age was 5 years [3,27-6,60] and the mean weight was 16 kg [11,56-21,28]. The mean cardiopulmonary bypass and aortic cross-clamping durations were respectively 109.6 minutes [82.9-136.3] and 54.3 minutes [40.5-68.1]. The global post-operative feasibility of longitudinal, circumferential and radial 2D strain was respectively 93%, 95% and 95% and was similar to the one of conventional parameters (97%). The intra-observer correlation coefficient of longitudinal, circumferential and radial strains were respectively 0.916 ($p < 0.001$), 0.880 ($p < 0.001$) and 0.701 ($p = 0.002$). The inter-observer correlation coefficient of longitudinal, circumferential and radial strains were respectively 0.885 ($p < 0.001$), 0.829 ($p < 0.001$) and 0.559 ($p = 0.020$). In the cohort, 13 patients had surgical closure of atrial septal defect. Circumferential 2D strain was significantly improved for these patients (-15.07 [-17.37;-12.76] versus -19.86 [-22.73;-17.00], $p = 0.028$).
Conclusions: 2D strain is feasible and reproducible for the assessment of left ventricular function after surgery for congenital heart disease. The reproducibility of this method is higher than the one of conventional parameters. Circumferential 2D strain significantly improves after atrial septal defect surgical closure. Further studies are needed to assess the clinical interest of this technique, especially for early diagnosis of post-operative myocardial dysfunction, and to determine the expected evolution of 2D strain values for each congenital heart disease.

P-232
Analysis of Long Term results for RVOT Homograft Reconstruction

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Introduction: Right Ventricle Outflow Tract (RVOT) Reconstruction has been successfully performed since the 1950s. There is a lot of debate regarding the best conduit for RVOT reconstruction. RVOT homograft (HMG) replacement is routine in many centers, but very few long term series are available in the literature.
Methods: Under the approval of the Ethics Research Committee, all patients' records were reviewed. Follow-up was obtained by patient's outpatient clinic visits. The main purpose was to evaluate HMG dysfunction (either severe stenosis or regurgitation), need for percutaneous intervention, HMG failure or death related to the procedure. Data analysis was performed in IBM SPSS.
Results: Between May 1995 and June 2013, 182 RVOT HMG reconstructions were performed. 56% of the patients were male. Mean age at surgery was 9 years old. 132 Aortic and 50 pulmonary homografts were used. The most common diagnosis was Fallot's Tetralogy in 79 patients, followed by Pulmonary Atresia and Truncus Arteriosus, with 38 and 14 patients respectively. Mean extracorporeal circulation time (ECT) was 125 minutes. Mean follow-up was 7,6 years (ranging from 6 months to 17 years). 18 patients needed homograft replacement. Freedom from homograft dysfunction in 2, 5 and 10 years was 96,7%, 91,7% and 68,6%, respectively. Freedom from pulmonary or aortic homograft dysfunction, in the same follow-up, was 96,7%, 96,7% and 83,9%, and 95,5%, 89,9% and 62%, respectively. There was no statistical difference in mortality between aortic and pulmonary homograft patients.
Conclusions: Homografts are an excellent choice when RVOT reconstruction is needed. Long time follow-up demonstrated better patency for pulmonary type. Lower age at surgery and higher than 115 minutes ECT where independent risk factors to death ($p < 0.05$). Non-Fallot diagnosis had a stronger tendency to statistical significance for death ($p = 0,056$).

P-233
Surgical Treatment for the Partial Anomalous Pulmonary Venous Connection: 23-Year Experience

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Introduction: Surgical treatment of partial anomalous pulmonary venous connection (PAPVC), although presenting no major problems in the short term outcomes, may lead to serious complications in the medium and long term outcomes. This study aim to evaluate the midterm results of surgical treatment of PAPVC.
Methods: A retrospective single-center study was performed with PAPVC patients operated between December/1988 and January/2011 in our institution. Medical records, surgical reports and results of complementary exams were analyzed. Descriptive statistics used mean ± 2 standard errors and the nonparametric Wilcoxon test for paired samples was used to assess post-surgical outcomes.
Results: Ninety-one patients, with a mean age at surgery of 13.6 ± 3.0 (min = 0, max = 54.3) years and a mean weight of 32.7 ± 5.2 (min = 3, max = 94) kilograms, were operated. The mean follow-up time was 29.2 ± 7.2 (min = 0, max = 107) months. 75.4% of the patients presented right PAPVC, 12.3% left PAPVC and 12.3% mixed type PAPVC. Twenty (22%) patients had other associated diagnoses, besides an atrial septal defect. The surgical technique was tunneling with pericardial patch in 72.5%, direct connection of the pulmonary veins to the left atrium in 17.5% and the Warden technique in 10% of cases. During follow-up, 7 (7.7%) patients underwent other cardiac surgery for correction of associated cardiac malformations. There were 2 (2,2%) late deaths, none of them related to direct complications of surgical treatment of PAPVC. The post-operative control showed decreased right ventricular overload ($p = 0.022$) and dilation ($p = 0.048$), as well as reduced tricuspid insufficiency ($p = 0.036$).
Conclusion: The medium-term results of the surgical treatment for PAPVC were satisfactory. It is necessary now, the long-term monitoring of these patients.

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P-234

Outcome of surgical intervention within 24 hours of life for hypoplastic left heart syndrome with intact or highly restrictive atrial septum

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*Objectives:*The surgical outcome of HLHS has been improving, however HLHS with intact/severe restrictive atrial septum (IAS/RAS) carry a poor prognosis. The purpose of this study is to clarify the surgical outcomes for this cohort.
Methods: We performed a retrospective review of 11 patients (8 female) diagnosed as HLHS and its variant with IAS/RAS who required urgent surgery within 24 hours of life from January 2003 to December 2013 at Mt. Fuji Shizuoka Children's Hospital. One patient who combined supra-cardiac TAPVC was excluded from this study. Norwood (NW) procedure was indicated as a first palliation for this cohort before 2007, however ASD creation with bilateral pulmonary artery banding (BPAB) was introduced after 2007 to obtain a quick and secure effect of left atrial decompression with minimal surgical stress on newborn babies. Follow-up data were available for 100% of the patients.
Results: Five were diagnosed prenatally and had planed delivery in our institute. Other 6 were born in other hospital and transferred immediately after their birth. Median gestational age and birth-weight was 38 (range, 36-41) weeks, 2.41 (1.71-3.02) kg, respectively. All 11 patients required urgent operation 7.5(0.75-20) hrs after their birth. Their SpO2 at OR was 62(43-81)%. As a first palliative surgery, two patients (before 2007) underwent NW procedure and 9 underwent ASD creation with BPAB. After ASD creation with BPAB, prostaglandin-E1 was continued and 4 survived to undergo a NW procedure at a median age of 39(24-54) days. We lost 5 out of 11 in perioperative period of NW procedure due to LOS (2), infection (1), pulmonary emphysema (1), and pneumorrhagia (1). Four accomplished bidirectional Glenn shunt and two of them have already undergone Fontan operation with adequate CVP and SaO2 during the median follow-up period of 0.59 years (7days-10.2years).
Conclusions: HLHS with IAS/RAS is a challenging combination. Our strategy to avoid postpartum NW procedure and to decompress the left atrium as soon as possible after birth by ASD creation with BPAB can bring a better prognosis for this cohort even though prenatally diagnosis is not available.

