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O1-1

Increased nuchal translucency and its relationship to fetal congenital heart disease

Vogel M. (1), Sharland G.K. (1), Zidere V. (2), Simpson J.M. (1), Miller O. (1), Allan L.D. (2)
Evelina Children's Hospital, London, UK (1); Kings College Hospital, London, UK (2)

Objective: Increased nuchal translucency (NT) at the 11–14 week scan has been shown to be associated with chromosomal anomalies and congenital heart disease. The rate of association has varied between 17 and 56% in different publications and suggested there is no link between increased NT and specific types of congenital heart disease (CHD). Our aim was to explore these aspects in more detail.
Methods: All fetuses with CHD, seen during a 9-year period between 1998 and 2006, were identified retrospectively from King's College (KCH) and Evelina Children's Hospital (ECH) databases. Data collected included the type of CHD, the NT measurement and the karyotype at each centre.
Results: There were 1626 fetuses with CHD where the NT measurement was known. The karyotype was known in 1102 of these. Of 536 fetuses with known normal chromosomes the NT was increased in 43%. Of 566 fetuses with known chromosomal abnormalities, NT was increased in 73%. The incidence of increased NT was higher in the KCH group than in the ECH group, this was related to the higher incidence of karyotype anomalies in the former group. Because of this, only cases with a known normal karyotype were included in the analysis. Five categories of CHD were chosen for more detailed study, coarctation of the aorta, hypoplastic left heart syndrome, transposition of the great arteries, atrioventricular septal defect and tetralogy of Fallot.
Of 119 cases with coarctation, 52 had an increased NT (44%). Of 60 cases of the hypoplastic left heart syndrome, 24 had an increased NT (40%). Of 92 cases of transposition, 29 had an increased NT (31%). Of 59 cases of atrioventricular septal defect, 32 had an increased NT (54%). Of 104 cases of tetralogy of Fallot, 40 cases had an increased NT (40%).
Conclusion: Major CHD is associated with increased NT in 30–54% of cases. The rate of association varies with the type of CHD present. Studies which examine this connection must exclude fetuses with chromosomal anomalies as these fetuses will skew the results, and must have sufficiently large groups of different types of CHD to make meaningful observations.

O1-2

Preventive effect of periconceptional folic acid supplementation on the risk of congenital heart defects: A registry based case-control study in the Netherlands

van Beynum I.M. (1), Kapusta L. (1), Bakker M.K. (2), den Heijer M. (1), Blom H.J. (1), de Walle H.E.K. (2)
Radboud University Nijmegen Medical Centre, Nijmegen, the Netherlands(1); EUROCAT-Registration Northern Netherlands, University Medical Centre Groningen, The Netherlands (2)

Evidence is emerging that multivitamins containing periconceptional folic acid supplementation protects against the occurrence of congenital heart defects (CHD). Postulating that folic acid is responsible for the reduction in CHD risk we used data from a large surveillance for birth defects (EUROCAT- Northern Netherlands registry from 1981 to 2006) to perform a case-control study to investigate the effect of periconceptional folic acid supplementation on CHD risk. We analysed data reported from January 1st 1996 until June 30th 2006. This was one year after the mass media campaign initiated to promote, to women capable of becoming pregnant, the recommended periconceptional folic acid supplements primary advised to reduce the risk of neural tube defects.
The cases consisted of mothers who delivered infants with isolated or complex heart defects, without any syndrome or genetic abnormality (N = 613). The control group consisted of mothers who gave birth to children with a known chromosomal or genetic defect or infants with other congential malformations (N = 2385). In both the case and control group, mothers of children with oral cleft, urinary tract, limb reduction and neural tube defects were excluded, because the risk of these defects are probably reduced by maternal folic acid supplementation. Potential confounding factors of periconceptional folic acid use included; maternal body mass index, education, maternal age at delivery of index baby, smoking behaviour and alcohol use during pregnancy were explored.

Generally, we observed a declining trend in the prevalence of CHD since 1993. Adequate use of periconceptional folic acid supplements revealed an odds ratio of 0.81 (95% CI 0.67–0.96) for all types of CHD. Subgroup analysis showed an odds ratio of 0.60 (95% CI 0.42–0.86) for isolated ventricular septal defects. Periconceptional folic acid supplements appear to reduce the prevalence of CHD with approximately 20%. Considering the relatively high prevalence of CHD worldwide the findings of this study are important for public health. From this study we might conclude that adequate periconceptional folic acid supplementation plays a role in the prevention of CHD.

O1-3
Fetal Pulmonary Venous Doppler Flow Pattern: Predicting the Need for Emergent Atrial Septostomy in Newborns with Hypoplastic Left Heart Syndrome(HLHS)

Jaeggi E.T., Chaturvedi R., Cavalle-Garrido T. Jaeggi M.
Division of Cardiology, The Hospital for Sick Children, Toronto, Canada

Background: In utero pulmonary venous (PV) Doppler flow has been correlated with the severity of atrial septal restriction and the need for emergent atrial septostomy (EAS) in newborns with HLHS. According to Michelfelder et al (Circulation 2005), a fetal forward/reverse PV Doppler flow velocity-time integral (PV VTI) ratio <5 most accurately (sensitivity: 88%; specificity: 97%;) predicted the need for EAS, required in 20% of their HLHS cases. We reviewed our contemporary experience with this entity.
Methods: The fetal and postnatal echocardiographic and clinical findings of 49 consecutive cases with actively managed HLHS were reviewed. PV Doppler assessments included maximal S-, D- and A-wave flow velocities (cm/s); A-wave duration (ms), S/D wave and forward/reverse PV VTI ratios. EAS <1 days of age and survival >28 days were used as clinical outcome variables.
Results: Three (6%) of the 49 cases required an EAS for an intact (n = 1) or severely restrictive atrial septum. The table indicates the median (range) fetal Doppler indices at the last trimester echocardiogram of newborns with/without EAS:

Parameters	Emergent AS (N = 3)		No Emergent AS (N = 46)		p-values
Fetal age at last echo (weeks)	36	(35–38)	35	(28–39)	NS
S-waves	53	(42–56)	42	(15–73)	NS
D-waves	0	(0–11)	18	(11–43)	NS
S/D wave ratio	n/a	(n/a–5.1)	2.2	(0.9–5.7)	NS
A-waves	47	(38–57)	21	(11–72)	< 0.001
A-wave duration (ms)	105	(90–112)	65	(30–88)	< 0.0001
Forward/reverse VTI ratio	1.5	(1.3–2.3)	7.7	(1–23.7)	< 0.001

All 3 cases requiring EAS had a forward/reverse PV VTI <2.5 on 3rd trimester echocardiography while this was the case in only 1 (2%) of 46 without a need of EAS (p = 0.002; sensitivity 100%; specificity: 98%). Moreover, the need of EAS was associated with increased neonatal mortality (100% versus 14%; p = 0.005).
Conclusions: Fetal PV Doppler flow parameters that represent atrial systole (A-wave peak velocity and duration, forward/reverse VTI ratio) differ significantly among cases with and without a need of neonatal EAS. Cases with a fetal forward/reverse PV VTI ratio <2.5 were most likely (3/4 cases) to die as neonates despite successful EAS.

O1-4
Haemodynamic assessment of prenatally treated arrhytmias in fetuses

Tomek V. (1), Jičínská H. (2), Škovránek J. (1), Gilík J. (1), Marek J. (1)
Kardiocentrum and Cardiovascular Research Centre, University Hospital Motol, Prague, Czech Republic (1); Department of Pediatric Cardiology, University Hospital Brno, Czech Republic (2)

Fetal isolated congenital atrioventricular complete block (CAVB) and supraventricular tachycardia (SVT) complicated by myocardial dysfunction and hydrops fetalis carries a significant risk of morbidity and mortality. The aim of our study was to assess the haemodynamic changes by prenatal echocardiography due to treatment of fetal CAVB and SVT.
Method: Cardiothoracic index (CTI), left ventricular shortening fraction (SF), index of atrio-ventricular velocities in inferior vena cava (AVI) and fetal heart failure score (FHS), were measured prospectively by prenatal ECHO before and after antiarythmic drug administration for CAVB and SVT.
The cohort consisted of 32 fetuses with structurally normal hearts presenting with documented fetal supraventricular tachycardia (heart rate >180/min, gestational age 22 to 35 weeks, median 26) and 12 fetuses with CAVB (mean ventricular rate 59/min, gestational age 19-28 weeks, mean 21).
Results: CAVB: Parameters of heart failure improved in all 12 fetuses treated (salbutamol,dexamethason): SF improved from 0.34 ± 0.05 to 0.41 ± 0.06 (p = 0.03), CTI from 0.35 ± 0.09 to 0.31 ± 0.06 (p = 0.01), and FHS from 7.82 ± 1.72 to 9.27 ± 0.65 (p = 0.01). Ventricular rate increased from 55.75 ± 7.36 to 63.25 ± 10.48 (p = 0.02).
SVT: Digoxin was used as first drug choice in all cases. Sotalol was added in 10 and 1 fetus was converted by intracordal infusion of amiodarone. In non-responding fetuses (N = 8), ECHO parameters of heart failure did not change significantly during the treatment. In responding fetuses (N = 24) all parameters improved: CTI from 0.330 ± 0.0561 to 0.278 ± 0.0481, SF from 0.284 ± 0.0632 to 0.352 ± 0.0687, AVI from 0.774 ± 0.201 to 0.401 ± 0.146) and FHS from 6.267 ± 2.017 to 9.800 ± 0.414; P < 0.001 in all. Pretreatment SF was significantly lower in non-responders (p < 0.001) whereas all other parameters before the treatment did not differ significantly between the groups.
Conclusions: Echocardiography allows for reliable monitoring of fetal heart and placental circulatory function. The regression of heart failure is achieved by the conversion to sinus rhythm in SVT and increase of ventricular rate in CAVB. Severe decrease of left ventricular systolic function is a predictor of unsuccessful treatment of SVT.
The study was supported by grants NR 9451-3/2007 and VZ00064203.

O1-5
Outcomes of Fetuses with Potential Multiple Left Heart Obstructive Lesions (Shone's Complex)

Han R.K., Yoo S.J., VanArsdell G.S., McCrindle B.W., Jaeggi E.T.
Hospital for Sick Children, Toronto, Canada

Background: Multiple left heart obstructive lesions have a wide spectrum of postnatal outcomes ranging from hypoplastic left heart syndrome requiring single ventricle palliation to biventricular circulation which may not require any intervention. Lack of

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predictive factors at the time of fetal diagnosis hampers our ability to counsel families appropriately.
Objectives: To determine the outcomes of fetuses diagnosed with potential multiple left heart obstructive lesions, and the value of fetal echocardiographic features in predicting postnatal outcomes.
Methods: Review of 45 fetuses (median gestational age at diagnosis: 27 weeks; range: 18–40 weeks) since 1999 with potential multiple left heart obstructive lesions (e.g. small but apex-forming left ventricle, Shone's complex) and with known postnatal outcomes. Excluded were obvious hypoplastic left heart syndrome and isolated aortic valve stenosis at fetal diagnosis. Offline measurements from serial fetal echocardiograms were normalized for gestational age as Z-scores and compared to postnatal outcomes.
Results: Postnatal diagnosis of Shone's complex was established in 9 patients (20%) while an additional 26 patients (58%) had small left heart structures at different levels. Additional postnatal diagnoses include restrictive foramen ovale (2), endocardial fibroelastosis (1), pulmonary valve stenosis (1), and normal hearts (6). Surgical intervention was required in the neonatal period in 21 patients (47%) of which 5 underwent single ventricle palliation and 16 had extended aortic arch repair only. Factors associated with single ventricle palliation were greater R:L disproportion at the level of the pulmonary and aortic valve annulus (1.7 vs. 1.5; $p = 0.01$), smaller initial mitral valve annulus Z-scores (−4.11 vs. −2.55; $p = 0.03$), and a trend in abnormal foramen ovale flow direction (60% vs. 18%; $p = 0.07$). On serial studies, fetuses with single ventricle palliation also developed greater R:L ventricular disproportion (+0.42 vs. −0.04; $p = 0.02$), primarily as a result of increased RV diameter Z-scores (+2.12 vs. −0.20; $p = 0.02$).
Conclusions: Fetal echocardiograms demonstrate greater R:L disproportion, smaller mitral valve Z scores, and higher likelihood of abnormal flow across the foramen ovale in patients who require single ventricle palliation. However, there was considerable overlap precluding a clear cutoff in values that would predict postnatal outcomes.

O1-6
A Fetal Cardiology Telemedicine Program: Acceptability, Social Factors and Impact on Maternal Anxiety.
McCrossan B.A., Sands A.J., Casey F.A.
Department of Paediatric Cardiology, Royal Belfast Hospital for Sick Children, UK

Introduction: The possibility of congenital heart disease in an unborn baby is an added stress for expectant mothers referred for fetal cardiology evaluation. Patients often wait weeks for an appointment and travel large distances for assessment at the regional centre.
This study aimed to evaluate the effect of a fetal cardiology telemedicine program on patient travel and subsequent days off work. This study also aimed to evaluate the acceptability of telemedicine and effect on maternal anxiety.
Methods: This was a prospective study over 16 months. An initial fetal echocardiogram (FE) was performed by the radiographer in the district general hospital (DGH). This was followed by a telemedicine consultation, incorporating a FE performed by the radiographer with live guidance and interpretation by the fetal cardiologist. The fetal cardiologist then counselled the parents. A third FE was performed later at the regional fetal cardiology centre (75 miles from DGH). A five point structured questionnaire was completed at DGH and regional centre relating to satisfaction with the consultation and preferred method of FE (5 point Likert scale). Anxiety levels were also assessed following each consultation using STAI questionnaires.
Results: Sixty-four remote FE consultations have been performed. The mean difference in distance travelled, return journey time, days off work to attend consultation and cost of travel to DGH and regional centre are shown in Table 1.

Table 1. Logistics of attending fetal cardiology appointments at district general hospital and regional centre.

Mean	DGH (Telemedicine)	Regional Centre ("Hands-on")	Paired sample Difference	P-value
Distance from home (km)	14.7	117.8	103.1	<0.001
Return journey time (mins)	44.7	235.9	191.7	<0.001
Cost of journey €	13.10	94.29	75.10	<0.001
Days off to attend consultation (including partner)	0.85	1.49	0.64	<0.001

Respondents preferred to have FE performed via tele-link at the DGH (Mean difference = 1.3/5, $p < 0.001$). Mean consultation ratings were very high for telemedicine and "hands-on" consultations, 23.5/25 and 23.3/25 respectively.
Anxiety levels were significantly reduced following the telemedicine consultation (mean STAI decrease = 9.4, $p < 0.001$). There was a trend towards lower STAI scores following the telemedicine consultation compared with "hands-on" consultation (Mean difference = 4.63, $p = 0.052$).
Discussion: FE facilitated by a live telemedicine link is highly acceptable to pregnant women and reduces anxiety. It is significantly time and cost saving for the patient. The cost to society of establishing a telemedicine service may, in part, be offset by a reduction in absenteeism in patients.