P-235

Extra-corporeal membrane oxygenation (ECMO) for children after cardiac surgery

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Objective: To review our experience in pediatric patients who supported by extracorporeal membrane oxygenation (ECMO) after cardiac surgery.
Methods: Between Jan. 1998 and Dec. 2013, there were 80 ECMO runs in 73 pediatric patients after cardiac surgery at Mt.Fuji Shizuoka Children's Hospital. Seven patients who required 2nd ECMO support during the same hospitalization were excluded (2 discharged alive, and 5 dead) and 66 patients were included in this study. Retrospective analysis of their clinical course were performed from hospital records.
Results: Median age and median weight was 1.9 month (day 0-21years) and 3.2 (1.6-70.4) kg, respectively. Thirty-three (50%) failed to wean off cardiopulmonary bypass and ECMO support was initiated in operating theater. Other 33 required ECMO support at ICU 34 hrs (27min-14weeks) after the end of operation, because of sudden deterioration requiring cardio-pulmonary resuscitation (CPR) with chest compression (n = 21) and for other reasons (n = 12). The duration of CPR (from the begging of CPR to ECMO run) was 47 (15-104) min. Almost all patients were assisted by V-A ECMO with both atrial and aortic cannulation under opened chest.
Forty-four (67%) had single ventricle physiology and 38 of them underwent palliative surgery. Median duration of ECMO support was 116(1.5-523) hrs and 5 patients required open-heart surgical re-interventions during its support. Overall 48(73%) were successfully weaned off ECMO (alive more than 24hrs after ECMO discontinuation) and 38(58%) were discharged alive. As a major complication, brain damage was found in 5 patients (all of them were resuscitated before ECMO support).
Successful ECMO weaning was not associated with ventricular physiology (UV vs BV, $p = 1.00$), treatment stage (palliation vs correction, $p = 1.00$) and E-CPR (CPR(+) vs CPR(-), $p = 0.770$).
Conclusion: Once the hemodynamics was properly established after cardiac surgery, ECMO can be a strong life-saving tool with few complications even in patients with single ventricle who dropped into circulatory collapse and resuscitation is required. Brain damage remains a major complication in E-CPR patients.

P-236

Hyperuricemia and renal dysfunction after cardiac surgery with extracorporeal circulation in pediatric patients

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Objective: Hyperuricemia and renal dysfunction are common problems after cardiac surgery with extracorporeal circulation (EC), but published data regarding incidence and severity of these complications vary widely. Our study was aimed at evaluating the incidence of significant hyperuricemia, the correlations with risk factors, and the efficacy of treatment with urate oxidase among children undergoing cardiac surgery with EC for various congenital anomalies of the heart.
Design: Retrospective chart review.
Setting: Pediatric patients in a university hospital setting.
Patients: Our review identified 175 consecutive patients aged from one day to 17 years who had cardiac surgery with cardiopulmonary bypass between January 2000 and July 2005.
Measurements and main results: The data collected from the records included demographic and biometric data, diagnosis, surgical risk factors, postoperative serum creatinine, uric acid and urea, and any medication of interest for the study. The mean (±SD) duration of EC and aortic clamping (AC) was 119 ± 51 and 65 ± 33 min, respectively. Serum uric acid, urea and creatinine levels all increased significantly during postoperative days 1-3. Overall, 26.1% of the patients had an increase in their serum uric acid level to 8 mg/dl or more, with no significant correlation with the duration of EC or AC. Only one patient required peritoneal dialysis postoperatively. Thirteen patients (7%) were treated with allopurinol, and six (3%) with urate oxidase (3 with the conventional enzyme, uricozyme, and 3 with the recombinant preparation, rasburicase). Serum uric acid levels fell promptly within hours in all patients treated with uric oxidase and reached subnormal concentrations within 24 hours. No adverse effects were observed.
Conclusion: These data suggest that urate elevations are common in the postoperative course after cardiac surgery and may reach

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significant levels. Urate oxidase may be effective and safe for the treatment of postoperative hyperuricemia in pediatric patients.

P-237

Long-term follow-up of repaired Tetralogy of Fallot using small infundibulectomy in developing countries children

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Objectives: There are still controversies regarding the surgical approach and the optimal age for surgery in Tetralogy of Fallot. In the developed world it's common to operate earlier as a one stage procedure. The definitive repair in infancy have been reported to carry low mortality and morbidity. However, in developing countries staged repair is still practised due to various limitations. The aim was to analyze the early post-operative clinical course of the RVOT obstruction enlargment, using small infundibulectomy.
Methods: 46 consecutive operated-Tetralogy of Fallot, underwent complete correction, were clinically and instrumental evaluated. The resection of RVOT was done by excising parietal band, dividing all obstructing muscle bands and excising the fibrous tissue, through a small infundibulectomy. The RVOT and the pulmonary artery were closed with autologous pericardial patch whenever required.
Median age at correction was 4.8 years.
Bi-ventricular volumes and ejection fraction, pulmonary regurgitation fraction, infundibular anatomy and function, pulmonary artery anatomy and flow, functional capacity and adverse outcomes were evaluated.
Results: RVOT reconstruction technique, transannular patch, right ventriculotomy size, duration of postoperative mechanical ventilator support and pumonary valve regurgitation grade by immediate postoperative echocardiography were significant risk factors of right ventricular dilatation.
Aggressive ventriculotomy predispose to RVOT aneurysms or akinetic regions.
Of the various RVOT reconstruction techniques, pulmonary valve repair through pulmonary arteriotomy and small infundibulectomy had the lowest incidence of right ventricular dilatation.
RV end-diastolic volume was low (146 ± 37 ml/m^2), pulmonary regurgitation fraction was low ($36 \pm 13\%$), ejection fraction of the infundibulum was high ($28 \pm 11\%$).
Conclusions: Our data show that early and late right ventricular dilatation after total repair in Tetralogy of Fallot is low, using small infundibulectomy.

P-238

Midterm Follow Up after Bi-ventricular Repair of the Hypoplastic Left Heart Complex

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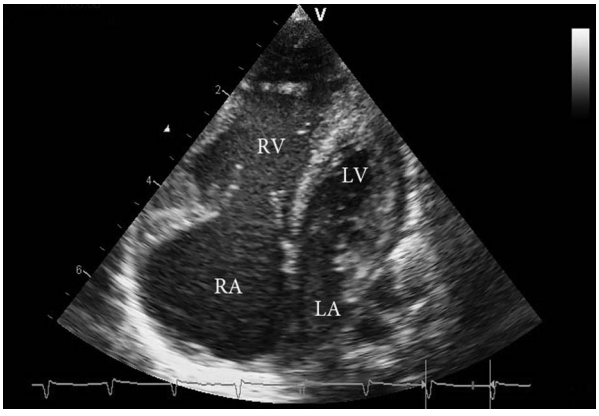
Introduction: In neonates with the hypoplastic left heart complex (HLHC), as defined by Tchervenkov Cl 1998, biventricular repair is considered superior to univentricular repair. The Z-scores of the mitral valve (MV)- and the aortic valvue (AoV)-annulus are primary factors for the choice of repair. Yet, predictive cutoff values for the feasibility and optimal outcome of biventricular repair are unknown. This study assesses the midterm outcome and the growth of the left heart structures after neonatal biventricular repair, and tries to evaluate a cutoff value for the MV- and AoV-annulus Z-scores.
Methods: HLHC-patients who underwent biventricular repair at our institution between 2004 and 2013 were reviewed retrospectively. MV-annulus, AoV-annulus, left ventricle inlet length and left ventricular internal diastolic dimension were measured by echocardiography before and at 6, 12, 24 & 48 months after biventricular repair to calculate the Z-scores. Using t-testing, the change in Z-scores before and after operation, and the influence of preoperative Z-scores on the need of re-interventions was assessed. The Z-score calculation method of Pettersen et al 2008 was used.