O2-1
Early Postoperative Arrhythmias in over 800 Consecutive Congenital Surgical Patients
Kansy A. (1), Rękawek J. (2), Knecht M. (2), Paszke M. (2), Bieganowska K. (2), Szymaniak E. (2), Burczyński P. (1), Kawalec W. (2), Brzezińska-Rajszys G. (2), Rytko D. (3), Manowska M. (3), Birbach M. (1), Maruszewski B. (1)
Department of Cardiothoracic Surgery, The Children's Memorial Health Institute, Warsaw, Poland (1); Department of Cardiology, The Children's Memorial Health Institute, Warsaw, Poland (2); Department of Anesthesiology and Intensive Care, The Children's Memorial Health Institute, Warsaw, Poland (3)

Background: Early postoperative arrhythmias are well recognized complications after Congenital Heart Surgical procedures.
Methods: Diagnosis and treatment of early postoperative arrhythmias were analyzed prospectively in 806 consecutive patients aged from 1 day to 18 years (mean 25.5 months), operated between January 2005 and December 2006 in our Institute. All children were admitted to the Intensive Care Unit (ICU) and continuous ECG monitoring was established. Risk factors, such as age, weight, and Aristotle Basic Score (ABS), opened sternum, CPB time, AoX time, use of DHCA were analyzed.
Statistical analysis using Students t-test, the Mann-Whitney U-test, or Yates Chi-square test was performed. Multivariate stepwise logistic regression was used to assess the risk factors for postoperative arrhythmias.
Results: Arrhythmias occurred in 124 pts out of 806 (15.4%). The most common types of arrhythmia were: SVT (39), JET

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(31), AV block (17). Such risk factors for arrhythmias as lower age (p = 0.0016*), lower body weight (p = 0.0013*), and higher ABS (p = 0.000001*), longer CPB time (p = 0.000001*), AoX time (p = 0.000001*) and use of DHCA (p = 0.00038*) were identified in a univariate analysis. The multivariate stepwise logistic regression showed statistical significance of: higher ABS (p = 0.000001*), lower body weight (p = 0.00005*), longer AoX time (0.0031*) compared to age p = 0.34, and CPB time (p = 0.21). The probability of arrhythmias was higer with opened sternum (p = 0.000001). The onset of arrhythmias was associated with higher mortality (p = 0.014), longer IPPV time (p = 0.000001) and longer LOS (p = 0.000001).
Conclusion: Younger age, lower body weight, and higher ABS, longer CPB time, AoX time, use of DHCA are the risk factors for postoperative arrhythmias. SVT and JET were the most common postoperative arrhythmias. The onset of arrhythmias is associated with higher perioperative mortality, longer IPPV time and lenght of stay.

O2-2
Restrictive enlargement of the pulmonary annulus at surgical repair of tetralogy of Fallot – 10 years experience with a uniform surgical strategy

Voges I., Schumacher M., Scheewe J., Fischer G., Jung O., Kramer H.H., Uebing A.
Department of Paediatric Cardiology and Cardiovascular Surgery, University Hospital of Schleswig-Holstein, Campus Kiel, Kiel, Germany

Introduction: Transannular patch enlargement (TAP) of the right ventricular outflow tract (RVOT) at repair of tetralogy of Fallot (TOF) aggravates pulmonary regurgitation (PR) with deleterious effects on long-term outcome.
It has been shown that the diameter of the pulmonary annulus (PAD) after TOF repair determines the extent of PR but does not lead to significant RV pressure load even in patients with a PAD close to the fourth lower standard deviation (SD) of normal.
Since 1997, therefore, we follow a surgical strategy that aims to use a TAP only in patients in whom the PAD is below the fourth lower SD. In addition, if patching of the annulus is needed it should be enlarged only to the second lower SD.
The purpose of this retrospective study was to evaluate whether this strategy was sufficient to reduce the frequency of TAP and to minimise postoperative PAD. The extent of any residual RVOT obstruction and the incidence of reoperation for significant RVOT obstruction (RVOTO) should also be assessed.
Methods: Early postoperative angiocardiographic and echocardiographic data of 217 TOF patients were analysed. 116 patients were operated between 1977 and 1996 without a uniform strategy (Group 1) and 101 patients underwent repair between 1997 and 2006 according to the strategy mentioned above (Group 2).
Results:

	Age at repair (Years; IQR)	TAP (%)	PAD (Z-value)	Vmax - RVOT (m/s)	Redo for RVOTO (n)
Group 1 1977–1996	1.7 (0.6–2.2)	68	−0.9 ± 1.6	2.1 ± 0.8	1
Group 2 1997–2006	0.8 (0.5–1.2)*	32†	−1.7 ± 1.6*	2.4 ± 1.1	3

$P < 0.05$ (t-Test); †$P < 0.05$ (Fisher's exact test)

Conclusion: Restrictive enlargement of the pulmonary annulus at TOF repair results in a decreased transannular patching rate; is effective in limiting postoperative pulmonary annulus size and does not result in significant RV pressure load with need for reoperation.
A limitation of pulmonary regurgitation with beneficial effects on long-term outcome can be expected when the extent of RVOT enlargement at repair is restricted.

O2-3
Is it there an optimal timing for surgical ligation of patent dustus arteriosus in preterm infants?

Vida V.L. (1), Lago P. (2), Salvadori S. (2), Boccuzzo G. (3), Padalino M.A. (1), Milanesi O. (2), Speggiorin S. (1), Butticci R. (1), Stellin G. (1)
Department of Pediatric and Congenital Cardiac Surgery, Padua, Italy (1); Department of Pediatric Cardiology, Padua, Italy (2); Department of Statistics, Padua, Italy (3)

Introduction: We sought to identify the optimal timing for closure of patent ductus arteriosus (PDA) and the cost/effectiveness of early surgical ligation in preterm infants.
Methods: From January 2001 to June 2007, all premature infants with PDA who were ≤32 weeks of gestational age (GA), ≤than 1.5 kilograms of body weight (BW) who received primary medical treatment with ibuprofen, were included. Age in days at PDA closure, GA, BW and mean arterial pressure less than 30 mmHg (MAP < 30) were related to hospital outcome, including hospital mortality, presence of necrotizing enterocolitis (NEC), acute renal failure (ARF), retinopathy (ROP) and chronic lung disease at 36th week (CLD). A logistic regression model was used to analyze the data.
Results: Two-hundred and one consecutive patients were included. Medical treatment was effective in 149 patients (75%). Fifty-two patients (25%) required surgical ligation after failed medical treatment; they had lower gestational age (25 ± 2.2 vs. 27 ± 2.5 weeks, p < 0.0001), lower body weight at birth (744 ± 186 vs. 892 ± 256 grams, p = 0.0002), lower Apgar scores at 5th minute (7.1 ± 1.6 vs. 7.5 ± 1.6 weeks, p = 0.07), and higher incidence of MAP < 30 (38/52, 73% vs. 56/149, 36%, p < 0.0001) than patients who respond to ibuprofen. The prolonged patency of the ductus arteriosus (>11 days) was significantly associated with and increased risk for NEC (OR = 3.3, p = 0.007), ARF (OR = 4, p = 0.04), ROP (OR = 3.45, p = 0.04) and CLD (OR = 3.6, p < 0.0001).
Conclusions: The most effective time for surgical PDA closure in our population is the day 12th of life. Surgical PDA ligation is an effective treatment and should be anticipated in selected patients who frequently fail medical therapy, in order to improve hospital outcome.

O2-4
Could Hyperglycaemia and Troponin I predict outcome in intensive care after congenital heart surgery in children?

Di Bernardo S. (1), Perez M.-H. (2), Stucki P. (2), Sekarski N. (1), Cotting J. (2)
Pediatric cardiology Unit, University Hospital Center (CHUV), Lausanne, Switzerland (1); Pediatric Intensive Care Unit, University Hospital Center (CHUV), Lausanne, Switzerland (2)

Objective: To establish if hyperglycaemia and cardiac Troponin I (cTnI) after congenital heart surgery on cardiopulmonary bypass in children could predict outcome in intensive care unit.
Methods: retrospective cohort study including 274 children (mean age 4.6 years; range 0–17 years-old). CTnI and glucose values were retrieved from our database. Integrated values (area under

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the curve (AUC)) were calculated for evaluation of sustained hyperglycaemia and then normalised per hour (48h-Gluc/h). Maximal cTnI, first glucose value (Gluc1) and 48h-Gluc/h were then correlated with duration of mechanical ventilation, ICU stay and mortality using cut-off values.
Results: The mean duration of mechanical ventilation was 5.1 ± 7.2 days and ICU stay was 11.0 ± 13.3 days, 11 patients (3.9%) died. Hyperglycaemia (>6.1 mmol/l) was present in 68% of children at admission and was sustained in 85% for 48 hours. The mean value of Gluc1 (7.3 ± 2.7 vs. 11.8 ± 6.4 mmol/l, $p < 0.0001$), 48h-Gluc/h (7.4 ± 1.4 vs. 9.9 ± 4.6 mmol/l/h, $p < 0.0001$) and cTnI max (16.7 ± 21.8 vs. 59.2 ± 41.4 mcg/l, $p < 0.0001$) were significantly lower in survivors vs. non survivors. Cut-off values and odds ratio are summarised in Table 1. Analyses for duration of mechanical ventilation and for length of stay in ICU are depicted in Table 2.

Table 1

ICU mortality	Odds ratio	95 % Confidence interval	p value
Gluc1 > 8.3mmol/l	18.5	2.2–158.4	0.0016
48h-Gluc/ h > 8.3mmol/l/h	2.0	0.7–10.8	NS
cTnI max > 35mcg/L	2.6	2.5–30	0.014

Table 2

	Mechanical Ventilation Time (days)			Length of ICU stay (days)		
	<8.3mmol/l	>8.3mmol/l	p value	<8.3mmol/l	>8.3mmol/l	p value
Gluc1	4.8 ± 6.0	6.8 ± 9.1	NS	9.7 ± 9.4	16.2 ± 21.7	0.001
48h-Gluc/h	4.5 ± 6.0	7.0 ± 8.1	0.008	9.6 ± 11.9	14.3 ± 14	0.007
	<35 mcg/l	>35 mcg/l		<35 mcg/l	>35 mcg/l	
cTnI max	4.3 ± 5.3	10.9 ± 10.9	0.0001	9.6 ± 10.4	19.7 ± 21.0	<0.0001

Conclusions: Hyperglycaemia is frequent after cardiopulmonary bypass and sustained in the first 48 hours. Admission glycaemia and cTnI max are associated with a high risk of mortality, prolonged duration of mechanical ventilation and prolonged length of stay in ICU.

O2-5
Perfusion of the Descending Aorta in the Norwood Procedure

Mair R. (1), Sames E. (1), Vondrys D. (1), Lechner E. (2), Vossen S. (2), Tulzer G. (2)
Department of Cardiac Surgery, General Hospital Linz (1); Department of Pediatrtic Cardiology, Children's Hospital Linz (2)

Background: For years standard perfusion method in the Norwood procedure has been deep hypothermia and circulatory arrest. Several techniques of antegrade cerebral perfusion have been introduced, to avoid ischemic damage to the brain. Intestinal organs and kidneys however are not perfused with these methods. Additional perfusion of the descending aorta provides continuous blood flow to all regions of the body, throughout the whole procedure.
Patients and Methods: Since Oct. 2003, 103 consecutive patients underwent a Norwood procedure in moderate hypothermia (25°C) using additional perfusion of the descending aorta (double arterial cannulation–brachiocephalic artery and descending aorta). 89 patients had different forms of HLHS, 8 patients had tricuspid atresia, TGA, hypoplastic aortic arch and restrictive bulboventricular foramen and 6 patients had other forms of single ventricle and systemic outflow tract obstruction. All patients were newborns: age: 9.18 ± 9.17 d(mean ± std), weight: 3.24 ± 0.42 kg(mean ± std).
Results: Survival in the whole group was 89.7% and increased from 83.4% in 2004 to 96.3% in 2007. Maximum intraoperative levels of serum lactate were 3.5 mmol/l and came up 10min after discontinuation of cardiopulmonary bypass.
Serum lactate levels at ICU admission were 4.53 (± 1.58)mmol/l and normalized (< 2 mmol/l) after a mean period of 9h
Conclusions: Additional perfusion of the descending aorta is a safe and feasible method. Excellent organ perfusion is indicated by low intra- and postoperative serum lactate levels. This might minimize the operative trauma and facilitate recovery.

O2-6
Attitudes and practices of North American paediatric cardiologists and cardiac surgeons in the management of hypoplastic left heart syndrome: a survey-based study

Prsa M. (1), Carnevale F. (1), Justino H. (2), Rohlicek C. (1)
Division of Cardiology, Department of Pediatrics, Montreal Children's Hospital, McGill University Health Centre, Montreal, Canada (1); Section of Pediatric Cardiology, Department of Pediatrics, Texas Children's Hospital, Baylor College of Medicine, Houston, USA (2)

Background: Hypoplastic left heart syndrome (HLHS) is associated with high morbidity and mortality. Management options for HLHS include staged palliative surgery, cardiac transplantation, and compassionate care without surgery. Over the past 25 years, clinical management of HLHS in North America appears to have shifted from predominantly compassionate care to surgical intervention. However, controversy continues as to the most appropriate management. There is limited contemporary data concerning the attitudes and practices of paediatric cardiologists and cardiac surgeons in this regard.
Objective: To determine which management options paediatric cardiologists and cardiac surgeons in North America discuss and which options they recommend when counselling parents after a newborn and antenatal diagnosis of HLHS.
Methods: Paediatric cardiologists and cardiac surgeons across the United States and Canada were asked to complete an anonymous, web-based survey about their attitudes and practices in the management of HLHS.
Results: Of 1621 physicians contacted 749 (46%) completed the survey. When counselling parents of newborns with HLHS, 99% of paediatric cardiologists and cardiac surgeons discuss a staged palliative surgery, 67% discuss cardiac transplantation, and 62% discuss compassionate care without surgery. Only a minority (15%) discusses all available options. Staged palliative surgery is recommended over cardiac transplantation or compassionate care without surgery by 77% of physicians. When counselling parents after an antenatal diagnosis of HLHS, 99% of paediatric cardiologists and cardiac surgeons discuss continuation of pregnancy with staged palliative surgery after birth, 53% discuss continuation of pregnancy with cardiac transplantation after birth, 57% discuss continuation of pregnancy with compassionate care after birth, and 75% discuss termination of pregnancy. Only 37% discuss all available options. Continuation of pregnancy with staged palliative surgery after birth is recommended over the other options by 56% of physicians.
Conclusions: Virtually all surveyed Canadian and American paediatric cardiologists and cardiac surgeons discuss a surgical

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intervention when counselling parents about the management of their child or foetus with HLHS. However, only a minority discusses all available options. Most physicians recommend a surgical intervention for management of HLHS. Our intention is to extend this study to Europe.

O3-1

Feasibility of Whole-Heart Steady-State Free Precession Magnetic Resonance Coronary Angiography in Neonates, Infants and Children with Congenital Heart Disease

Tangcharoen T., Bell A., Hegde S., Botnar R., Beerbaum P., Schaeffter T., Razavi R., Greil G.
King's College London, Division of Imaging Sciences, London

Introduction: Whole heart steady-state free precession (SSFP) Magnetic Resonance Coronary Angiography (MRCA) has been successfully used in adult patients for assessment of coronary artery anomalies and the detection of coronary artery stenosis. Imaging of frequently observed abnormal origin, diameter and course of coronary arteries in neonates, infants and children with congenital and acquired heart disease would be beneficial for preoperative assessment and follow-up studies including postoperative assessment after surgical or other interventional procedures. Also patients with Kawasaki disease would benefit from a noninvasive x-ray free imaging method.
Methods: One hundred patients (mean age 3.9±3 years, range 2 months–11 yrs, 57 males) with congenital heart disease were imaged with a Philips Intera 1.5T MR scanner under general anesthesia. After injection of contrast agent (Magnevist® 0.2 mmol/kg), a vector ECG triggered, free-breathing, 3D-SSFP whole-heart approach with navigator gating and a T2 prepulse was used with isotropic image resolution (1.0–1.3 mm^3). The acquisition window was adapted to the rest period of the heart (end-systole n = 56, end-diastole n = 44). Image quality of the coronary arteries were assessed by two independent observers (image score 0 (non visible) to 4 (excellent)). Vessel length and sharpness were evaluated using custom made software (Table 1).

Table 1:

Parameters	Left main trunk	Left anterior descending	Left circumflex	Right coronary artery
Image quality	2.6±1.4	2.5±1.4	1.6±1.3	2.5±1.5
Vessel length (mm)	8.7±4.5	31.3±16.3	25.2±19.4	54.3±24.8
Vessel sharpness (%)	40±14.3	39±15.5	28±20.1	38±18.6

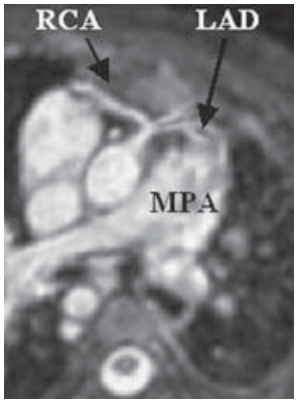

Figure 1: Single orifice of the coronary artery system with the left anterior descending coronary artery (LAD) running across the main pulmonary artery in a 2 years old patient with Tetralogy of Fallot.

Results: All studies were completed without adverse effects (mean acquisition time 4:34 min; mean navigator efficiency 52%). The origin and proximal courses of left and right coronary arteries were visualized with diagnostic image quality (≥2) in 73% of all patients and 95% of patients ≥5 years. In eight patients abnormal coronary artery origins were detected by MRCA (Fig. 1). Quantitative values are summarized in Table 1.
Conclusion: The successful use of whole heart MRCA for the assessment of coronary arteries in 100 infants and children with congenital heart disease was demonstrated. This technique may be used for delineation of abnormal size, origin and course of the coronary arteries in this group of patients.

O3-2

Accuracy of 3D-Real-Time Echocardiography for the assessment of small volumes and distances

Herberg U. (1), Brand M. (1), Trier H.G. (2), von Hahn R. (3), Bernhardt C. (1), Breuer J. (1)
Department of Pediatric Cardiology, Bonn, Germany (1); TIMUG, Bonn, Germany (2); TÜV München, Germany (3)

Introduction: In children with CHD, ventricular volume and function determine therapy and outcome. 3D-Real-Time-echocardiography (3DE) is a promising tool to assess ventricular volume and function. Unfortunately, the exactness of 3DE has not been validated for small structures as the neonatal or infant heart. Objective was to assess the accuracy of 2D- and 3D-generated distances and volume-measurements using phantoms.
Methods: The phantom CIRS555A for small distances und CIRC555 volumetric target phantom (6.9 ml) were used to assess the influence of data generation (transducer, frequency, focus, resolution) and data analysis (postprocessing) on volume and distance measurement. Inter- and intraobserver-variability were determined. Ultrasound-System: Sonos7500 Philips, Transducer: X4-1, S8, Curved array. Workingstations Q-Lab5.1 (Philips), MedCom.
Results: Distances: Inter- and intraobserver-variability was 1–2%. In all measurements, distances were determined significantly too small if they were positioned vertically to the transducer (−6%). Volumes: 2D- and 3D-generated volume measurements underestimated the true volume of the phantom significantly (2DE: 8–10% = −0.5–0.7 ml; 3DE 8%). 2D-generated volume calculations are dependent on measurement of distances and areas: therefore the vertical measuring error produced significant differences of the calculated volumes in dependency of the change of the position of the examined object ($p < 0.05$) (7% i.e. 0.5 ml). 3D-generated volume measurements were not dependent on the position of the examined object and showed a significant lower inter- and intraobserver-variability than 2D-calculated volumes (2D 6–10% = 0.4–0.7 ml; 3D 1–3% = 0.1–0.2 ml). Data postprocessing had significant influence on volume-determination (working station, change of contrast).
Conclusions: Compared to true values, 2D and 3D measurements underestimate distances and volumes, this has to be taken in account if these measures are compared to other techniques. 3D volume and distance measurements can be performed with low inter- and intraobserver-variability also in small objects. In contrast to 2DE, 3D volume measurements are independent on the position of the examined object and data acquisition shows minor influence on volume measurements.
Being aware of its limitations, 3D RT echo can be used as reliable tool for the non-invasive and serial assessment of small hearts and for decision making in neonates and infants with CHD.

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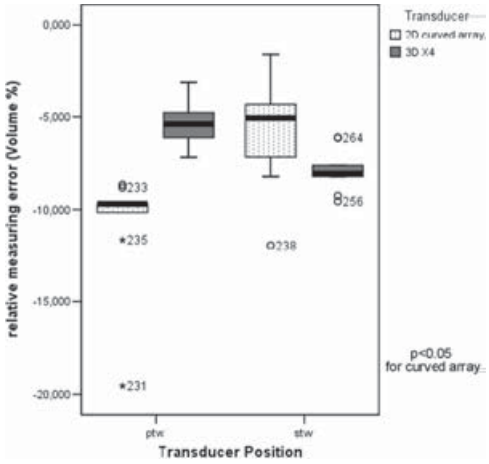

Figure: Relative error of volume measurement in 2D- and 3D-generated volumes dependent on tranducer position

O3-3
MRI Catheter Assessment of Total and Differential Pulmonary Vascular Resistance in Single Ventricle Physiology

Bell A. (1)(2), Greil G. (1)(2), Beerbaum P. (1)(2), Schaeffter T. (1), Razavi R. (1)(2)
Kings College London (1); Guy's and St Thomas' NHS Foundation Trust (2)

Introductions: In patients with single ventricle physiology, pulmonary blood flow may be derived from more than one source, making assessment of pulmonary vascular resistance (PVR) difficult by conventional methods. MRI Catheter combining phase contrast MRI (PC MRI) with invasive pressure measurement and has been shown accurately measure of the PVR. We demonstrate the application of MRI catheter to measure the total and differential PVR in this patient group.

Methods: MRI cardiac catheterisation was performed in a combined XMR suite with a Philips 1.5T Achieva MR scanner and a single plane cardiac X-Ray unit under general anaesthesia with ventilation to normocarbia. Invasive pressure measurements were obtained simultaneously with PC-MRI flows. Imaging planes were positioned according to the vascular anatomy. Differential pulmonary blood flow was measured distal to all sources of blood flow. For those patients with multiple sources of blood flow, PVR was calculated by the equation: $1/PVR_{total} = 1/PVR_{left} + 1/PVR_{right}$. Data is expressed as mean (±standard deviation), means are compared with a 2-tailed t-test.

Results: Between 2005 and 2007 MRI catheter with measurement of differential and total PVR was undertaken in 12 patients, median age 3.1 (0.8 to 41.7) Of these, seven patients had multiple sources of pulmonary blood flow. Patient details including diagnosis, differential PVR and outcome are described in table 1. Mean PVRtotal was 2.20 (±0.5) WU.m2, mean PVRright was 5.7 (±2.1) WU.m2 and a mean PVRleft 4.3 (±1.8) WU.m2. The difference between these two did not reach statistical significance (p = 0.08). 9 of the 12 patients were being investigated for suitability to progress to the next stage of fontan palliation. 5 have had successful surgery, 2 have been are awaiting surgery, and a further 2 had alternative palliative procedures to deal with concurrent problems.

Conclusion: Measurement of PVR in patients with complex single ventricle physiology can be undertaken with MR catheter. Preliminary surgical outcomes in this patient group have been favourable. MRI catheter offers the advantage of quantification of differential and total PVR, even when there is more than one source of pulmonary blood flow.

Table 1. Patient diagnosis, PVR and outcome

	Age (Years)	Diagnosis	Palliation	Pulmonary Blood Flow Sources
1	3.1	HLHS, Chylothoraces	TCPC	SVC, IVC
2	3.0	HLHS, Ascites	TCPC	SVC, IVC
3	12.5	DILV, TGA	SCPC	SVC
4	11.5	Right atrial isomerism, AVSD Pulmonary atresia, PLE	TCPC	SVC IVC
5	13.7	Mitral valve dysplasia Hypoplastic left ventricle, DORV	PA Band MBTS	MPA Shunt
6	1.5	Pulmonary atresia, intact ventricular septum	Pulmonary valvotomy Ductal stent	MPA Duct
7	41.7	DILV, TGA	Right MBTS Pulmonary valvotomy	MPA Shunt
8	15.9	DILV, TGA, Pulmonary stenosis Mitral regurgitation	None	MPA
9	1.1	Tricuspid atresia with VSD	PA Band	MPA
10	0.8	Left atrial isomerism, TAPVD AVSD with dominant left ventricle	Repair of TAPVD PA Band	MPA
11	1.9	HLHS Severe Tricuspid Regurgitation	SCPC	SVC
12	1.9	HLHS Severe Tricuspid Regurgitation	SCPC	SVC

	Age (Years)	PVR (WU.m^2)			Outcome
		Right	Left	Total	
1	3.1	4.4	3.6	2.0	Successful conservative management
2	3.0	6.8	5.0	2.9	Recreation atrial fenestration Resolution of ascites
3	12.5	3.3	4.3	1.9	Successful TCPC
4	11.5	9.3	1.9	1.6	Medical treatment PLE
5	13.7	6.9	2.7	1.9	Successful SCPC
6	1.5	6.0	3.3	2.1	Successful SCPC
7	41.7	5.5	3.2	2.0	Awaiting SCPC
8	15.9	7.3	4.9	3.0	Over sew mitral valve and PA Band
9	1.1	2.8	3.8	1.6	Successful SCPC
10	0.8	8.6	3.2	2.3	Successful SCPC
11	1.9	4.6	6.9	2.8	Awaiting SCPC
12	1.9	3.3	8.3	2.4	Repair tricuspid valve

Abbreviations: PVR–Pulmonary vascular resistance, HLHS–Hypoplastic left heart Syndrome, TCPC–Total cavopulmonary connection, SVC–superior vena cava, IVC–Inferior vena cava, MPA–Main pulmonary artery, DILV–Double inlet left ventricle, AVSD–atrioventricular septal defect, PLE–Protein losing eneteropathy, SCPC–Superior cavopulmonary connection, MBTS–Modified Blalock Taussig Shunt, TGA–Transposition great arteries, TAPVD–Total anomalous pulmonary venous drainage.

O3-4
The impact of pre-existent myocardial remodelling on the ventricular function after Tetralogy of Fallot repair

Farah M.C.K., Castro C.R.P., Moreira V.M., Guerra V.C., Binotto M.A., Riso A.A., Marcial M.B., Lopes A.A., Aiello V.D.
Heart Institute (InCor), University of São Paulo Medical School, Brazil

Introduction: We sought to investigate prospectively the histopathological myocardial remodelling in patients submitted to the correction of Fallot's Tetralogy, in order to detect possible factors associated to postoperative echocardiographic findings of systolic or diastolic ventricular dysfunction.

Patients and Methods: 23 consecutive Fallot patients (14 males), aged 12 to 186 months (mean = 39.6, median = 23 months) were enrolled in the study. Tissue Doppler ecocardiographic analysis (isovolumic acceleration-IVA, systolic myocardial velocity-S' and

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early diastolic myocardial velocity-E') was performed in three moments for both ventricles: before surgery, within the first three postoperative (PO) days and later, between the 30th and 90th PO days. During surgery, besides the anomalous infundibular bands resected, subendocardial biopsy samples from the right ventricular (RV) inflow tract and of the left ventricle (LV), through the ventricular septal defect, were obtained for histopathological morphometric evaluation: degree of cell hypertrophy, interstitial collagen (Sirius-red) and capillarity (immunohistochemistry against Factor-VIII). Troponin-T levels were measured before and after surgery. This study was approved by the Ethical Committee of our Institution.
Results: Troponin-T levels increased postoperatively in all patients (27.7 ± 18.6 ng/ml and 15.9+11.3 ng/ml - second and third PO days) and correlated positively with the cardiopulmonary bypass and cross clamping times ($p = 0.019$ and 0.018 respectively). IVA of the RV decreased significantly at the third echocardiographic evaluation ($p = 0.006$) and correlated negatively with the diameter of the RV cardiomyocytes from the inflow tract ($r = -0.59$; $p = 0.006$). E' measured at the RV free wall decreased significantly in both PO periods ($p < 0.001$) and showed a weak but significant negative correlation with the percentage of interstitial myocardial collagen ($r = -0.525$; $p = 0.044$). S' measured at the RV decreased significantly in the PO period but did not present any correlation with myocardial histopathological characteristics. The capillary area fraction did not differ among the biopsy samples analyzed. E' and S' measured at the LV did not change throughout the study.
Conclusions: Myocardial remodelling present preoperatively, as judged by the morphometric histopathological evaluation of cell hypertrophy and interstitial collagen, influenced respectively the medium term PO systolic and diastolic right ventricular function of repaired Fallot patients. Further studies are necessary to define its influence in long term follow-up.

O3-5
Correction of right ventricular loading conditions improves left ventricular systolic and diastolic function
Lurz P., Puranik R., Nordmeyer J., Khambadkone S., Frigiola A., Coats L., Marek I., Bonhoeffer P., Taylor A.M.
UCL Institute of Child Health and Great Ormond Street Hospital, London for Children, UK

Objectives: The Interdependences of right and left ventricular function is poorly understood, especially in the context of unfavourable right ventricular loading conditions. Left ventricular (LV) function has been shown to predict survival in patients with Tetralogy of Fallot and pulmonary valvar dysfunction. Here we assessed the impact of percutaneous pulmonary valve implantation (PPVI) on LV systolic and diastolic function.
Methods: In 40 patients with pulmonary stenosis/regurgitation and an indication for PPVI, change in LV volumes, ejection fraction and stroke volume were assessed on magnetic resonance (MR) before and with in one month post PPVI. The septal wall contour at end-diastole and end-systole was evaluated and quantified by calculation of the septal curvature on MR short axis images. LV filing parameters were determined using both mitral valve E/A ratios and tissue Doppler assessment of the lateral mitral annular velocities with echocardiography.
Results: Percutaneous pulmonary valve implantation led to a significant increase in indexed LV end-diastolic volume (68.9 ± 14.8 to 74.2 ± 15.3 ml/m2; $p < 0.001$) and stroke volume (41.9 ± 10.1 to 47.1 ± 9.9 ml/m2; $p < 0.001$). In addition, there was an improvement in LV ejection fraction (61.3 ± 9.2 to 64.2 ± 7.3%; $p < 0.001$). Relief of RV pressure and volume overload resulted in a more favourable septal shape as indicated by a higher curvature at end-diastole (0.129 ± 0.04 to 0.207 ± 0.06; $p < 0.001$) and end-systole (0.15 ± 0.09 to 0.26 ± 0.08; $p < 0.001$). On echocardiography, the mitral inflow E/A ratio increased form 1.9 to 2.1 ($p = 0.037$), the E to E' ratio from 9.7 ± 4.0 to 11.2 ± 3.4 ($p = 0.004$), indicating raised LV filling pressures.
Conclusion: PPVI resulted in a significant improvement in global LV function. Improvement in septal curvature indicates more favourable ventricular interaction post relief of RV pressure and volume overload.