Results: Nineteen patients were included. The follow-up ranged from two to 98 months. The 30-day mortality was zero. The midterm survival rate was 95% and 85% of the patients were classified as NYHA I. One patient died due to a non-cardiac related cause after two months. Seven patients (37%) required a total of 8 re-interventions, due to recurring or residual obstructive lesions (4 re-CoA, 2 supravalvular AS and 2 subvalvular AS). Within six months after biventricular repair, the Z-scores almost normalized but remained small thereafter ($p < 0.001$).
Conclusion: Neonatal biventricular repair is successful even in HLHC patients with AoV- and MV-annulus Z-scores equal to -5.8 and -4.73 respectively. It can be concluded that the left heart structures undergo temporary compensatory growth after neonatal biventricular repair as within the first six months after repair the Z-scores almost normalize. Nevertheless re-appearance of stenotic lesions requires re-intervention in 37% of the HLHC population. However the re-appearance of stenotic lesions was not significantly correlated to the magnitude of the pre-operative MV- and AoV-annulus Z-scores.

P-239

Surgical repair of ventricular septal defect; contemporary results and risk factors for a complicated course

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Introduction: The ventricular septal defect (VSD) is the most common congenital malformation of the heart and therefore surgical closure of a VSD is the most common pediatric open heart surgery. This surgical procedure is relatively safe but there are studies that suggest a higher risk at complications with low weight and young age. The purpose of this study was to see if there was any correlation between young age/low weight and complications and to evaluate the safety of this procedure.

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Methods: This retrospective study examined a consecutive series of 243 VSD closures at a single institution. Because mortality is low in nearly all centers for repair of these defects, we focused on morbidity and identified drivers of risk via multivariable linear regression modeling.
Results: 243 patients who underwent surgical closure of a VSD were included. The median age at operation was 168.0 days (range 17–6898), the median weight at operation was 6000 grams (range 2100–102000). No deaths occurred. Only two patients (0.8%) had a permanent heart block requiring pacing. In total only seven patients (2.9%) of the patients had a major adverse event. A lower weight at operation resulted in a longer stay and a longer ventilation time, but not did not increase the risk of complications or major adverse events.
Conclusions: Contemporary results of a surgical VSD closure are excellent. The procedure is safe with major adverse events occurring in only 2.9%. Low weight at operation is not associated with an increased risk of complications or major adverse events but is associated with increased ventilation time and a longer hospital stay. Therefore surgical VSD closure is a safe procedure in all patients regardless of their weight.

P-240
Contemporary results of Coarctation repair: Can we do better?
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Introduction: Aortic coarctation occurs as an isolated anomaly or in association with other complex malformations. Resection of isthmus and extended end-to-end anastomosis has become the gold standard for surgical repair of coarctation.
Methods: We reviewed 100 consecutive primary coarctation repairs performed since 2003 at our clinic. Coarctation co-existing with complex intra-cardiac defects such as d-TGA, Shone's complex and others (10 out of 100) were excluded. Median age and weight were 32(1-1878) days and 4(1.5-20) kg. Coexisting defects included bicuspid aortic valve (20), arch hypoplasia (34), ASD/VSD (9). In 7 patients with significant LV dysfunction at time of diagnosis temporary stenting of the isthmus followed by early stent removal and resection was performed as soon as the LV function recovered. Muscle sparing posterior thoracotomy was the approach used. Three patients underwent arch augmentation using subclavian/left common carotid flap and 3 using autologous pericardial/xenopericardial patch. Eight patients with VSD underwent concomitant PA banding. Median ischemic aortic clamp time was 25 (15-43) min. Follow-up was complete with a median duration of 38 (2-80) months.
Results: The hospital mortality was 0%. Two patients with a well corrected aortic arch and isthmus and a corrected AVSD died after 20.5 and 4.5 months due to pulmonary hypertensive crisis and unknown cause, respectively. Incidence of paraplegia/paresis was 0%. The median hospital stay was 11 (2-147) days – few having been discharged to the referring hospital. The freedom from reoperation and reoperation/reintervention on the arch and isthmus was 90(+10, -18.6) % and 87.6(± 6.6) % at 60 months respectively. Seventeen patients till date have undergone subsequent intra-cardiac repairs for ASD, VSD, AVSD, sub-aortic membrane, and mitral valve repair/replacement.
Conclusion: Surgery for aortic coarctation (isolated and with simple intra-cardiac shunts) can be performed with incrementally low operative risk. The rate of reoperation and re-intervention is low. Most recoarctations can be managed with catheter intervention. Further studies will help develop better selection strategies so as to further reduce the need for reoperation by subjecting them to a more aggressive repair using cardiopulmonary bypass.

P-241
Does the hypoplastic arch grow after resection of Coarctation and extended end-to-end anastomosis?
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Introduction: Arch hypoplasia coexists in about 40% of children presenting with Aortic Coarctation. Resection and extended end-to-end anastomosis is an elegant operation to repair Coarctation without subjecting a neonate to the risks of cardiopulmonary bypass (CPB). We sought to analyze the long-term growth of the arch after such an operation.
Methods: 29 patients with hypoplastic arch and Coarctation undergoing resection and extended end-to-end anastomosis without CPB (out of a total of 139 consecutive Coarctation repairs performed between 2002-2010) were reviewed. Median age and weight were 7 (range 0-442) days and 3.1 (0.98-10) kg respectively. Z-value of the transverse arch preoperatively was a Median of -4.6(-9.4 to -1.4). Median follow-up (Fup) was 82.3 (28.1-119.7) months.
Results: There was no early or late death. 2/29 (7%) patients needed surgical re-enlargement of the arch through a sternotomy at 6 & 14 months and therefore were excluded from further Fup. 1/27 patient required balloon dilatation for recoarctation at 2.3 months and was retained in the study. Z-value of the transverse arch improved from a median of -3.5 (-7.6 to -0.5) early postoperatively to -2.6 (-7.3 to -0.6) at follow-up (Growth spurt from 3.1 to 19.5[9-44] kg). All (barring 1) patients were free of antihypertensive medication. 3 patients had signs of LV hypertrophy, often due to association with bicuspid aortic valve, sub- aortic stenosis and AV septal defect.
Conclusion: The hypoplastic arch grows with age after extended end-to-end anastomosis, but some arches remain small for age. Further studies are needed to identify the morphological subgroup with small proximal arch diameter that may be best corrected through a sternotomy and use of CPB.

P-242
Dobutamine stress echocardiography in children after anatomical correction of transposition of the great arteries - this method is safe or not?
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Objectives: Anatomic correction of transposition of the great arteries is associated with risk of coronary complications in the postoperative period. Dobutamine stress echocardiography is non-invasive method for detection of coronary abnormalities after arterial switch operation. The aim of the study was to evaluate the safety dobutamine stress echocardiography in children after anatomical correction of TGA.
The study group consisted of 76 patients after anatomical correction of transposition of great arteries in the neonatal period (89% of patients). The age of patients ranged from 3 years to 16 years (mean 8 years). There were 51 (67%) boys and 25 (33%)

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girls. In the group of 53 children with simple transposition of the great arteries, in 23 cases, it was a complex transposition (with ventricular septal defect or pathology of the aortic arch).
Methods: All patients underwent dobutamine stress echocardiography to assess left ventricular function compared with the result of coronary angiography. The test was performed according to the approved protocol (with atropine in selected cases). We analyzed the course of the test and the frequency and type of side effects associated with the administration of dobutamine.
Results: The negative result of dobutamine stress echocardiography (without induced wall motion abnormalities) was in 54 patients, positive in 15 (20%) and non-diagnositc in 3 (4%). The test interrupted because severe discomfort or arrhythmia in 4 cases (5%).
Twenty four (32%) of the 76 patients developed side effects during dobutamine infusion that included headache (5), abdominal pain (4) with or without nausea and vomiting (2), nonspecific chest pain (2) and arrhythmia (11). These side effects were transient and resolved spontaneously with discontinuation of the infusion. Gender and age of the patients, type of the TGA, abnormalities of the coronary arteries and atropine-protocol did no affect the incidence of side effects during the test (NS). Arrhythmia occured more frequently in patients with complex type of the TGA ($p = 0.02$) and non-negative test result ($p = 0.02$).
Conclusions: Dobutamine stress echocardiography is a safe diagnostic method in children.
Side effects during dobutamine infusion are transient and reversible without treatment.

P-243
Diagnostic utility of rigid body rotation in children with left ventricular non-compaction: a two-dimensional speckle tracking study

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Background: The reproducibility of accepted echocardiographic criteria of left ventricular non-compaction (LVNC) is poor. 'Rigid body rotation' (RBR) or reversed LV rotation has been suggested as a novel diagnostic parameter in adults. However, no study has examined RBR in children with LVNC. We sought to explore the prevalence of LV rotational abnormalities in children with LVNC, and the feasibility of assessing it.
Methods: We searched our echocardiographic database to identify all children who fulfilled the diagnostic criteria for LVNC between January 2010 and December 2013. All subjects underwent a normal conventional echocardiographic study. LV Short-axis views at the base and apex were obtained. LV rotation (LVrot) was assessed using two-dimensional speckle tracking echocardiography. Twenty healthy subjects served as controls.
Results: We identified 13 children with LVNC. Three patients were excluded because of poor image quality (feasibility: 10/13 patients (77%)). The study comprised 10 children with LVNC. Associated congenital heart disease was found in 7/10 patients (70%). Mean age was 6.1 years (range 1 month – 16 yrs). LV ejection fraction was reduced in 4/10 children. LVrot was abnormal in all patients (100% vs 0%, $P < 0.001$). LVrot was decreased in 2 patients, and augmented in 1 patient. A RBR pattern was noted in 7/10 patients (70%; reversed apical-rot $n = 4$, reversed basal-rot $n = 3$). The patient with augmented LVrot developed a RBR pattern during follow-up.
Conclusions: Feasibility of LVrot assessment in children with LVNC was good. A typical RBR pattern was found in most children with LVNC. However, a RBR pattern may develop during follow-up. This novel echocardiographic feature may have diagnostic implications.

P-244
Takotsubo cardiomyopathy in the very young

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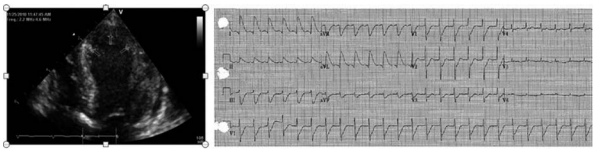
Introduction: Takotsubo cardiomyopathy, apical ballooning or "broken-heart" syndrome is characterized by a transient severe dysfunction of the apical segments of the left ventricle affecting the distribution of more than one coronary artery in the absence of angiographic evidence of obstructive coronary artery disease. It is associated with ST changes and T wave inversion, mild elevation of cardiac enzymes, and a sudden emotional and/or physical stress as the precipitating factor. The majority of the reports have been in adults, especially in postmenopausal women. It has rarely been described in children.



Methods: Case series of two cases of Takotsubo cardiomyopathy presenting in very young patients.
Results: We present two young patients of less than 5 years old who developed features of Takotsubo cardiomyopathy that recovered spontaneously. First case (16 months) presented with cyclical vomiting and the second case (2 years) with head trauma and a right subdural hematoma. Both patient presents with clinical features of heart failure, ECG changes of ischemia, cardiac enzyme elevation and echocardiographic evidence of apical hypokinesia. Both patients were treated with conventional cardiac failure support and had complete recovery of their left ventricular function with normalization of their electrocardiogram, echocardiogram and cardiac enzymes within days. A follow up evaluation a few months later confirmed resolution of this condition.
Figure 1. Echocardiogram and ECG at presentation (case 2)
Conclusions: Takotsubo or stress-induced cardiomyopathy may occur in children, and it should be considered in any individual at any age presenting with the features mentioned above. Special consideration should be given when there is a known significant emotional/physical event that can provoke stress and increase in catecholamine load.

P-245
Outcome of parvovirus B19 myocarditis in children

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Background: The advent of polymerase chain reaction (PCR) testing for the presence of viral genomes has led to identification of parvovirus B19 (PVB19) as a cause of viral myocarditis. This is the first reported series of parvovirus myocarditis in children in the United Kingdom.
Objective: To report the presentation, clinical course and outcome of PVB19 myocarditis in children.

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Methods: Data was collected through a retrospective review. Patients were referred with overt clinical signs of cardiac failure. Echocardiography confirmed impaired systolic left ventricular function. Together with patient history and clinical phenotype of a viral infection myocarditis was diagnosed. PVB19 viral genome was detected by PCR from serum prior to administration of intravenous immunoglobulin. Endo-myocardial biopsy was only performed in patients who required extra-corporeal membrane oxygenation (ECMO) or at the time of post-mortem. Samples were tested for PVB19 viral genome and assessed histologically to identify lymphocytic infiltration. Primary end point was death or transplantation.
Results: 17 (4 females, 7 males) patients presented at median age of 1.3 years (0.4-15.4 years) in cardiac failure with fractional shortening 15% ± 3%. Eleven patients required mechanical ventilation and intravenous inotropes. Intravenous immunoglobulin was administered in 9 patients. Seven required extra-corporeal mechanical oxygenation. All patients (n = 3) with one days' prodrome died. All patients (n = 4) with ST segment elevation died. Event free survival occurred in 65%. Five (29%) patients died, 1 underwent heart transplantation. Four patients died within 6 days of admission. There was a late, sudden death at 8 weeks in a patient with resistant ventricular tachycardia with persistence of PVB19 genome in serum.
Complete recovery of cardiac function occurred within median of 12 months (range: 1–48) in 45% patients (5/11). 6/11 patients were asymptomatic at follow-up. Five patients had echocardiographic normalisation of cardiac function on angiotensin-converting enzyme inhibitor and/or carvedilol and 1 patient had mild impairment of function on medications at 7 years.
Conclusion: PVB19 can cause a devastating viral myocarditis in children. If they can be fully supported through the illness, recovery of the myocardium can occur without the need for cardiac transplantation, even those who develop overwhelming circulatory failure.

P-246

Features and outcomes of Acute Myocarditis in children

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This study was to assess features and outcomes of children with acute myocarditis.
Methods: Patients <18y with acute myocarditis (proved by virology and/or MRI and/ or complete recovery of myocardial function) were included. Clinical data, echocardiographic parameters and outcomes were collected and cases divided in groups I (<2y), II (2 to 10y) and III (>10y).
Results: 72 patients were included (1983 to 2012), 30males, aged 4.1 ± 5.1y (med 1.5y): 43 in group I, 17 in II and 12 in III. Heart failure was present at onset in 57cases (78%): 8 cardiogenic shock (12%), 30 severeHF (44%) were more frequent in groups I (56%) and II (46%) than in III (17%, $p < 0.0001$), while chest pain (15.5% of all) was more frequent in III (83%). LVSF at diagnosis was 18.4 ± 9% (med16%): 16% and 15% in groups I and II vs 30.5% in III ($p = 0.0001$). Aortic VTI was 11.4 ± 5.8 cm (med10): 8 cm and 11 in groups I and II vs 17 in group III ($p < 0.05$). Mitral regurgitation was present in 76.5%, pericarditis in 16.4%, thromboembolic events occurred in 5cases (7%), arrhythmias in 7(10%). Virus was positive in 27cases = 37.5% (1 virus in 24, >1 viruse in 3). Nine patients died (13%) within 2months post-diagnosis (2days to 8.6months), 1 was transplanted (3rd month), 19 have sequellae (27.5%), 40 completely recovered (58%), at FU = 5.5 ± 5.6y (med 4y). Inotrope support was needed in 34cases (47%): 51%, 59% and 16% of groups I, II and III respectively ($p < 0.0001$). Six patients (8.3%, 1 in groupIII) needed mechanical circulatory support (3ECMO, 3VAD), within day-14 from onset: 1 died on support, 5 were weaned-off (duration of support: 4d to 3mos). Survival was 96%, 90%, 87.5% and 86% at 1month, 3months, 6months, 2y and 10y of FU. All patients in group III survived. Ten-year survival was 81.4% in group I and 88.2% in II (p = NS). LVSF improved from 18.4 ± 8.9% (med16%) at onset, to 24.6 ± 10.3% (med23.5%) at 1stmonth, 26.5 ± 8.6% (med26.5%) at 3rdmonth, 30.7 ± 8.6% (med29.6%) at 6thmonth, 38 ± 7% (med37%) at last FU.
Conclusion: Acute myocarditis in children has favourable outcomes despite early mortality. Myocardial dysfunction and heart failure are less frequent in patients >10 years of age. Mechanical circulatory support successfully lessens mortality. Myocardial contractility can progressively improve within the first 6months after onset of disease.