O3-6
Assessment of Right Ventricular Function with the Pressure-Volume Conductance System after Surgery for Congenital Heart Disease – The Value of Volume
Uebing A. (1), Kramer H.-H. (1), Schlangen J. (1), Scheewe J. (1), Steendijk P. (2), Fischer G. (1)
Department of Paediatric Cardiology and Cardiovascular Surgery, University Hospital of Schleswig-Holstein, Campus Kiel, Kiel, Germany (1); Department of Cardiology, Leiden University Medical Center, Leiden, The Netherlands (2)

Introduction: In patients after repair of tetralogy of Fallot (ToF) or complete palliation of hypoplastic left heart syndrome (HLHS), the right ventricle (RV) is exposed to abnormal volume or pressure load. We aimed to assess RV function in both patient groups using load independent indices from pressure-volume loop analysis. Furthermore, we meant to relate load independent RV function (i.e. contractility) to RV end diastolic and end systolic volumes (RV EDV and RV ESV) as ventricular volumes and derivatives such as ejection fraction are commonly used to judge on ventricular performance.
Methods: 46 predominantly paediatric patients ($n = 28$ after repair of ToF; $n = 18$ after Fontan-type palliation of HLHS) were examined with the pressure-volume conductance system. In four, analysis of pressure-volume data failed for technical reasons. RV contractility was quantified using end systolic elastance (EES).
Results: In both patient groups EES correlated significantly with RV EDV and RV ESV index (ToF: $r = -0.64$, $P = 0.0003$ and $r = -0.64$, $P = 0.0005$, respectively; HLHS: $r = -0.75$, $P = 0.0005$ and -0.64, $P = 0.008$, respectively). Only in ToF patients, RV ejection fraction correlated with EES whereas in HLHS patients it did not (ToF: $r = 0.55$, $P < 0.01$; HLHS: $r = -0.2$, $P = NS$).
Conclusion: RV end diastolic and end systolic volumes are valid estimates of load independent RV function in patients with RV volume and pressure load after surgery for congenital heart disease. This result supports current clinical practice to use right ventricular volumes to guide on decision making in the follow up of these patients.

O4-1
20 years single center experience in pediatric heart transplantation: Long term results
Behnke-Hall K. , Bauer J., Thul J., Akintuerk H., Valeske K., Horn M., Bartram U., Kraemer U., Schranz D.
Children's heart center Giessen Germany

Introduction: Heart transplantation (Htx) is sometimes the last therapeutic option for children with end stage congenital or acquired heart disease. Since 1988 we have acquired 20 years of experience. New strategies in operative and drug management

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have been the major improvement for effecting the outcome. The long term results of our patients were analysed to find a clearer definition of causes of morbidity and mortality.
Methods: We reviewed the records of our pediatric transplant patients treated in our clinic between 1988 to 2008, with special interest in survival, mortality and major morbidity.
Results: In 20 years, we performed 134 heart transplantations. Five patients underwent retransplantation. 134 patients are currently in our program, including five patients transplanted in different centers. The majority of children that were transplanted had hypoplastic left heart syndrome (53) or cardiomyopathy (53). Age at time of transplant was mostly under one year (81). 25 patients died. 19 of those within the first year after transplant. Causes of death were graft failure (7), rejection (10) and others (8).
Probability of survival 10 years after transplantation is 78% for all patients, which shows a tremendous improvement from a five year survival rate of 71% (1988–1994) to 92% (2000–2006).
Substantial morbidity in long term course is acute rejection (171), hypertension (n = 86), chronic renal insufficiency (n = 13), malignancies (n = 11) and others. We treated 171 acute rejections in 85 patients. Chronic renal insufficiency is stable in 11 patients, while two underwent kidney transplantation. Malignancies consist of posttransplantation lymphoproliferative disease (8) and others (3). Rare, but apparent immunosuppressive therapy related diseases are found in seven patients: osteonecrosis, diabetes, sarcoidosis, unclassified myopathy, Evans-Syndrome (n = 2) autoimmunhemolytic anemia.
Conclusions: Morbidity in long term course is mostly caused by adverse effects of immunosuppressive therapy. Survival rate improved substantially in the last years and is now over 90% five years after transplant.
Hopefully in the future will be even more changes to a better outcome by focusing on morbidity and by instituting new immunosuppressive strategies.

O4-2
Results from the Swedish Prospective Screening Study in Newborns With Pulse Oximetry – 39 878 babies in the Region of Västra Götaland

de-Wahl Granelli A. (1), Mellander M. (1), Sunnegårdh J.(1), Sandberg K. (1), Wennergren M.(2), Eriksson M. (3), Segerdahl N. (3), Ågren A. (4), Ekman-Joelsson B-M. (4), Bejlum C. (5), L Inganäs L. (5), Östman-Smith I. (1)
The Queen Silvia Children's Hospital (1); The East Hospital (2); Göteborg; SÄS Borås (3); SKAS Skövde (4); NÄL Trollhättan (5), Sweden

Many babies with duct-dependent congenital heart disease (CCHD) are not detected by routine neonatal physical examination (PE) alone. Saturation screening has been proposed as an aid for early detection of CCHD, but so far the screening studies have been too small to enable a proper evaluation.
Purpose: To conduct a prospective screening study large enough to evaluate its contribution to early detection of CCHD, and compare with the detection-rate of PE alone.
Methods: Prospective screening of all babies in the 5 newborn nurseries in our region from September 2004 to March 31st 2007. A pulse oximetry reading was obtained from the right hand and one foot using identical new-generation oximeters (NGoxi), before the neonatal physical examination (PE). Our cut-off values were a saturation < 95% in both right hand and foot or a hand/foot difference > ± 3%. The protocol definition of screening-positive was: All babies with one optimal measurement ≤90%, and all babies with 3 repeated measurements below the cut-offs; all positives should be referred for echocardiography. Saturation protocols were blind to the doctor when saturations were > 90% until the findings on PE and any suspicion of CCHD were documented in a protocol. Exclusion criterion was admission to the neonatal intensive care unit (NICU).
Results: 39 878 newborns were included in our screening study and 31 of them had CCHD. PE detected 73% of the CCHD cases, versus 87% when combined with NGoxi-screening. 4 babies were missed on both PE and NGoxi-screening; 3 with Coarctation of the aorta and one with IAA. No baby with duct-dependent lung circulation or transposition was missed with NGoxi-screening. False-positive rate was 0.26%; among the "false" positive cases we found 13.5% with a ductus and/or foramen ovale, 11% infections, 11% other congenital heart defects and 9% with lung problems.
Conclusion: A pre- and postductal saturation screening with NGoxi before discharge from the newborn nurseries increased the detection of CCHD with 14% to 87%. The false-positive rate was low enough to cause only an average of 8 extra echocardiograms/unit and year (vs PE alone that generated 57) and included babies that benefited from early treatment or follow-up.

O4-3
Sudden death in children with cardiomyopathy: Results from a national population-based study of childhood cardiomyopathy

Bharucha T., Davis A.M., Turner C., Ramsay J., Robertson T., Daubeney P.E.F., Nugent A.W., King I., Weintraub R.W.
Royal Children's Hospital, Melbourne, Australia

Background: The incidence and risk factors for sudden death in children (SCD) with cardiomyopathy (CM) have not been well defined.
Methodology: The National Australian Childhood Cardiomyopathy Study is an ongoing cohort study, which includes all children in Australia with primary CM who were diagnosed at 0–10 years of age, between 1987–1997. Sudden cardiac death (SCD) was defined as a sudden and unexpected death in children who were not in congestive heart failure and were not hospitalised. For this analysis, subjects were included if they survived at least 30 days from presentation. Indexed echocardiographic measurements at latest follow-up were compared between surviving subjects and those with SCD.
Results: There were 264 children identified who met the study criteria. The mean duration of follow-up was 7.98 years for all subjects. The incidence of sudden death relative to each CM type, for all cases and as a proportion of deaths, is shown in the Table. SCD was significantly associated with CM type, for all cases (p = .003) and when only those subjects who died were considered (p = .05).

Table 1. Incidence of SCD by CM type.

	DCM (n = 151)	HCM (n = 79)	RCM (n = 7)	LVNC (n = 27)	Total (n = 264)
SCD	6	4	1	6	17
SCD as %age of all cases in CM type	3.9	5.1	14.3	22.2	6.3
SCD as %age of all deaths in CM type	15.4	26.7	50	54.5	24.6
Average annual rate of SCD	0.54	0.54	2.62	2.57	0.79

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In patients with dilated CM, subjects with SCD had a lower mean FS z score compared to survivors (−9.55 vs. −1.62) and a higher mean LVEDd z score (5.6 vs. 1.652; p = 0.0002 for both. Familial dilated CM was also associated with a higher risk of SCD (p = .0005). 10 additional subjects (8 with HCM and 2 with LVNC) underwent ICD implantation for primary prevention (3 cases) or symptoms and/or arrhythmias (7 cases).
Conclusions: This study defines the incidence of sudden death in children with CM. Subjects with restrictive cardiomyopathy or LV non-compaction, and those with dilated cardiomyopathy who have severe systolic dysfunction or a positive family history are at greatest risk of sudden death.

O4-4
Effect of medical therapy in childhood hypertrophic cardiomyopathy with risk factors for heart-failure related death
Östman-Smith I. (1), Keeton B. (2), Wettrell G. (3)
Division of Paediatric Cardiology, Queen Silvia Children´s Hospital, Gothenburg, Sweden (1); Wessex Cardiothoracic Centre, Southampton, United Kingdom (2); Division of Paediatric Cardiology, University Hospital, Lund, Sweden (3)

Introduction: About half of all childhood deaths in hypertrophic cardiomyopathy (HCM) are caused by congestive heart failure, but hitherto much research has focussed on sudden arrhytmia deaths. A recent cohort study identified separate risk factors for sudden arrhythmia death and heart-failure related death. This study was undertaken to evaluate the effect of therapy on patients at high risk for heart-failure (CCF) related deaths.
Methods: From a complete geographical cohort of 132 HCM patients diagnosed in life below 19 years of age the patients with identified risk factors for heart-failure deaths (left ventricular wall-to-cavity ratio (LVCAVR) > 0.30, CCF-symptoms below 2 years of age) were identified. 72 patients fulfilled these criteria, with median age at diagnosis 1.2 years (interquartile range 0.4–16.7 years), LVCAVR 0.41 (0.31–0.49), and total follow-up 8.8 (3.4–15.4) years; 35 had associated Noonan or Leopard syndrome (49%). Four therapeutic strategies could be identified: No therapy or diuretics (NST; n = 14), conventional dose beta-blockers (1–4 mg propranolol equivalents/kg BW; n = 20; CDBB), high-dose betablocker (>4.5 mg/kg propranolol; n = 31; HDBB) and calcium-channel blockers (verapamil 2.0–12.0 mg/kg; n = 8; CCBL). There were no significant differences in risk factors between the different treatment groups.
Results: There were a total of 17 CCF-deaths, but they were very unequally distributed between treatment groups: CCBL group had 75% CCF-mortality, NST-group 43% CCF-mortality, CDBB-group 20% CCF-mortality, and HDBB-group 6.5% CCF-mortality. Comparisons with Fisher´s exact test shows that the CCBL-group has significantly worse survival than the total group (p = 0.006) with odds ratio (OR) for CCF-death of 9.7 (95%CI 1.8–52.6), and worse survival than HDBB (p = 0.0002), OR = 43.5 (5.1–373) or CDBB (p = 0.0089), OR = 12.8 (1.8–64.6). HDBB-group also had significantly better survival than the NST-group (p = 0.007), OR = 0.09 (0.02–0.55). A notable feature of the CCBL-group was a high proportion of late CCF-deaths, 67% of CCF-deaths as compared with 17% in the NST-group, 25% in the CDBB-group, and 0% in the HDBB-group.
Conclusions: Calcium-blocker therapy is deleterious in HCM-patients at high risk for CCF i.e. with generalized left ventricular hypertrophy, and should be avoided in such patients. Beta-blocker therapy reduces mortality in CCF in such patients, with a high-dose regime appearing to confer most benefit.

O4-5
Sildenafil in the management of the failing Fontan circulation
Reinhardt Z. (1), Bhole V. (1), Uzun O. (2), Wright J. (1), Linter K. (1), Onuzo O. (2), Wilson D. (2). Stumper O. (1)
The Heart Unit, Birmingham Children's Hospital – NHS Foundation Trust, Birmingham, UK (1); Department of Paediatric Cardiology University Hospital Wales, Cardiff, UK (2)

Background: Sildenafil is increasingly being used in the management of pulmonary arterial hypertension. Its role in patients with congenital heart diseases is less well defined and as yet has only been sporadic.
Aim: Present our experience with Sildenafil treatment in patients with a failing Fontan circulation.
Patients and methods: Retrospective review of 12 symptomatic patients after a Fontan procedure who received treatment with Sildenafil.
Results: Three patients suffered from protein losing enteropathy (PLE), three patients presented with bronchial casts, two had severe desaturations post fenestrated Fontan procedure, two had prolonged chylous effusions, one had previous failure of Fontan and take-down and one had arrhythmias and end-stage failure and required conversion to an extracardiac Fontan.
Sildenafil was used in the dosage of 15 to 25 mg three to four times per day.
PLE and alpha1-antitrypsin levels improved in all 3 patients on Sildenafil treatment. One of these patients had concomitant catheter creation of a fenestration, as did 2 patients presenting with bronchial casts and both patients with persistent chylous effusions. All 3 patients with bronchial casts improved significantly on Sildenafil treatment. Chylous effusions decreased after Sildenafil and stent enlargement of fenestration. In the 2 patients with severe post-op cyanosis there was an improvement of resting and exercise saturations. Sildenafil was used in 2 patients perioperatively in the creation/conversion of an extracardiac Fontan with good effect. There were no significant side effects requiring Sildenafil withdrawal over a treatment period ranging from 2 months to 1.4 years. One patient died from end stage CHF. Sildenafil was electively stopped after optimizing haemodynamics in 4 patients with recurrence of PLE in 1.
Conclusions: Sildenafil can be used safely and effectively in the treatment of patients with a failing Fontan circulation. In this study it was mainly used as an adjunct to catheter or surgical therapies addressing haemodynamic lesions. Our initial observations suggest that randomised controlled trials of Sildenafil treatment in children with a Fontan circulation should be undertaken.