P-247

Left ventricular pump function in Duchenne muscular dystrophy

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Introduction: Patients with muscular dystrophy develop a left ventricular dysfunction during life. Our impression was that left ventricular dysfunction starts during puberty, with an initial increase of the left ventricular diastolic diameter, which was smaller than expected for the body length. Whereas other echo dimension seemed within the normal range. We analyzed our data base to verify our impression.
Method: We had 102 patients with muscular dystrophy assessed echocardiographicaly. A total of 150 examinations were done with an age range from 3,8 to 21,5 years with a median of 11,2 years (mean 11,5 +/- 3,7 years). We measured the aortic, pulmonary valve diameter in 2 d-echo and left ventricular diameters and function in the classical m-mode. The analysis was done using prizm 6.0.
Results: The left ventricular pump function started to decline after the age of 12 years. In some patients being on beat-blockers and ACE-inhibitors it remained reduced with a fs of about 25%, in others it declined further, in some patients close to end of life situation the medication had to be discontinued due to severe hypotension. Interestingly there was a linear growth of the left ventricular end-diastolic and end-systolic diameter of 1 with a set off of 12 mm. The end-diastolic diameter was normal for body length. The aortic root grew as expected with the body length.
Discussion: To our surprise were the left ventricular diameter within the normal range throughout the observation period. The declining pumpfunction seems to be due to a linear increase of the left ventricular end-systolic diameter; which showed a linear growth with the end-diastolic diameter. The end-diastoic

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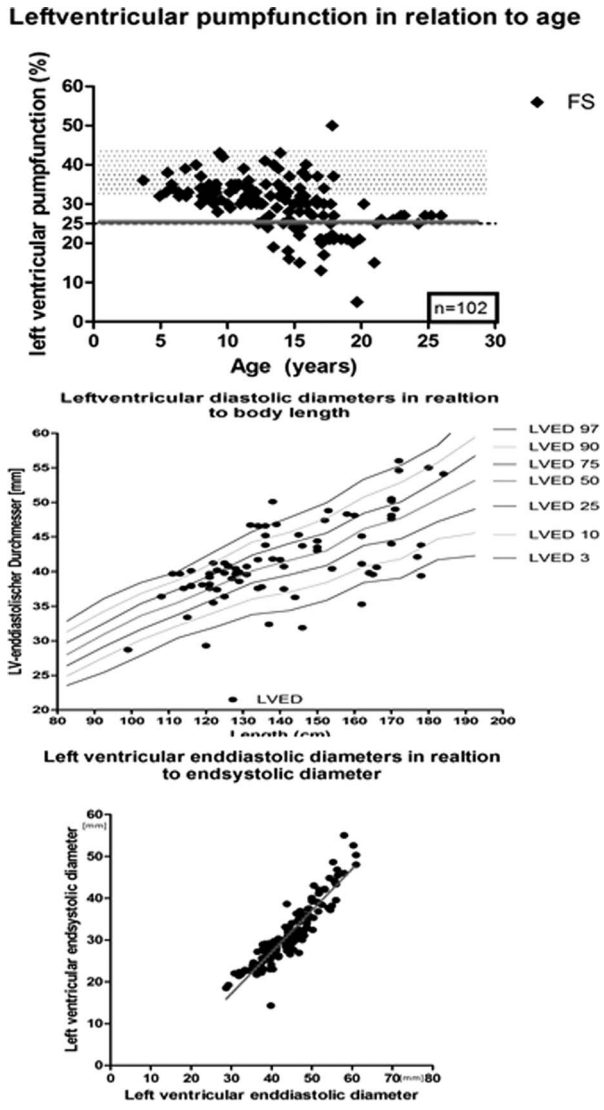
diameter stayed within the normal range, so not comparable to a classic dilating cardiomyopathy were all diameter are out of the normal range.



P-248

Hyponatremia at discharge independently predicts re-hospitalization in adult heart failure patients with congenital heart disease

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Introduction: Hyponatremia is a powerful predictor of adverse outcome in patients with acquired heart disease. However, a little information is available regarding association between hyponatremia and outcome in adult patients with congenital heart disease (ACHD). Therefore we investigated the relationship plasma sodium concentration (Na) and re-hospitalization in heart failure patients with heart failure.
Method: A total 85 ACHD patients who had unexpectedly hospitalized due to cardiovascular events were retrospectively reviewed. Relationship between Na at discharge and cardiac events (death of any cause or unexpected hospitalization related to cardiovascular event after discharge) was evaluated.
Results: The mean age was 28.7 years, 56% were male, 29% had single ventricular physiology and 16% had cyanosis. The mean Na was 138.7 ± 2.5 (mEq/L). The Na was associated with New York Heart Association functional (NYHA) class and use of diuretics. During a mean follow up of 3.3 ± 2.7 years, 51 patients (61%) had cardiac events. Na was significantly lower in ACHD with cardiac events (138.2 ± 0.35 vs. 139.3 ± 0.44 mEq/L, $P = 0.048$). Of the significant predictors of cardiac events on univariate analysis (age, gender, NYHA class, B-type natriuretic peptide, use of diureics), low Na, as well as NYHA class and use of diuretics, was the independently associated with re-hospitalization due to cardiac events (hazard ratio: 0.89, 95% confidence interval 0.74-0.95, $P = .049$).
Conclusion: Hyponatremia was an independent predictor of re-hospitalization in ACHD patients with heart failure.

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Acute vasodilator testing with 100% oxygen can predict adverse outcomes in children with idiopathic and heritable pulmonary arterial hypertension

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Objectives: Acute pulmonary vasodilator testing (AVT) appears to be important for children with idiopathic pulmonary arterial hypertension (IPAH) to identify those who have a favorable outcome. Although several different vasodilators including intravenous epoprostenol and inhaled nitric oxide have been used, the pulmonary vasoractivity assessed by 100% oxygen has not been well established. The aim of this study was to the prognosis value during AVT with 100% of oxygen in children with IPAH and heritable PAH (HPAH).
Methods: A retrospective study was designed to evaluate the in 50 children (younger than 19 years of age) with IPAH/HPAH. A positive response to the AVT is defined as a reduction of pulmonary vascular resistance index (PVRI) ⩾20%, unchanged or increased cardiac index (CI), and decrease or no change in the ratio of pulmonary vascular resistance to systemic vascular resistance (Rp/Rs ratio). The adverse events included hospitalization due to heart failure, lung transplantation, and cardiac mortality.
Results: The median age was 15 years with 22 males and 28 females. Twenty-two children (44%) were acute vasodilator responders. All children with HPAH were non responders. Although responders had lower brain natriuretic peptide levels (20.4, 4.5-203 pg/ml vs 54.8, 7.6-244.9 pg/ml, $p < 0.05$), hemodynamics at baseline including mean pulmonary arterial pressure, PVRI, CI, and Rp/Rs ratio were not significant difference between responders and non-responders. During follow-up period (median 20 months), 27 (54%) children had an adverse event. Cumulative event-free survival rate in responders was significantly higher than those in non-responders (2-year event-free survival rate; 87% vs 33%, log-rank test, $p < 0.05$).
Conclusions: Although hemodynamic data at baseline was not significant difference between responders and non-responders, non-responders may have worse outcomes compared with responders. AVT with 100% oxygen is safe, convenient, and useful for predicting adverse outcomes in children with IPAH and HPAH.