O4-6
Cardiac function and outcome of patients with infantile Pompe's disease treated with recombinant human alpha-glucosidase (rhGAA) in Germany – a retrospective survey
Schmidt D. (1), Schranz D. (1), Hagel K.J. (1), Hennermann J.B. (3), Smitka M. (4), Tarusinov G. (5), Marquardt T. (6), Haase C. (7), Trübel H. (8), Motz R. (9), Fröhlich G. (10), Neubauer B.A. (2), Kampmann C. (11), Mengel E. (11), Hahn A. (2)
Pediatric Heart Center, University Gießen, Germany (1); Department of Neuropediatrics, University Gießen, Germany (2); Department of Pediatrics, Charité Campus Virchow-Klinikum, Berlin, Gemany (3); Children's Hospital, University Dresden, Germany (4); Pediatric Heart Center, Duisburg, Germany (5); Department of Pediatrics, University Hospital Münster, Germany (6); Department of Pediatrics, University

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Hospital Jena (7); Department of pediatric cardiology, Children's Hospital Wuppertal University of Herdecke , Germany (8); Department of pediatric cardiology, Children's Hospital Oldenburg, Germany (9); Children's Hospital, Memmingen, Germany (10); Children's Hospital, University of Mainz, Germany (11)

In infantile Pompe disease, virtually absent activity of lysosomal acid a-glucosidase leads to progressive accumulation of glycogen in the heart and skeletal muscles. Rapidly worsening hypertrophic cardiomyopathy results in cardiac failure and death usually within the first year of life. First clinical studies showed that enzyme replacement therapy (ERT) can reduce cardiac hypertrophy and prolong survival, but data about the effects on cardiac function and outcome of treated individuals are limited.
Methods: We analyzed retrospectively the clinical charts, echocardiographic data, and laboratory parameters of 13 German patients receiving ERT.
Results: Age at onset of first symptoms was 1.5 ± 1.0 months, age at diagnosis 3.8 ± 3.0, and age at start of ERT 4.9 ± 4.1 months. 4 out of 13 subjects (31%) died during ERT. Mean age at death was 18.6 ± 6.3 months while the mean age of the surviving patients is 34.5 ± 24.1 months. Before start of ERT, all patients had hypertrophic cardiomyopathy as assessed by left-ventricular mass index (LVMI), or IVS- and LVPW-thickness. In 7 out of 8 subjects, for whom serial echocardiographic data were available, a significant reduction of cardiac hypertrophy with age was attained.
LVMI completely normalized in 4 patients. In one subject shortening fraction remained substantially reduced (< 20%) although LVMI normalized.
One individual deceased after 72 weeks of treatment due to progressive cardiac hypertrophy despite augmentation of recommended rhGAA dosage.
Brain-natriuretic peptide (BNP) determined in three individuals was massively elevated (>2000 ng/l) in two of them, normalized during therapy (< 50 ng/l), and reflected amelioration of cardiac function earlier than echocardiography.
Conclusion: These findings confirm prolonged survival in infants with Pompe disease receiving ERT, and demonstrate that significant improvement of cardiac function can be achieved in most, but not in all patients. BNP may be a valuable parameter for surveillance of cardiac function during ERT.

O5-1
Radiofrequency ablation in infants

Sarquella Brugada G., Torner S., Esquirol M., Prada F., Mortera C., Bartrons J., Rissech M., Jimenez L., Carretero J., Brugada J.
Arrhythmia Unit, Cardiology Section, Hospital Sant Joan de Déu, Barcelona, Spain

Objective: The objective of this study was to compare the safety and efficacy of radiofrequency ablation in infants (< 15 kg) versus the rest of the paediatric population.
Methods: Between 1992 and 2007, radiofrequency catheter ablation was performed to 800 patients ≤18 years in a single institution and by a single operator. Among them, 55 were infants (weight ≤15 kg) and 745 children (>15 kg). Analysis of efficacy, complication rate and procedure characteristics were analyzed and compared between both groups.
Results: Mean weight was 9.2 ± 4.4 kg (range 1.5–15) in infants and 45.7 ± 15.1 kg (range 17.2–100) in children. Mean age was 1.3 ± 1.1 years (range 5 days – 4 years) in infants and 12.7 ± 3.4 years in children. Efficacy was 94% in infants and 95% in children (p = ns). Recurrence rate was 6% and 5% respectively (ns). One major complication – mitral valve regurgitation that required surgical plasty - occurred in an infant and 4 minor complications (0.3%) occurred in children (ns). Only 1 catheter was used in 80% of the infants, and 2 catheters were used in 83% of children. Duration of the procedure (51 ± 27 vs. 52.8 ± 38 minutes) and radiation exposure (10.9 ± 8 vs. 12.7 ± 11.1 minutes) were similar in both groups. In Infants the most frequent diagnostic was orthodromic tachycardia (30%), incessant supraventricular tachycardia due to a slowly conducting accessory pathway (22%) and auricular tachycardia (19%). In children the diagnostics were WPW (50%), orthodromic tachycardia (19%) and AV nodal reentrant tachycardia (15%).
Conclusions: Radiofrequency ablation can be performed successfully and safely in an infant population. In experienced hands, the technique can be performed with low radiation exposure times, few catheters, few complications and low recurrence rates.

O5-2
Catheter ablation of common supraventricular tachycardia substrates in children in the era of non-fluoroscopic navigation

Papagiannis J., Tsoutsinos A.I., Kourkoveli P.E., Kantzis M., Rammos S.
Onassis Cardiac Surgery Center, Athens, Greece

Introduction: Catheter ablation of supraventricular tachycardia (SVT) substrates is highly successful, but in the past required significant exposure to radiation. This study was undertaken to assess the effect of a non-fluoroscopic navigation (NFN) system in the outcome of catheter ablation of common SVT substrates.
Methods: 146 patients (ages:12.4 yrs average, 11yr median and 3yrs min value) underwent 156 catheter ablation procedures using the Nav-X Ensite system over a period of 4 years. The tachycardia substrates were: accessory pathway (AP) (n = 94), and AV nodal reentrant tachycardia (AVNRT) (n = 52). Thirty two patients with AVNRT underwent cryoablation, 2 pts underwent RF and cryoablation whereas the rest 21 pts underwent radiofrequency ablation. The effect of the learning curve of the NFN system was assessed by comparing the first half of the study period with the second half. A p value < 0.05 was considered statistically significant.
Results: The success rate was high and similar in both periods: 100% vs. 99% for AVNRT and 97.5% vs. 96.8% for AP. The recurrence rate was also similar in both periods: 15% vs. 11.4% for AVNRT and 13.5% vs. 17.4% for AP. The fluoroscopy time, procedure time, and recurrence rate according to tachycardia substrate and study period is shown in the table. There is a significant reduction in fluoroscopy time for both groups of patients, without any significant effect on the procedure time.

	First half of study period			Second half of study period		
AVNRT	20 pts			32 pts		
		mean ± SD	range	mean ± SD	range	
	Fluoro time (min)	9.15 ± 4.8	2.3–22	4.13 ± 2.7	0.8–12	P:0.022
	Proc.time (hrs)	2.7 ± 0.67	1.5–4.5	2.48 ± 0.53	1.5–3.5	p:0.126
AP	35 pts			59 pts		
		mean ± SD	range	mean ± SD	range	
	Fluoro time (min)	9.3 ± 6	1.9–28.8	7.25- ± 6.8	0.4–33	p:0.001
	Proc.time (hrs)	3.1 ± 1.2	1.5–7	2.6 ± 0.8	1.5–5	p:0.144

Abbreviations: fluoro: Fluoroscopy time; Proc. Procedure; Rec.: Recurrence

Conclusions: The use of a non-fluoroscopic navigation system for catheter ablation of common SVT substrates has lead to a significant reduction in exposure to radiation, without compromising the

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efficacy and safety of the procedure. A learning curve exists in the use of the NFN system.

O5-3
Reproducibility of Left Ventricular Dyssynchrony Evaluation by Echocardiographic 2D Strain in the Young

Janousek J. (1), Razek V. (1), Salameh A. (1), Gebauer R.A. (2)
Department of Pediatric Cardiology, University of Leipzig, Heart Center, Leipzig, Germany (1); Kardiocentrum and Cardiovascular Research Center, University Hospital Motol, Prague, Czech Republic (2)

Introduction: Speckle tracking derived 2D strain may be used for evaluation of segmental ventricular contraction timing and mechanical dyssynchrony and be helpful in the indication of CRT. Reproducibility of this technique has, however, not been validated in young patients.
Methods: 20 patients aged median 4.6 (0.4–25.7) yrs with either right ventricular (RV, N = 13) or left ventricular (LV, N = 7) pacing for complete atrioventricular block reflecting 2 different types of LV mechanical activation sequence were evaluated by speckle tracking-derived 2D strain. Time to peak systolic longitudinal strain (SL) was assessed in apical 4-chamber and long axis views in 4 basal and 4 mid-ventricular segments, time to peak systolic radial (SR) and circumferential strain (SC) in 6 segments in the parasternal short axis view. Intra-observer reproducibility of measurements performed in 2 consecutive cardiac cycles was assessed.
Results: Of the 78 recorded single cycle loops 7 (9.0%) were primarily rejected because of low grey scale image quality or rate. Reliable measurements could be obtained in 255/312 (81.7%) of SL, 161/192 (83.6%) of SR and 170/192 (88.5%) of SC segments analyzed (p NS) resulting in 108, 70 and 78 cycle pairs available for evaluation, resp.. Correlation between measurement 1 and 2 increased from $R2 = 0.809$ ($p < 0.001$) for SL to $R2 = 0.814$ ($p < 0.001$) for SR and $R2 = 0.835$ ($p < 0.001$) for SC. Bland-Altmann plot revealed 6/108 (5.6%) of paired SL, 5/70 (7.1%) of SR and 7/78 (9.0%) of SC measurements being outside of the ± 2SD range (p NS). Reproducibility of time to peak SR decreased with LV ejection fraction ($R2 = 0.434$, $p = 0.008$). Reproducibility of dyssynchrony indices (maximum difference and standard deviation of time to peak segmental strain) was lowest for SL ($R2 = 0.286$, $p = 0.018$ and $R2 = 0.397$, $p = 0.004$, resp.) and highest for SC ($R2 = 0.730$, $p < 0.001$ and $R2 = 0.735$, $p < 0.001$, resp.).
Conclusions: Measurement of time to peak systolic segmental SC is the most reproducible method for evaluation of mechanical LV dyssynchrony in the young. SR measurements are negatively influenced by low ejection fraction and reproducibility of SL derived dyssynchrony indices is low. (R.A.G. was supported by grant NR/9472-3 of the Ministry of Health, Czech Republic)

O5-4
Low Rate of inappropriate Shocks in paediatric Patients with ICD by preventing Strategies for Tachycardias and individual Device Programming

Botsch M., Franzbach B., Krings G., Opgen-Rhein B., Will J. C., Berger F.
Departement of paediatric cardiology and congenital heart failure, Charité Medical Centre, Berlin, Germany

Introduction: Recent studies in pediatric patients with implantable cardioverter defibrillators (ICD) report a high percentage of inappropriate therapy up to 50% of delivered shocks, which lead to reduction of quality-of-life and in individual cases to the rejection of the therapy.
Methods: The data of all of our patients (pts) with previously implanted ICD were retrospectively analyzed for cardiac disorders, ICDs, leads, programming, anti-arrhythmic drugs, ablation procedures and device discharges.
Results: In total 33 pts underwent ICD implantation between January 2001 and November 2007. The underlying cardiac disorders included congenital heart disease (36%), primary electrical disease (27%), cardiomyopathy (24%) and idiopathic ventricular tachycardia (12%). Implant indications were fast VT (55%), documented VF (12%), syncope (18%), resuscitation (9%) and others (6%). The mean age at implantation was 16.5 years (range 8-36). During operation there were no major complications. In all pts the approach was transvenous. Dual chamber ICDs were implanted in 19, biventricular ICDs in 2 and single chamber ICDs in 12 pts. Medication after implantation was mainly beta-blocker (60%), Amiodarone (18%) and none (15%). Ablation procedure underwent 9 pts due to atrial flutter or ventricular foci. In all pts the programmed VF-zone covered the life-threatening ventricular tachycardia, additional zones were programmed in 16 pts. In total 63 shocks (range 1–34) were delivered in 11 pts (33%) during a cumulative follow up of 972 months (range 1–78). All therapies were delivered for tachycardias. There were 61 shocks (97%) appropriate for documented VT/VF. Only two shocks (3%) were inappropriate, each of which caused by atrial flutter: in a pt with LQTS with a single chamber ICD and in a pt with Brugada-syndrome with a dual chamber ICD. During follow up 3 pts died due to progressive heart failure, none of the deaths were primarily related to the arrhythmia.
Conclusion: In children and young adults ICD therapy is effective in prevention of life threatening arrhythmias. The combination of prevention strategies for tachyarrhythmias by specific drug therapy and ablation procedures and individual Device programming cause a low incidence of inappropriate shock delivery.

O5-5
Left Ventricular Apical Pacing in The Young: Preserved Synchrony and Left Ventricular Function over Mid-Term Follow-Up

Janousek J. (1), Gebauer R.A (2), Tomek V. (2), Razek V (1), Gebauer R. (2), Matejka T. (2), Kostelka M. (3)
Department of Pediatric Cardiology, University of Leipzig, Heart Center, Leipzig, Germany (1); Kardiocentrum and Cardiovascular Research Center, University Hospital Motol, Prague, Czech Republic (2); Department of Cardiac Surgery, University of Leipzig, Heart Center, Leipzig, Germany (3)

Introduction: RV epicardial pacing is frequently used in young children because of limited venous access to the heart but may lead to LV mechanical dyssynchrony, remodeling and failure. Alternative pacing sites have not been well evaluated yet. We thought to evaluate left ventricular (LV) function in epicardial LV apical as compared to right ventricular (RV) epicardial pacing the young.
Methods: LV synchrony and function was evaluated in 16 consecutive pediatric patients with LV apical pacing for complete atrioventricular (AV) block and compared to 12 RV epicardial paced controls matched for age at implantation (median 5.7 vs. 1.2 months, NS) and block etiology (congenital/surgical: 7/9 vs. 6/6 patients, NS).

Results: LV paced patients were superior to their RV paced counterparts in terms of LV synchrony (interventricular mechanical delay: mean 7 ± 21 vs. 51 ± 21 ms, p < 0.001; septal to posterior wall motion delay: median 0 vs. 70 ms, p = 0.003, maximum difference in time to peak segmental longitudinal LV strain: 73 ± 24 vs. 124 ± 39 ms, p < 0.001, standard deviation of time to peak segmental longitudinal LV strain: 26 ± 8 vs. 47 ± 14 ms, p < 0.001) and LV function (shortening fraction: 40 ± 7 vs. 25 ± 11%, p < 0.001; ejection fraction: 58 ± 8 vs. 42 ± 14%, p < 0.001 and LV end-systolic volume index: 22 ± 17 vs. 34 ± 30 mL/m2 BSA, p = 0.004). Although follow-up was longer in the RV paced group (median 61.5 vs. 22.2 months, p = 0.002), none of the dyssynchrony indices correlated significantly with the duration of pacing. Pacing site (p < 0.001) but not the duration of pacing (p NS) was the most significant multivariate predictor of LV ejection fraction.
Conclusions: LV apical pacing is superior to RV epicardial pacing in terms of preservation of LV synchrony and function and should be preferred in the young. (R.A.G. was supported by grant NR/9472-3 of the Ministry of Health, Czech Republic)

O5-6
Mapping of Intraatrial Reentrant Tachycardias by Remote Magnetic Navigation in Patients with D-Transposition of the Great Arteries after the Mustard or Senning Procedure

Pflaumer A. (1), Wu J.(2), Deisenhofer I. (2), Ücer E. (2), Estner H. (2), Hess J. (1), Zrenner B. (2), Hessling G. (2)
Pediatric Cardiology (1) and Cardiology (2), Deutsches Herzzentrum Muenchen, Klinik an der TUM, Munich, Germany

Introduction: Mapping of Intraatrial Reentrant Tachycardias (IART) still presents a challenge in complex congenital heart disease. Acess to both atria is necessary in IART after atrial switch operation in those patients. Remote magnetic navigation (RMN) combined with a 3-dimensional (3D) electroanatomical mapping system may improve safety and reduce fluoroscopy times in this population. We present our initial experience with RMN for mapping of intraatrial reentrant tachycardia.
Methods: 3D mapping of the systemic venous atrium (SVA) and the pulmonary venous atrium (PVA) was performed using RMN (Niobe®) in conjunction with 3D mapping (Carto RMT®) in 4 patients after the atrial switch procedure (Mustard n = 1, Senning n = 3) for D-Transposition of the Great Arteries. The maps were fused with a CT-based 3D anatomy and ablation was performed using this map.
Results: All patients had cavotricuspid- isthmus dependent IART with a mean atrial cycle length of 305ms. Mapping of both atria (PVA retrogradely by passing the aortic and tricuspid valve) was feasible and safe. The procedure time for IART mapping ranged from 210–320 minutes (min) with a mean of 251 min. The fluoroscopy time for IART mapping ranged from 16.8 to 45.0 min (mean 30.3 min) for patients, and ranged from 12.3 to 19.2 min for physicians. No procedural complications occurred.
Conclusions: Precise mapping of IART in the complex anatomical structures after an atrial switch procedure was feasible and safe using RMN. The maneuverability of the catheter was possible even with a retrograde access crossing two valves. Further reduction of procedural and fluoroscopy times for both patients and physicians seems possible.