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Hostility is associated with cardiovascular reactivity in mental stressors

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Purpose: Exaggerated/diminished reactivity to stress and hostility as a psychological factor have been extensively studied as the cardiovascular risk factors. We aimed to study autonomic reactivity in the interaction with hostility to different mental stressors.
Methods: Seventy one healthy students (40 girls, age: 22.9 ± 08 yr.) were examined. ECG signal and electrodermal activity (EDA) were continuously monitored: rest (P1) – Stroop test (P2) – recovery phase (P3) – mental arithmetic test (P4) – recovery phase (P5) – negative emotion (P6) – recovery phase (P7). Heart rate variability (HRV) parameters and EDA (sympathetic index) were analyzed from each 5-minute period. HRV parameters: RR interval, spectral power in high frequency band (HF-HRV) reflecting cardiovagal control; symbolic dynamics: 0V% - cardiac sympathetic index, 2LV% - cardiovagal index. Trait hostility was evaluated using Cook-Medley Hostility Scale.
Results: The parameters - logHF-HRV, 2LV% were significantly lower and EDA was significantly higher during all stressors compared to rest ($p < 0.05$). Mean RR interval and 0V% index were significantly higher only in cognitive stressors (P2, P4) compared to rest (P1; $p < 0.01$). The 0V% and EDA were significantly higher in recovery phases (P3, P5, P7) compared to rest (P1; $p < 0.05$). Correlation analysis showed a positive correlation between EDA and hostility in negative emotion and its recovery phase ($r = 0.254$, $p = 0.045$; $r = 0.249$, $p = 0.049$, respectively), and negative correlation between logHF-HRV and hostility in both cognitive stressors ($r = -0.327$, $p = 0.011$, $r = -0.260$, $p = 0.039$, respectively).
Conclusions: Our study confirmed a cardiovagal withdrawal (lower logHF-HRV, 2LV%) and a sympathetic arousal (higher EDA) in response to all mental stressors, while ß-adrenergic activity (0V%) increased only to cognitive stressors. Contrary to cardiovagal indices, EDA and 0V% remained higher in recovery phases indicating a potential sympathetic overactivity during stress profile. Importantly, hostility correlated positively with sympathetic reactivity (EDA) to negative emotion, and negatively with cardiovagal reactivity (logHF-HRV) to cognitive stressors reflecting a dependence of personal trait on the type of stressor. We suggest that detailed stress study in the interaction with subjective characteristics could illuminate the pathway via which psychosocial factors may contribute to cardiovascular risk.
Support: VEGA 1/0087/14, APVV 1/0235/12.

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Palliative care in pediatric cardiology – when should it be considered?

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Introduction: Pediatric palliative care program started in 1994. Recently there is more discussion about the necessity of introducing the palliative care approach in pediatric cardiology. The aim of this study was to evaluate patients with congenital heart defects (CHD) who were under the hospice care in our institution between 1994 and 2013, and in the whole country in 2012.
Material: During 19 years, 63 children with CHD were under the home palliative care. There were 19 children with isolated CHD and 44 with chromosomal aberrations. in the whole country in 2012.
In the whole country there were 80 patients with CHD, 28 with isolated and 52 with chromosomal aberrations
Results: Period of palliative home care lasted from 3 to 1269 days, mean 161 days.
In our institution the diagnosis changed between 1994–2013. Until 2000–9 teenagers with complicated CHD who were not operated on due to pulmonary hypertension or severe pulmonary hypoplasia were admitted mainly due to possibility of home-based oxygen supply and hemodilutions. Since 2001: 10 children with post-operative complications, 3 after prenatal diagnosis, 4: T21 and pulmonary hypertension, 47 with lethal chromosomal aberrations, mainly T13, T18, and complicated T21. In the country the diagnosis were similar: single ventricle physiology after different stage of treatment (17), hypoplastic pulmonaty arteries (7), T13(7), T18(30) and complicated T21.
Since 1999 perinatal palliative care was established, for fetuses with lethal chromosomal aberrations complicated by congenital heart defects or with unoperable isolated CHD. 37 patients were admitted to the hospice program after prenatal diagnosis and consultation. None of those children was operated on. One newborn with HLHS diagnosed prenatally was under the hospice care for 35 days.
Conclusions: Palliative care should be considered in all children with life limiting conditions. In patients with complicated CHD in whom surgical treatment failed such options should be discussed with parents. Palliative care should be the method of choice for fetuses and neonates with lethal chromosomal disorders whose parents were against termination of pregnancy. Such approach should be discussed with parents if severe inoperable isolated CHD is diagnosed in fetuses.

P-252

The impact of an interstage monitoring programme on parents of children with a hypoplastic left heart syndrome before and after bidirectional cavopulmonary anastomosis

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Background: Studies have shown the importance of home monitoring for children with a hypoplastic left heart syndrome (HLHS) to reduce the interstage mortality between stages I and II. But very little is known about the parents' experience and impact on quality of life from coping at home with the responsibility of an interstage monitoring programme including saturation measures and body weight gain.
Methods: This multidisciplinary project used a prospective sequential mixed methods study design to analyse the medical course and psychosocial impact on the parents of an interstage monitoring programme. The parents rated their quality of life with the Short Form Health Survey questionnaire (SF-36) and the German version of the Impact of Family Scale (FaBel) one and 5 weeks following discharge, both before and after bidirectional cavopulmonary anastomosis operation (BDCPA). Semi-structured interviews were conducted twice, 5 weeks after hospital discharge before and after BDCPA. The interviews were analysed using the qualitative content analysis of Mayring.
Results: Between February 2011 and December 2012 ten infants (2 female) with HLHS (7 of 10) or other types of univentricular

heart malformations (3 of 10) and their parents (one single mother) were included. Prenatal cardiac diagnosis was made in seven infants. There were no interstage deaths, but one child could not be discharged after BDCPA due to severe congestive heart failure. SF-36 subscales in the mental health summary were low, especially for vitality (37.0 ± 19.46 for mothers and 43.12 ± 25.9 for fathers) before BDCPA and did not change significantly after BDCPA. FaBel values showed gender differences to the disadvantage of the mothers in most subcategories, with a significant difference in the daily and social burden ($p < 0.047$). The most important category in the interview was "Becoming a family" to cope and find a fragile emotional balance. The parents judged the interstage monitoring primarily as a protective intervention.
Conclusions: For families with HLHS the psychosocial burden before and after the interstage period remains fairly high, but being at home as a family is a crucial experience for parents and reassures them in their parenthood. Health professionals should identify overburdened parents and provide assistance and psychosocial support.