No. Patients	Procedure time (minutes)			Fluoroscopy time (minutes)						
	Mapping	Ablation	Total	T1	T2	T3	T4	T5	T(pt)	T(phys)
1	265	200	490	7.3	0.5	5.0	3.0	2.5	19.3	14.8
2	320	30	370	7.2	8.4	14.8	7.8	1.0	40.1	23.0
3	210	160	435	7.2	2.0	12.0	6.3	7.2	27.5	26.4
4	210	222	450	8.3	3.7	16.0	17.0	16.0	71.1	40.3
mean	251	153	436	7.5	3.7	11.2	8.5	6.7	39.3	29.8

Table 1: Procedural Data: T1 = the fluoroscopy time for puncture and catheter placement; T2 = the fluoroscopy time for SVA mapping; T3 = the fluoroscopy time for retrograde access to RV, T4 = the fluoroscopy time for PVA mapping; T5 = the fluoroscopy time for ablation. T (pat) = T(1+2+3+4+5): the total fluoroscopy time for the patient; T (phys) = T(1+3+5): the total fluoroscopy time for the physician.

O6-1
Closure of atrial septal defects with the Solysafe occluder

Daehnert I. (2), Djukić M. (1), Parezanović V. (1), Vukomanović G. (1), Ilisić T. (1), Stefanović I. (1), Bernhard J. (3), Jovanović I. (1)
University Childrens Hospital, Belgrade, Serbia (1); Herzzentrum, University of Leipzig, Germany (2); CARAG AG Medical technology, Baar, Switzerland (3)

Introduction: To report early and intermediate results of a prospective mono-center study with the Solysafe Septal Occluder, a new self-centering device for transcatheter closure of atrial septal defects (ASD). The initial European experience with this device presented at last years AEPC meeting in Warsaw [Ewert P, et al. Cardiol Young 2007; 17(Suppl 1):18–19] is currently in press [Ewert P, et al. ASD and PFO closure with the Solysafe Septal Occluder - results of a prospective multicenter pilot study. Catheter Cardiovasc Interv 2008]. This was the second trial investigating this device with a slightly modified design including two larger additional device sizes and studying exclusively ASD.
Methods: Prospective single-center study. Interventions were performed under general anaesthesia. Stretched ASD diameter was measured by conventional sizing procedure. Device placement was guided by transesophageal echocardiography and fluoroscopy. Follow-up visits were performed pre-discharge and 1, 3, 6 and 12 months after implantation. 52 patients with a median age of 10 years (range 4–53) underwent interventional ASD closure with Solysafe devices. Median size of stretched defects: 17mm (range 7–30). Median procedure time: 37min (range 24–111). Median fluoroscopy time: 9.1min (range 3.5–24.1). Devices used: 9 size 15, 18 size 20, 12 size 25, 9 size 30 and 4 size 35.
Results: There were no intra-procedural complications. One day after implantation one patient showed device migration to the pulmonary artery. Device removal and ASD closure performed surgically revealed a non-existing inferior rim of the defect at a distance of 10mm, excluded damage of intraatrial structures and proved the explanted device still correctly configured and without damage. All other patients were discharged one day after intervention, none had residual shunt. Early closure rate was 51/52. Follow-up is three months in 17, six months in 15 and one year in 19 patients and was uneventful in all. Late occlusion rate is 100% (51/51).
Conclusions: With the self-centering Solysafe Septal Occluder, ASD with stretched diameters up to 30 mm can effectively be closed with very high occlusion rates. As in all other devices, sufficient rims are needed for stable device anchoring and errors of judgement can result in early device migration.

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O6-2

Dilatable pulmonary artery band in low-weight or complex congenital heart disease neonates allows to avoid or postpone subsequent surgery

Gewillig M., Rega F., Boshoff D.E., Eyskens B., Mertens L., Delhaas T., Meyns B.
Paediatric Cardiology & Cardiac Surgery, University Hospitals Leuven, Belgium

Objectives: Banding of the pulmonary artery PA remains good early palliation in low-weight or complex congenital heart disease neonates. A fixed band needs to be removed surgically when the patient outgrows his pulmonary blood flow. We evaluated the efficiency and safety of dilatable bands to avoid or postpone further surgery.
Methods: 2 types of "handmade" bands were evaluated:
1/ pulmonary trunk: non-resorbable nylon 2mm cord around PA, overlay side-by-side at desired tension, sewn with polypropylene 6/0; ends of nylon cord folded together and fixed with 3medium titanium clips; band fixed to trunk with 2separate polypropylene 5/0 stitches; this design allows "dilatable" restriction;
2/ branch PA during hybrid procedure (ductal stenting & bilateral banding): open ring of 3.0–4.0 mm Gore-Tex reclosed with one 7/0 polypropylene stitch; this design for branch PA allows "breakable" band.
Balloon dilation was done with high-pressure non compliant balloons, starting with small size until desired result.
Patients & Results: 17 patients (mean birth weight 2.5 kg (0.35–4.5) were divided in 4 groups:
Group 1: 4pts: large VSD or complex CHD requiring additional surgery; progressive dilation (2 balloons, max 8mm) after 22 (3–59) weeks allowed to postpone further surgery;
Group 2:8 pts: large VSD & coarctation (4), spontaneous restriction of VSD; band became redundant and was blown away after 44 (7–91)weeks with a 10–16 mm balloon; 2 patients required later VSD closure;
Group 3: 4 pts: hybrid procedure for complex CHD with hypoplastic or interrupted arch. In 1pt (BW 1.6 kg) dilation after 8.7 weeks with 3.5 mm balloon resulted in high flow, requiring rebanding; in 3 other patients balloon dilation with 3.5–4.0 mm balloon after 10–19 with good result;
Group 4:1pt: neonatal balloon dilation AS & hybrid procedure because borderline LV; adequate growth and function of LV after 31 weeks; successful closure duct and balloon dilation 8 mm of bands as final procedure.
Conclusions: A dilatable band is an attractive technique. The "dilatable" design with staples allows for predictable progressive dilation from early on; in this series the "breakable" designs for branch PA resulted in unrestrictive flow when dilated < 9 weeks; after 10 weeks sufficient restriction persisted if desired.

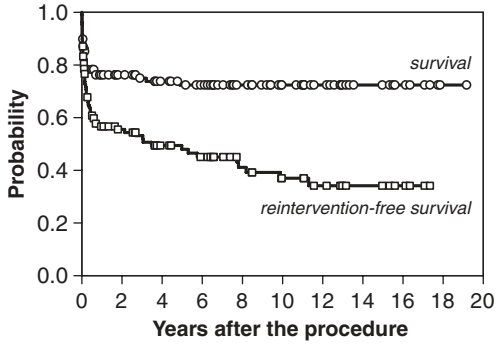
O6-3

Long term (up to 19 years) results of balloon valvuloplasty of critical neonatal aortic stenosis

Reich O.(1,2), Tax P.(1), Tomek V.(1), Gilík J.(1), Škovránek J.(1,2), Chaloupecký V.(1)
Kardiocentrum and Cardiovascular Research Centre, University Hospital Motol (1); Department of Paediatrics, 2nd Medical School, Charles University (2), Prague, Czech Republic

The purpose of this study was to assess the efficacy, long-term results, and predictors of outcome of balloon valvuloplasty of critical neonatal aortic stenosis

Methods: The procedure was performed in 108 consecutive newborns aged 0 to 28 (median 2) days. The indication for treatment was peak Doppler gradient > 75 mmHg in 64 (59%) and duct-dependent systemic circulation and/or left ventricular dysfunction in 44 (41%) patients. The balloon to annulus diameter ratio was 0.98 ± 0.09 (± SD). Follow up ranged up to 19.2 years (median 4.5; in survivors 7.4 years).
Results: Twenty eight patients (26%) died, 15 of them (14%) early after the procedure. Reinterventions on the aortic valve were necessary in 39 patients (36%): repeated valvuloplasty in 9 patients (8%) and surgery in 30 (28%), in 10 of those after a previous revalvuloplasty. Actuarial probability of survival 19 years after the valvuloplasty was 72.4 ± 4.5% and of reintervention-free survival 17 years after the procedure 34.2 ± 5.9% (± SE). By multiple logistic regression, independent predictor of death was procedure indication based on duct-dependent systemic circulation and/or left ventricular dysfunction (odds ratio 4.67, 95% bounds 2.43 to 8.98, $p < 0.001$); independent predictors of death or need for reintervention were the duct-dependent systemic circulation and/or left ventricular dysfunction (odds ratio 2.53, 95% bounds 1.43 to 4.48, $p < 0.001$) and aortic annulus Z-score (odds ratio 0.26, 95% bounds 0.09 to 0.79, $p < 0.001$)
Conclusions: Balloon valvuloplasty is capable of saving more than 70% newborns with critical aortic stenosis, however only one third of the newborns will survive childhood without further aortic valve interventions. Duct-dependent systemic circulation along with the left ventricular dysfunction and a small aortic annulus predict unfavourable outcome of the treatment.
Supported by grant VZ00064203-6302



O6-4

Hybrid Closure of Muscular Ventricular Septal Defects in Children; Initial Experience

Thanopoulos B.D., Kourtesis A., Paphitis Ch., Eleftherakis N., Azariadis G.,
"Aghia Sophia" Children's Hospital, Athens, Greece

Introduction: In spite recent advances in percutaneous techniques device closure of muscular ventricular septal defects (MVSDs) in small infants has significant limitations. The aim of this study was to report initial experience with hybrid (intraoperative closure) closure of MVSDs in 11 consecutive patients using the Amplatzer MVSD occluder.
Methods: The age of the patients ranged from 1–12 months and the weight from 2.8–8 Kg. After a median sternotomy the right ventricle was punctured and an introducer sheath was advanced through the MVSD over a wire into the left ventricle. The occluder was advanced through the sheath and it was deployed under transesophageal echo guidance.
Results: The MVSD diameter ranged from 7–10 mm. The device size varied from 8–12 mm. The procedure was successful in

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all patients. In one patient with an associated perimembranous defect the MVSD was closed using a periatrial approach. Transient ventricular tachycardia during the procedure was observed in two very small infants. No other early or late major complications occurred. At the end of a 2-year follow-up complete echocardiographic closure was observed in 10/11 (91%) of the patients.
Conclusions: Hybrid closure of MVSDs using the Amplatzer MVSD occluder it is safe and effective alternative to surgical closure for small infants with MVSDs. Further studies are required to document its efficacy, safety and long-term results in a larger patient population.

O6-5
Single- Centre Experience Of Stenting The Ductus Arteriosus Using Three Different Types Of Stents
Tzifa A., Tsapakis E., Rosenthal E., Qureshi S.A.
Evelina Children's Hospital, London, UK

Objective: Stenting of the arterial duct has been established as an alternative palliative approach to maintain ductal patency. We sought to assess the technical difficulties, complication rate and long term outcome of three different types of stents used in our institution over the past 15 years for stenting of the arterial duct in patients with duct-dependent pulmonary flow.
Methods: Retrospective review of the medical records of 30 consecutive patients who underwent cardiac catheterization for stenting of the arterial duct using Tower, Corinthian and Liberte stents between 1991 and 2006. The patients were grouped according to the type of stent implanted. Procedural details, complications and short- and long term outcome were recorded.
Results: Thirty patients had attempted PDA stenting using Tower (group I, n = 15), Corinthian (group II, n = 9) or Liberte stents (group III, n = 6). PDA stenting was successful in 24/30 patients (10/15 of group I, 8/9 of group II and 6/6 of group III). Catheter or surgical reinterventions to maintain ductal patency were required only in patients from group I (n = 4). Complication rate was 60% for group I (6/10 patients), 50% for group II (4/8 patients) and 16% for group III (1/6 patients). 3/10 Tower (30%), 5/8 Corinthian (62.5%) and 6 Liberte stents (100%) are still patent at a median of 138, 72 and 12 months, respectively.
Conclusion: The availability of lower profile, more flexible stents with better longitudinal and radial strength has led to higher success rates and better procedural and clinical outcomes in patients who undergo arterial duct stenting.

O6-6
Catheter Closure of Fontan Fenestrations – A Caution
Stumper O., Bhole V., Ofoe V., deGiovanni J.G., Miller P.A., Dhillon R., Wright J.G.C.
The Heart Unit, Birmingham Children's Hospital - NHS Foundation Trust, Birmingham, UK

Objective: To review the indications and the outcome of transcatheter closure of fenestrations after the Fontan procedure.
Background: Fenestrations of the Fontan circuit are frequently created to improve postoperative outcome and recovery. The resultant right-to-left shunt leads to chronic desaturations and poses a risk factor for systemic thromboembolic events. Catheter closure of persistent fenestrations has been advocated but leads to a reduction in cardiac output. In patients with late Fontan failure de-novo catheter fenestration has proved to be beneficial in most patients.
Patients and Methods: Between 1/1988 and 7/2004 406 patients underwent a modified Fontan procedure at a median age of 4.7 yrs (IQR 3.8–7.1). Fenestrations were performed in 216pts (53%). Early mortality was 4.4% (n = 18), 4 patients required takedown (1%). Late mortality was 5.4% (21/388) , late takedown 1 , Htx 3 patients.
Results: 37/216 patients (16.2%) underwent transcatheter closure of the Fontan fenestration. Indications for closure were resting saturations < 85% in air and further reduction in saturations with minor exercise. Temporary test occlusions were performed and device occlusion was performed if the right atrial pressure remained < 18 mm Hg during occlusion.
Complete device occlusion was carried out in 24, custom-made partial occlusion devices were used in 12 and one patient had closure by implantation of covered stents. There were no procedural complications.
During the follow-up there were 4 serious complications (10.8%). Two patients developed protein losing enteropathy 6 weeks and 3 years after device occlusion – one of these required de-novo re-fenestration with significant improvement. One patient developed bronchial casts 4 months after occlusion and one patient died from end-stage congestive heart failure and low output 6 months post procedure.
Conclusion: We experienced a high rate of serious complications after transcatheter closure of Fontan fenestrations in the context of very conservative indications (resting saturations < 85%). In severely cyanosed patients after a fenestrated Fontan procedure meticulous haemodynamic assessments have to be carried out. Partial occluder devices should be preferred. In the context of progressive late post-operative desaturation in the setting of a stable fenestration, alternative treatment modalities suchas a trial of pulmonary vasodilator should be considered.