P-253
Double Aortic Arch Anomalies – Anatomic Variations and Their Clinical Impact (15 Years Single Centre Experience)
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Introduction: Edwards' hypothetic double aortic arch (AA) can describe all existing morphological variations: with persistence of both complete AA or with an atretic segment or abnormal interruption of one arch. Clinically dominant are ring-forming subtypes, surrounding trachea/oesophagus and causing symptoms from compression; but appreciating other AA anomalies is important as well.
Methods: Retrospective analysis of all consecutive patients with double AA variations managed during 15 years at our institution (one national centre with 1.2 million paediatric population). This long-term study, similarly like a registry, reflected epidemiology of these rare anomalies.
Analyzed were 27 patients (16F/11M); defining double AA morphological subtypes: Group 1 (ring-forming), Group 2 (w/o vascular ring), Group 3 (with persistent 5^{th} AA). Analyzed were associated congenital heart defects (CHD) and non-cardiac diseases as well.
Results: In Group 1 were 17 patients (6 with complete double AA, 5 with right AA+dorsal left AA atresia, 6 with right AA and aberrant left subclavian artery+left arterial duct/ligament) with median age at diagnosis 5 months. Only 2 patients (11.8%) in this group had an associated CHD but in 5 patients (29.4%) also non-cardiac genetic syndromes were present. Fifteen patients (88.2%) manifested with compression symptoms (stridor and/or dysphagea) and underwent surgical transsection of the minor AA or ligament.
In Group 2 were7 patients with AA anomalies w/o a vascular ring (with arterial duct from arterial trunk, contralateral to AA side). All presented as newborns and were associated with cyanotic duct-dependent CHD. No non-cardiac pathology was found in this group.
In Group 3 were 3 patients with persistent 5^{th} AA; with median age at diagnosis 5 months. All patients had other CHD, as well as non-cardiac diseases; and AA anomaly was only an accessory finding.
Conclusions: Our study showed population occurrence-rate of all double AA anomalies 0.023% per thousand children; ring-forming subtypes 0.014%. Understanding the double AA concept is essential to define exactly vessel structures. Although rare, it is of immense clinical importance not only in patients with vascular ring but also when differentiating arterial duct from other collateral pulmonary blood supply in newborns with cyanotic (especially duct-dependent) CHD.

P-254
Evaluation of cardiac muscle microvessel density in children diagnosed with cyanotic heart defects
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Introduction: The cause of cyanotic congenital heart defect or its classification, provoking systemic modifications in internal organs is chronic hypoxia. Tissue hypoxia is mostly the reason to response as an adaptive to angiogenesis. An extent of that process in children's cardiac muscle diagnosed with congenital cyanotic heart defects is not well established.
In line with the above, the aim of the research was to estimate cardiac muscle microvessel density (MVD) in children with cyanotic (study group) and non-cyanotic (control group) heart defects and to evaluate prognostic significance of MVD value in evolution of ventricular dysfunction observed in the post-operative period.
Methods: The study group included 42 children diagnosed with heart defects. The control group comprised 33 patients with non-cyanotic heart defect. Histological material included cardiac muscle sections obtained from interventricular or interatrial wall during surgical correction.
An indirect immunocytochemical procedure with the use of monoclonal mouse anti-human antibodies against CD31 and CD34 was employed to estimate MVD (number of microvessels per $1\,mm^2$).
Results: The mean cardiac muscle MVD in the study group amounted to 596.7 ± 32.6 microvessels per $1\,mm^2$ and it was not significantly different from the mean MVD in the control group (461.2 ± 30.5).
In non-cyanotic heart defects an inner area of subendocardial meshwork was estimated with only 75.3 ± 7.0 microvessels per $1\,mm^2$. An adequate area in cyanotic heart defects had a significantly higher MVD value and was calculated with 92.8 ± 10.9 microvessels per $1\,mm^2$ ($p = 0.0082$). No significant correlations between MVD value and ventricular dysfunction in studied children were found.
Conclusions: The mean cardiac muscle MVD in the study group amounted to 596.7 ± 32.6 microvessels per $1\,mm^2$ and it was not significantly different from the control group (461.2 ± 30.5). Interestingly, in non-cyanotic heart defects an inner area of subendocardial meshwork was estimated with only 75.3 ± 7.0 microvessels per $1\,mm^2$. An adequate area in cyanotic heart defects had a significantly higher MVD value and was calculated with 92.8 ± 10.9 microvessels per $1\,mm^2$ ($p = 0.0082$). No significant correlations between MVD and ventricular dysfunction in studied children were found.

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Aorto-left ventricular tunnel: Think about and treat early

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Introduction: Aorto-ventricular tunnels are rare, congenital, extracardiac channels between the ascending aorta and the cavity, mostly of the left ventricle (LV).
Depending on the size of the tunnel, symptoms vary between none and overt cardiac failure. Echocardiography shows diastolic backflow from the aorta into the LV, leading to LV enlargement in spite of a normally developed aortic valve. We present two cases with aorto-LV tunnel diagnosed at different ages.
Case Reports: In the first patient, diagnosis was made in the 35th week of gestation and confirmed after birth by echocardiography. After an unremarkable postnatal adaptation, the newborn quickly developed cardiac failure and, on echo, progressive LV dilation and decreased contractility. Surgical repair with pericardial patch closure was performed at the age of 18 days without complications and with immediate normalisation of LV size and function. After a follow-up of 4.5 years, LV and aortic valve function is normal.
The second patient was assessed for a heart murmur at the age of 5 years, and diagnosis of aortic regurgitation was made. During follow-up, severe arterial hypertension with remarkably low diastolic blood pressure values and a progressive LV dilation due to regurgitant flow into the LV were documented at regular cardiologic examinations. Correct diagnosis of aorto-LV tunnel was finally established by a congenital cardiologist at the age of 15 years, as all clinical signs of aortic run off with severe LV dilation and a slightly reduced contractility were present (EF 50%). Cardiac surgery with patch closure of the aortic ostium and direct closure of the ventricular ostium was immediately initiated. The postoperative course was prolonged due to a moderately decreased LV function and postpericardiotomy syndrome. The patient was discharged on cardiac medication, and 6 months later LV size and function (EF 52%) are still abnormal with moderate aortic regurgitation present.
Conclusion: Aorto-LV tunnel is a rare lesion mimicking aortic valve regurgitation. Diagnosis should be suspected in the presence of LV dilation and severe run off in the aortic arch without obvious aortic valve lesions. Early diagnosis and surgical intervention prevent long term sequelae such as LV dysfunction.

P-256

Arterial stiffness – a novel important cardiovascular risk factor in the pediatric essential and white coat hypertension?

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The term ,,arterial stiffness'' denotes alterations in the mechanical properties of arteries, which may represent higher cardiovascular risk in middle-aged and older adults. Recently, the question regarding the arterial stiffness in patients with risk factors for the early progression of systemic atherosclerosis in vulnerable adolescent age-period are rare. Thus, the aim of this study was to evaluate the arterial stiffness and early atherosclerotic changes in adolescents suffering from essential and white coat hypertension using noninvasive markers.
Methods: Twenty-eight patients (14 boys) with essential hypertension (EH) and white coat hypertension (WCH) without clinical symptoms of other cardiovascular diseases and age/gender-matched controls were examined. The age-period of all subjects ranged from 17 to 18 years. Evaluated parameters - cardio-ankle vascular index (CAVI), brachial-ankle pulse wave velocity (baPWV) and carotid-femoral PWV (cfPWV) - were assessed using the system VaSera 1500 (Japan). In addition, mean heart rate was evaluated.
Results: Mean heart rate and baPWV were significantly higher in adolescents with WCH than healthy subjects ($p < 0.01$). However, CAVI and cfPWV mean values did not differ significantly between EH (5.35 ± 1.06; 8.7 ± 3.0, respectively), WCH (5.58 ± 0.79; 7.7 ± 2.5, respectively) and control groups (5.02 ± 0.71; 7.9 ± 2.0, respectively).
Conclusions: Our results of indcreased brachial-ankle pulse wave velocity associated with tachycardia indicate a potential sympathetic overactivity as a major pathomechanism leading to the development and stabilisation of hypertension. Furthermore, no significant differences in other parameters between individual diagnoses at this age-period were found. We suggest that further research regarding the potential atherosclerotic changes using the sensitive noninvasive parameters in pediatric hypertension is important.
Support: VEGA 1/0087/14.