O7-1
Contraception and contraceptive counselling for women with congenital cardiac disease – still an unresolved problem?
Kaemmerer H. (1), Vigl M.(2), Bauer U. (2), Niggemeyer E. (2), Nagdyman N. (3), Rattenhuber J. (4), Trigas V.(1), Kaemmerer M. (1), Seifert-Klauss V. (4), Schneider KTM.(4), Hess J. (1), Berger F. (3)
Deutsches Herzzentrum München, Munich, Germany (1); Competence Network Congenital Heart Defects, Berlin, Germany (2); Deutsches Herzzentrum Berlin, Berlin, Germany (3); Frauenklinik der Technischen Universität München, Munich, Germany (4)

Introduction: The number of women with congenital cardiac disease entering sexual maturity is rising steadily. Advice on pregnancy related risks and contraception has to consider cardiologic and gynaecological aspects and is of vital importance in this patient group.
Methods: Questionnaire-based observational survey over 12 months in two tertiary care centres, including 540 adult female patients with a median age of 29 years [18–75]. The study population was grouped into functional classes according to Perloff (functional class I: 51.0%; II: 40.8%; III/IV: 7.2%).
Results: Only 56.9% of the patients received information concerning methods and risks of contraceptive methods and in only 56.3% of these cases the issue was addressed by the treating physician without being asked. Out of the 65 women (12.0%) considered to be at the highest risk in case of pregnancy, 19 (32.2%) did not get any information on contraception. 8.0% of all patients became pregnant despite using a contraceptive method. A total of 173 out of the 540 patients (32.0%) were regarded to have a contraindication against the use of combined oral contraceptives. Despite of this, 34 patients (19.6%) are using this method for

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contraception at the moment. The median age at first sexual intercourse was 17.0 years, which is similar to the general German population (17.5 years) and makes a timely counselling necessary.
Conclusions: Despite the eventual and - in some groups - life threatening risks in case of pregnancy, knowledge and counselling on contraception is poor in this patient group. Women with congenital cardiac disease could profit from pharmacological and technical advancements in contraceptive methods and an effective and suitable contraception can be found in almost all cases.

O7-2
Pregnancy After Atrial Switch Operation For Transposition Of The Great Arteries
Trigas V. (1), Bauer U. (2), Oechslin E. (3), Nagdymann N. (2), Rattenhuber J. (4), Seifert-Klauss V. (4), Schneider K. T.M. Deutsches Herzzentrum München (1); Deutsches Herzzentrum Berlin (2), Universitätsspital Zürich (3); Frauenklinik der Technischen Universität München (4), Stern (1), Berger F. (2), Hess J. (1), Kaemmerer H. (1) Deutsches Herzzentrum München (1), Deutsches Herzzentrum Berlin (2), Universitätsspital Zürich (3), Frauenklinik der Technischen Universität München (4)

Objectives: Due to improved treatment and surgical options, the number of women with congenital cardiac disease reaching reproductive age has dramatically increased in recent decades.
This study looks into the course of pregnancy after atrial switch operation (ASO) in patients with transposition of the great arteries (TGA).
Methods: In total, data on 60 pregnancies in 34 women (pat.) were collected. Patient age at first pregnancy ranged from 16 to 34 (median: 25) years, at last pregnancy from 19 to 35 (median: 27) years. Before the first pregnancy, all women were in a functional class I or II. The median interval between ASO and first pregnancy was 23 years (range: 14–20 years), between first pregnancy and last postpartum follow-up 4.7 years (range: 0.5–17 years).
Results: In total, 16 abortions were observed, 11 of which were spontaneous. Abruptions occurred five times due to medical or social reasons. 43 pregnancies were carried to full term. In 16 cases, delivery happened spontaneously, while vaginal surgical delivery took place in 4 cases and cesarean section in 23 cases.
In 6 patients (18%), the FC deteriorated during pregnancy. Relevant pregnancy related complications included progressive dyspnoe (n = 4), cardiac decompensation (n = 1), and obstetrical complications (n = 21). After the last pregnancy, 23 patients were still in FC I, 8 were in FC II and 2 were in FC III; one patient deceased.
In two cases, resuscitative measures had to be taken during/after the delivery.
12 children had a birth weight < 2500 g. There was no congenital cardiac defect in any of the offsprings.
Conclusions: TGA after ASO leads to serious risks for both mother and child. Due to this, patients have to be informed about potential pregnancy-related risks early.
A close monitoring of the course of pregnancy by cardiologists and obstetricians is mandatory; these must have specific experience in this field and be well acquainted with the hemodynamics of the underlying disease.

O7-3
Pulmonary Artery Growth and Pressure late after Fontan Operation
Ovrutskiy S., Ewert P., Alexi-Meskishvili V., Hölscher K., Miera O., Peters B., Hetzer R., Berger F.
German Heart Institution Berlin, Germany

Objective: We evaluated the growth of the central and lower lobe pulmonary arteries (PA) and the development of the pulmonary artery pressure (PAP) after Fontan operation.
Methods: Forty children in whom Fontan operation (extracardiac conduit in 22 and lateral tunnel in 18) was performed at median age of 3.9 (1.5–13.3) years underwent angiographic measurements of the central and lower lobe PA diameter and the mean PAP analysis before Fontan operation and during the follow-up (median 4.7; range 1.3–15.1 years). The median patient age at follow-up was 8.8 (range 3.2–28.6) years. Body surface area (BSA)-dependent PA index (PAI) and lower lobe index (LLI) were calculated and values obtained before Fontan operation and at follow-up were compared.
Results: Median BSA was 0.60 preoperatively and increased to 0.89 during the follow-up ($p < 0.001$). While parallel somatic development of the children in percentile terms was documented by BSA measurements, the PA showed no gain in diameter at all (median RPA diameter of 9 mm preoperatively vs. 10.5 mm post-OP, $p = 0.32$ and median LPA diameter of 9.7 mm preoperatively vs. 10.0 post-OP, $p = 0.33$). Consequently, the BSA-dependent PAI und LLI (preoperative median 242 and 138 mm2/m2, respectively) decreased highly significantly over the follow-up period (median 179 and 112 mm2/m2, respectively, $P < 0.001$). No significant changes in PAP were observed between pre- and postoperative (median 10.5 mmHg vs. 12 mmHg, p = n.s.). The lowest PAI was noted in patients who had the longest follow-up ($P = (0.029)$). We found a correlation between a low PAI and unfavourable Fontan hemodynamic, especially in association with PAP > 15 mmHg ($n = 12$, $p = 0.004$).
Conclusions: Growth and development of the pulmonary arteries after Fontan operation are clearly reduced despite somatic growth. This phenomenon, especially combined with elevated pulmonary artery pressure may lead to an increase in pulmonary vascular resistance and limit optimal Fontan circulation in children over a long time period.

O7-4
Modifications of cardiopulmonary capacity are not associated with changes in quality of life in patients with congenital heart disease
Ehlert N., Hess J., Hager A.
Department of Paediatric Cardiology and Congenital Heart Disease, Deutsches Herzzentrum München, Technical University Munich, Germany

Objective: The intention of surgical and medical treatment is to improve the cardiovascular status and exercise ability. However, do improvements of exercise ability also result in a better quality of life?
Patients and Methods: 194 patients (85 female, 109 male, age 14–81 years) with various congenital heart defects were investigated twice. At each time, they completed the SF-36 quality of life survey and, afterwards, they performed a symptom-limited cardiopulmonary exercise test. In-between the two tests, 48 patients had a change in their medicational regime, 34 open heart surgery, 10 a catheter intervention, and 102 had no changes in their medical management for at least 6 month.
Results: Patients were classified into 3 groups. 47 patients showed an increase in peak oxygen uptake for more than 10%, in 88 patients it remained similar (± 10%) and in 53 patients it declined for more than 10%.
Comparing the 3 groups, there were no significant changes in any of the scales of quality of life. Even in self-reporting physical functioning, there was no difference between those three groups. (Kruskal Wallis Test p = .909).
Conclusions: Improvements or worsening of physical exercise ability do not automatically result in an improvement or worsening of quality of life and have to be evaluated and analyzed separately.

Patients with congenital heart disease do not recognize objectively measurable improvements or worsenings of their physical capacity in their subjective evaluation.

O7-5

The awareness of illness and quality of life in adolescents after corrective surgery for congenital heart disease

Maryniak A., Ziolkowska L., Ondruch A., Bielawska A., Kawalec W.
The Children's Memorial Health Institute, Warsaw, Poland

Background: After surgery for congenital heart disease (CHD) many patients suffer from residual defects and possible impairment of their quality of life (QoL). The aim of our study was to assess, how adolescents after surgery evaluate their illness, current ailments and QoL.
Methods: We analyzed 232 pts, aged 16–19 yrs, after surgery for CHD (mainly VSD, ASD, TOF, CoA). A questionnaire comprising of open questions about their own heart disease, experienced ailments, doctor's orders, limitations in style of life was used. QoL was assessed using the SF-36v2 health survey. It measured 8 health domains: 1) physical functioning; 2) role limitations due to physical health; 3) role limitations due to emotional problems; 4) vitality; 5) emotional state; 6) social functioning; 7) pain; 8) general health perceptions. The range of 0 to 100 points may be assigned to each domain – the higher the score means the better QoL.
Results: About 24% of the adolescents were not capable of describing properly their heart disease, they did not know their diagnosis. 30% of pts suffered from clinical symptoms, 38% reported serious limitations in their style of life, though the majority of them did not specify what those limitations referred to. 8% of pts declared no need of further medical control. Results obtained from SF-36 questionnaire indicate that the patients did not perceive significant negative impact of their heart disease on physical, emotional and social functioning (73-80 points). However, they evaluated their general state of health as rather poor (53), likewise emotional state (62) and vitality (62). There was statistically significant correlation ($p < 0.009$) between the presence of clinical symptoms and QoL - patients reported ailments had significantly lower outcomes in all eight domains. These results suggest dominant impact of experienced ailments on the quality of life evaluation.
Conclusions: The majority of analyzed adolescents after surgery for CHD experience cardiac symptoms, but their knowledge about disease and its impact on their activity was poor. This points to the necessity of special medical and psychological care for those patients. Emphasis should be put on preparing adolescents to take over responsibility of their health condition.

O7-6

Long term follow up of adolescents and adults (GUCH) after modified Fontan operation

Ovrutskiy S., Ewert P., Miera O., Alexi-Meskishvili V., Peters B, Hetzer R., Berger F.
German Heart Insitute Berlin, Berlin, Germany

Introduction: Since the introduction of single ventricle palliation more patients reach adulthood and need specialized supervision and therapy. We analyzed hemodynamics, physical performance and morbidity in GUCH patients after modified Fontan operation.
Methods: Fifty-three patients, who underwent Fontan operation between 9/91 and 9/2007, reached adolescent (15–18 years) or adult age (Range: 15–47, Median 27 years) during the median follow up of 8 (Range: 0.6–15) years. Sixteen of them were operated on as adults (16–37 years). The intraatrial modification was performed in 38 patients and extracardiac Fontan operation (ECFO) in 15. The hemodynamics was analyzed by heart catheterization and magnet resonance image. The cardiopulmonary capacity was controlled by spiroergometry with monitoring of the exercise (Wmax) and oxygen consumption capacity (VO2max). Further, necessity for cardiac medication and the incidence of arrhythmias were checked.
Results: There were three late deaths (mortality 5.6%). Patients who underwent heart catheterization ($n = 33$) showed stable Fontan hemodynamics with low pulmonary artery pressure (median 11 mmHg) and low transpulmonary gradient (median 6 mmHg). The cardiac index, measured by MRI was in median good with 2.6 l/min/m², but in adolescent better as in adults (3.1 vs. 2.4 l/min/m², $p = 0.027$). Wmax and VO2max decreased significantly during the follow up (W/kg: 2.1 ± 0.6 (64.9 ± 16.2% of healthy coevals) early postoperatively vs. 1.6 ± 0.7 W/kg (49.3 ± 23.3%) late postoperatively, $p = 0.001$ and VO2max of 26.5 ± 7.7 ml/min/kg (59.8 ± 14.9%) early vs. 21.8 ± 8.3 ml/kg/min (52.8 ± 17.7%) late, $p = 0.005$). 12 patients (22%) developed tachyarrhythmias. 11 patients, all after intraatrial Fontan operation, required a permanent pacemaker due to bradyarrhythmias. Medical treatment of heart failure was necessary in 28 patients (53%). All patients were on an anticoagulation regime. No clinically relevant thromboses were noted but one thromboembolic event occurred in one patient after intracardiac Fontan.
Conclusions: Grown up patients after lateral tunnel or extracardiac Fontan operation reach adulthood with stable hemodynamics and low morbidity. The incidence of arrhythmias after ECFO is lower. Regular checks of the patients' physical exercise capacity and hemodynamics are necessary to optimize the cardiac medication for progressive heart failure and to identify candidates for later heart transplantation.

O8-1

Prevalence of Congenital Heart Disease (CHD) in Newborns in Germany: Preliminary Data of the PAN Study

Lindinger A. (1), Schwedler G. (2), Olchvary J. (1), Hense H.W. (3)
Clinic of Pediatric Cardiology, University of the Saarland, Homburg, Germany (1); Competence Network of Congenital Heart Defects (2); Institute of Epidemiology & Social Medicine, University Muenster (3)

Introduction: The PAN study is a nationwide investigation of the prevalence of CHD in Germany. Particular attention is paid to the impact of gender, maternal age, gestational age and birth weight of the newborns as potential determinants for CHD.
Methods: Prospective study design with a total study period from July 2006 to June 2008. Data were provided by a total of 259 participating institutions: 34 departments of pediatric cardiology, 155 children´s hospitals and 70 outpatient departments. Diagnostic method was echocardiography in nearly all cases.
Results: A total of 6133 infants with CHD born 07/2006 – 06/2007 were entered into the database. The preliminary CHD prevalence for this study period was calculated as 0.9%.
A ventricular septal defect (all types) was the most common lesion (50.6%), followed by atrial septal defects (14.2%), pulmonary stenoses (5.7%), aortic coarctation (3.3%) and aortic stenoses (2.0%). The most common cyanotic lesions were tetralogy of Fallot (2.5%) and transpositions of the great arteries (2.5%). A single ventricle (all types) was identified in 3.0% with half of them being hypoplastic left heart syndrome.
Of all cases, 57% were diagnosed within the first month of life and 86% within the first three months. Female gender was slightly more common (53.1%) with a predominance in `minor´ (57.0%)

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and 'moderate' (51.4%) cardiovascular defects while a striking predominance of male infants was seen in the more complex lesions (59.3%).
A prenatal diagnosis was reported for 32% of all CHD cases, and pre- and postnatal diagnoses were identical in 90%. A birth weight below 1500g or 2500g, respectively, were both associated with a threefold incidence of CHD, and the incidence was doubled for multiple pregnancies.
Conclusion: First preliminary data from the PAN survey seem to confirm prevalence rates of about 1% of CHD. Diagnosis is made early in life in Germany; however, the rate of prenatal diagnosis is still fairly low. CHD is clearly more common in small for date infants and multiple pregnancies.
This work is supported by the Competence Network for Congenital Heart Defects funded by the Federal Ministry of Education and Research, FKZ 01GI0601.