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Characteristics of Cardio-Ankle Vascular Index (CAVI) in healthy Slovak children

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Purpose: Recently, the cardio-ankle vascular index (CAVI) is considered as a novel noninvasive index of the arterial stiffness from the beginning of the aorta to the ankle. CAVI represents an important marker of early atherosclerotic changes that is significant for a consequent evaluation of the atherosclerosis severity. We aimed to determine the CAVI values in the group of otherwise healthy children and adolescents necessary for a comparison with CAVI values in pathological states. Moreover, the second aim was to assess the age and gender influence on the CAVI in healthy children and adolescents.
Methods: We examined 117 healthy Slovak children at the age from 7 to 21 years (58 boys) without clinically observed cardiovascular risk factors. CAVI values were evaluated using the system VaSera 1500 (Japan).
Results: The CAVI normal values are presented in graphical forms for total group, and separately for boys and girls. In healthy children, the CAVI increased linearly with age from 7 to 21 years. Gender analysis did not show significantly differences between boys and girls at this age-period. Additionally, CAVI values were independent on the blood pressure measurement at the same time.
Conclusion: Our study firstly presented the CAVI normal values for Slovak population of children and adolescents at the age from 7 to 21 years. Importantly, the CAVI was dependent on the age during this developmental period. Concluding, our CAVI values can be used for detection of the early atherosclerotic changes in children with essential hypertension.
Support: VEGA No. 1/0087/14.

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Acute Effects of Maximal Endurance Exercise on Arterial Stiffness in Children, Adolescents and Young Adults

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Objective: Measures of arterial stiffness are surrogates for cardiovascular health and predict cardiovascular events. Arterial stiffness is responsive to acute physiologic stressors like exercise. However, the acute effects of intensive exercise on post exercise arterial stiffness and associations to the fitness level measured by peak oxygen uptake are commonly unknown especially in the pediatric cohort.
Patients and methods: 119 healthy male children, adolescents and young adults (mean age 16.3 ± 5.4 years, BMI 19.7 ± 2.6 m^2/kg) underwent evaluation of their arterial stiffness, as well as central and peripheral blood pressure after resting 5 minutes in supine position using the oscillometric Mobil-O-Graph. Afterwards they performed a cardiopulmonary exercise test (CPET). Finally, 5 minutes after terminating CPET a second measurement of arterial stiffness parameters was obtained.
Results: Measures of arterial Stiffness are increased 5 minutes after acute exercise. Systolic blood pressures was still increased (118 ± 9 mmHg to 123 ± 11 mmHg; $p < .001$) as well as central blood pressure (108 ± 10 mmHg to 114 ± 12 mmHg; $p < .001$). The stiffness parameters augmentation index (12.9 ± 15.9% to 17.1 ± 10.7%; $p = .014$) and pulse wave velocity (5.0 ± 0.46 m/s to 5.3 ± 0.54 m/s; $p < .001$) were also still enhanced.
Unfortunately, no significant association of peak oxygen uptake to arterial stiffness at baseline or the change during exercise could be found after correcting data for age by partial correlation.
Conclusions: Arterial stiffness is increased shortly after maximal endurance exercise. However, no association could be found between exercise capacity, measures of arterial stiffness and changes in stiffness parameters after acute exercise. Longer post-exercise observation studies are needed to evaluate the effects of exercise on cardiovascular stiffness parameters that, maybe, are comparable to post-exercise hypotension.

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Prevention in school: one day workshop about cardiovascular health

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Introduction: The average german student spends 9 hours a day sitting in school, doing homework, in front of the computer or TV. When chips and chocolate bars for lunch are added we face a horror scenario that explains increasing numbers of overweight and obese children. To make children aware of positive effects of a healthy lifestyle, we set up a prevention program that can be included in every school setting.
Methods: The prevention program ,,Sternstunden der Gesundheit'' was conducted in the school district of Berchtesgadener Land, Germany. It consisted of a noninvasive ultrasound measurement of carotid intima media thickness (cIMT) and parameters of arterial stiffness (pulse wave velocity, augmentation index, stiffness index β). Health-related fitness (HRF) was assessed using the FITNESSGRAM® test battery. Students were tested for strength, flexibility and aerobic capacity. Anthropometric measures consisted of body weight and height, waist circumference, body mass index, waist-to-height and waist-to-hip ratio. Children and their parents filled in questionnaires about nutritional behavior and family history. The additional program was adapted according to age and school type. Children learned about the circulatory system, prevention of cardiovascular disease, healthy nutrition, performed ultrasound measures and chemical food analysis. The study was funded by ''Sternstunden e.V''. and ''Landratsamt Berchtesgadener Land''.
Results: 1017 children from 14 schools and 47 different classes respectively, aged 7-17 years (483 boys/ 534 girls) participated in the program ''Sternstunden der Gesundheit''. HRF data was calculated for the entire study population, vascular data was analyzed for 736 children (330 boys/ 406 girls). The program was very well accepted by pupils, teachers and parents and gave insight into cardiovascular health data of an entire district.
Conclusion: The program ''Sternstunden der Gesundheit'' can be applied in any school setting due to numerous variations in content and can be implemented within a regular school day. Ultrasound measurements are a cost-intensive factor and need trained examiners but provide important data for diagnosis of atherosclerosis. Further analysis will include assessment of normative values for cIMT and vascular stiffness data, gender comparisons of HRF for age, body composition and different school types and the association between vascular data and HRF.

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Lipid profile and body weight in relation to infant feeding

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Introduction: The process of atherosclerosis (according to the latest research of inflammation), begins to develop in the first years of life. High levels of cholesterol in breast milk could lead to reprogramming of its metabolism and prevent the development of hypercholesterolemia in the adult age. Long-term duration of breastfeeding can protect the child from infection, adequate advance on weight, prevention of obesity.
Aim: examination of the serum lipids profiles and C reactive protein in infants fed breast milk and formula milk, duration of breastfeeding in relation to lipoproteins and body mass index.
Methods: prospective clinical study was performed from 06. 2011. till 10. 2013., in Pediatric Clinic, Clinical Center of Sarajevo. Study included 100 patients, and formed two groups according to age: 6 months and 12 months, and analysis lipids, CRP, anthropometric parameters in relation to diet. Its realized the detailed information about the period of pregnancy, birth weight, duration of breastfeeding, and the beginning of complementary food. Mothers were evaluated: diet during breastfeeding, maternal body weight before and after pregnancy.
Results: High-density lipoprotein (HDL) were higher in breastfeed infant. (p-0.024) Total cholesterol and other lipoproteins are not significantly changed, compared to the infant nutrition. Longer duration of breastfeeding leads to an increase in total cholesterol (p-0.001), low density lipoproteins (p-0.003) and C/HDL) p-0.015; resulting in a positive effect on reprogramming of cholesterol metabolism. Different diet did not influence the development of overweight or obesity. Longer duration of breastfeeding affects the reduction of the control in CRP. ($p = 0.045$). Breastfeeding significantly reduce maternal body weight after 3 months of breastfeeding (p-0037)
Conclusion: breastfeeding have, proven, short and long-term benefits. The results of our study showed insignificance in the lipids profile, body weight compared to the deferent infant diet.
Keywords: breastfeeding, lipids, CRP, body weight.