O8-2

Mid term outcome of patients with pulmonary atresia, ventricular septal defect, pulmonary arteries hypoplasia and major aortopulmonary collaterals after a combined multistage strategy of anatomical repair

Dragulescu A., Ghez O., Kreitmann B., Chetaille P., Metras D., Fraisse A.
Timone Children's Hospital, Marseille, France

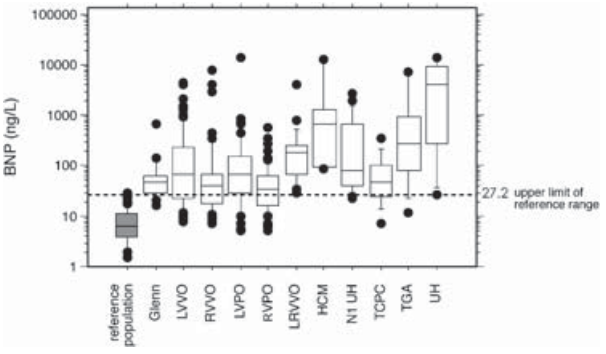
Objectives: To report the mid-term results of pulmonary artery rehabilitation in pulmonary atresia-ventricular septal defect (PAVSD), hypoplastic pulmonary arteries (PAs) and major aortopulmonary collaterals (MAPCA's).
Patients and Methods: Since 1990, 19 patients (11 males) with PAVSD, profound PAs hypoplasia (mean Nakata index was $27.5 \pm 16.7\ mm^2/m^2$) and MAPCA's benefited from the medico-surgical strategy of PAs rehabilitation which includes 3 steps:
1. Surgical RV-PA connexion early in infancy to favor antegrade flow and to promote angiogenesis.
2. Interventional catheterization for PA dilation and MAPCAs occlusion.
3. Surgical repair with PA reconstruction, VSD closure and valved RV-PA conduit
The ultimate goal is to achieve biventricular, anatomical repair with satisfactory hemodynamics (RV to aortic pressure ratio < 0,8).
Results: The initial RV-PA connection was performed at a median age of 5 (0.1–25.2) months with one operative death and was followed after a median time of 4.3 (1.2–15.1) months by the 2nd step of interventional catheterizations (mean: 2.3 ± 1 per patient). Thirty-two PA angioplasties were performed with 10 stent implantation and 14 MAPCAs were occluded. Significant PAs growth was obtained in all the cases with a mean Nakata index of $209.5 \pm 90\ mm^2/m^2$ ($p < 0.001$) after the 2nd step. Surgical repair was performed at a median age of 2.2 (0.6–10.3) years with elective fenestration of the VSD patch in 4 patients. During the mean follow-up of 8 ± 4.2 years, PAs rehabilitation was pursued in 11 patients with residual PA stenosis or MAPCAs. Twenty-six PA angioplasties were performed with 7 stents implantation and 9 residual MAPCAs were occluded. Three patients with poor hemodynamic results and RV dysfunction died, of which one after RV-PA homograft replacement. At last visit, the 15 survivors are in NYHA class I ($n = 11$) or II ($n = 4$) with a satisfactory hemodynamic result in 11 cases.
Conclusion: Medico-surgical PAs rehabilitation represents an aggressive management but is successful in the majority of the cases in this very complex and difficult-to-treat group of patients. Late deaths may occur, often in patients with a dysfunctional and hypertensive right ventricle. Precise right ventricular function assessment seems mandatory during the patient's long term follow-up.

O8-3

Diagnostic Accuracy and Clinical Relevance of BNP Assay in Pediatric Patients with Congenital Heart Disease

Cantinotti M., Vittorini S., Storti S., Prontera C, Vellani A., Ait Ali L., De Lucia V.
G. Monasterio Foundation and Institute of Clinical Physiology-National Research Council, "G. Pasquinucci" Hospital, Massa, Italy.

Introduction: Although B-type natriuretic peptide (BNP) assay is recommended for the diagnostic evaluation of heart failure, its clinical usefulness in pediatric cardiology is still controversial. We evaluated the diagnostic accuracy of BNP assay in congenital heart disease (CHD), and its clinical relevance in relation to different cardiovascular haemodynamics in pediatric patients.
Methods: BNP measurement was performed in 173 healthy subjects (HS) and in 357 consecutive patients with CHD. We identified 5 major subgroups of haemodynamical conditions: left ventricle (LV) volume overload (LVVO) (DORV, mitral insufficiency, PDA, VSD, truncus arteriosus), right ventricle (RV) volume overload (RVVO) (anomalous pulmonary venous return, ASD, tricuspid insufficiency), LV pressure overload (LVPO) (aortic coarctation, aortic stenosis), RV pressure overload (RVPO) (DORV and aortic stenosis, palliated pulmonary atresia and VSD, pulmonary stenosis, TOF, VSD with pulmonary hypertension or pulmonary stenosis), biventricular volume overload (LRVVO) (atrio-ventricular septal defect). Patients with Glenn, hypertrophic cardiomyopathy (HCM), cavo-pulmonary connection (TCPC), univentricular heart (UH) univentricular heart after first stage of Norwood palliation (N1 UH), transposition of the great arteries (TGA) were considered as separate groups. Logarithmic transformation of data was used for statistical analysis with parametric tests (ANOVA and Scheffè analysis).
Results: BNP was higher ($p < 0.0001$) in CHD patients (median 52.0 ng/L, range 0.45-13932 ng/L), considered as a whole, than in reference population (median of 6.1 ng/L, range 1.0–29.7 ng/L). BNP assay showed a good diagnostic accuracy by ROC analysis in discriminating between HS and CHD patients (AUC 0.954, SE 0.008). Lower BNP values were found in RV pressure overload (median 34.0 ng/L, range 5.0-567 ng/L) than in LV volume overload (median 64.6 ng/L, range 8.0–4300 ng/L), LV pressure overload (median 64.0 ng/L, range 0.45–13729 ng/L), or biventricular volume overload (median 171.7 ng/L, range 28.8–4012 ng/L). LRVVO vs. RVPO $p = 0.0312$; TGA vs. RVVO $p = 0.0369$; HCM vs. RVPO $p = 0.009$; $p < 0.001$ for: UH vs. LVVO, UH vs. RVVO, UH vs. LVPO, TGA vs. RVPO, UH vs. RVPO.
Conclusions: BNP assay is a useful diagnostic tool in pediatric CHD patients. Lower BNP mean values were found in conditions of right ventricle pressure overload than left ventricle volume or pressure overload and biventricular volume overload.



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O8-4

Identification of high-risk patients among children with hypertrophic cardiomyopathy

Ziolkowska L., Kawalec W., Turska-Kmiec A., Pregowska K., Boruc A., Kowalczyk M.
The Children's Memorial Health Institute, Warsaw, Poland

Background: Hypertrophic cardiomyopathy (HCM) is the commonest cause of sudden cardiac death (SCD) in children. The aim of study was to evaluate the risk factors for SCD in children with HCM hospitalized in our institution.
Methods: Retrospective analysis of 76 pts, mean age 10.7 ± 5.1 yrs with HCM diagnosed from 1991 to 2007. Patients demographics, clinical symptoms, family history of HCM and SCD as well as the results of echocardiography, ECG, 24-hour ECG, chest X-ray, SPECT, ambulatory BP monitoring, exercise test were examined. Data from all patients have been analyzed regarding the presence of major and minor risk factors for SCD according to ACC/AHA/ESC 2006 recommendations.
Results: The age at diagnosis of HCM ranged from 1 month to 16 yrs, mean 6.1 ± 5.6 yrs, in 21 pts HCM was diagnosed before the age of 1 yr. Screening for familial HCM revealed 34 (45%) positive cases, of whom 79% had a first degree affected family member.
The major risk factors for SCD were present in 31 (41%) pts: cardiac arrest in 3 (4%) pts, sustained VT in none of pts, family history of SCD in 15 (20%), syncope in 10 (13%), LV thickness ≥30 mm in 3 (4%), abnormal exercise BP in 13 (17%) and nonsustained VT in 8 (11%) pts. Among 31 children with the major risk factors, 17 pts had one, 11 children had two (in 2 pts ICD was implanted as a primary prevention), 2 pts had three (ICD was implanted as a secondary prevention) and 1 pt had five risk factors (ICD was implanted as a secondary prevention). All children are treated with β-blockers, 5 pts underwent surgical myectomy. The minor risk factors such as myocardial ischemia was found in 40 (53%) children, LVOTO in 22 (29%) pts, competitive sports in 4 (5%) pts. The atrial fibrillation was not found in our group of patients.
Conclusions: (1) The major risk factors for SCD were present in 41% of children, of whom 14 (18%) had ≥ two risk factors. (2) High-risk patients with HCM ought to be prospectively identified and therapy should be considered even if they don't have symptoms.

O8-5

Effect on quality of life and other subjective parameters of a novel formulation of bosentan in children with pulmonary arterial hypertension: FUTURE-1 study

Berger R.M.F. (1), Beghetti M. (2), Barst R.J. (3), Jais X. (4), Acar P. (5), Fraisse A. (6), Ivy D. (7), Schulze-Neick I. (8), Bonnet D. (9), Galie N. (10), Haworth S.G. (8)
Beatrix Children's Hospital, Groningen, The Netherlands (1); University Hospital of Geneva, Geneva, Switzerland (2); New York Presbyterian Hospital, New York NY, United States (3); Hôpital Antoine-Béclère, Clamart, France (4); Hôpital des Enfants, Toulouse, France (5); Hôpital de la Timone, Marseille, France (6); University of Colorado/Children's Hospital, Denver CO, United States (7); Great Ormond Street Hospital for Sick Children, London, UK (8); Necker Hospital for Sick Children, Paris, France (9); University of Bologna, Bologna, Italy (10)

Objectives: FUTURE-1 primarily evaluated the pharmacokinetics (PK) of a new paediatric oral formulation of bosentan in children with pulmonary arterial hypertension (PAH) and compared it with that of adult PAH patients from a previous study. Exposure after 2 and 4 mg/kg BID dosing was found to be similar indicating non-linear PK, and bosentan was well tolerated (Beghetti et al, ESC 2007). Here, results on quality of life (QoL), WHO functional class (FC), and Global Clinical Impression are reported.
Methods: This single arm, prospective, multicentre study enrolled children (2–12 years) with idiopathic PAH (IPAH) or familial PAH (FPAH). Patients were treated with the new bosentan oral, dispersible, quadrisected, paediatric formulation at 2 mg/kg BID for 4 weeks, then 4 mg/kg BID until week 12. Exploratory endpoints were changes in SF-10 health-related QoL survey, WHO functional class (FC), and Parent's and Physician's Global Clinical Impression.
Results: In total, 36 patients were enrolled: 31 IPAH/5 FPAH; 21 male; mean age 6.8 years; mean weight 22.3 kg; WHO FC II/III: 64%/36%. 1 patient was excluded due to protocol violation. Mean changes from baseline to Week 12 in the SF-10 Physical and Psychological summary scores were similar, with a trend for improvement. WHO FC improved in 5 patients, remained stable in 19 patients, and worsened in 1 patient. Changes in Global Clinical Impressions are shown in Table 1.

Table 1: Global Clinical Impression

	n	significantly better	better	unchanged	worse	significantly worse
Parents	35	7	11	13	1	2
Physicians	35	2	13	18	1	1

Conclusions: In the majority of patients the use of this new paediatric formulation of bosentan was associated with an improved or stable clinical condition, as consistently observed in SF-10 evaluation, WHO FC analysis, and Parent's and Physician's Global Clinical Impression assessments.

O8-6

Increased risk for congenital heart defects after periconception exposure to medicines and low dietary nicotinamide in nicotinamide N-methyltransferase polymorphism carriers

van Driel L.M.J.W. (1,2), Smedts H.P.M. (1), Helbing W.A. (2), Isaacs A. (3), Lindemans J. (4), Uitterlinden A.G. (4,5,6), van Duijn C.M. (6), de Vries J.H.M. (7), Steegers E.A.P. (1), Steegers-Theunissen R.P.M. (1,2,3,6).
Obstetrics and Gynaecology/Division of Obstetrics and Prenatal Medicine, Erasmus Medical Centre, Rotterdam, The Netherlands (1); Paediatric Cardiology, Erasmus Medical Centre, Rotterdam, The Netherlands (2); Clinical Genetics, Erasmus Medical Centre, Rotterdam, The Netherlands (3); Clinical Chemistry, Erasmus Medical Centre, Rotterdam, the Netherlands (4); Internal Medicine, Erasmus Medical Centre, Rotterdam, the Netherlands (5); Epidemiology, Erasmus Medical Centre, Rotterdam, the Netherlands (6); Human Nutrition, Wageningen University, Wageningen, the Netherlands (7).

Introduction: Genetic and nutritional factors in the homocysteine and detoxification pathways play a role in the aetiology of congenital heart defects (CHDs). The recently identified nicotinamide N-methyl transferase (NNMT) gene and its substrate nicotinamide are implicated in both pathways. Our objective was to determine CHD risk in association with the NNMT G/A genotype, maternal periconception medicine use and/or dietary nicotinamide intake.
Methods: A case-control study of 292 children with complex CHD and 316 nonmalformed children and both parents was conducted in the Western part of the Netherlands. CHD phenotypes comprised of tetralogy of Fallot (n = 30), transposition of the great arteries (n = 51), atrioventricular septal defect (n = 31), perimembranous ventricular septal defect (n = 81), coarctation of the aorta (n = 28), aortic valve stenosis (n = 6), pulmonary valve stenosis (n = 51) and

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hypoplastic left heart syndrome (n = 16). At the study moment of approximately 16 months after the index-pregnancy, mothers filled out standardized questionnaires on periconception medicine use and a food frequency questionnaire on dietary nicotinamide intake. Mothers, fathers and children were genotyped for the NNMT G/A polymorphism (rs694539). The data were analyzed by univariate and multivariate logistic regression analysis using the dominant model.
Results: Periconception medicine use and a low dietary intake of nicotinamide (≤13.8 mg) was associated with CHD risk, (OR (95%CI) 1.5 (1.0–2.2) and 1.6 (1.1–2.3), respectively. No significant association was found between the NNMT AG/AA genotypes and CHD risk in mothers (0.9 (0.7–1.3)), fathers (1.1 (0.8–1.6)), and children (1.1 (0.8–1.6)). However, periconception medicine use, nicotinamide intake, and the NNMT AG/AA genotype in mothers or children showed risk estimates up to 2.2 (0.8–5.7) and 4.0 (1.3-12.0), respectively.
Conclusions: The NNMT A-allele of the mother or child additionally contributes to CHD risk in combination with periconception exposure to medicines and/or low dietary nicotinamide intake. These findings provide new insights into the complex aetiology of CHDs, and may be important for future preconception counselling in order to prevent CHDs.

O9-1
FoxO3 mediates hU-II-induced MMP-2 expression in pulmonary artery smooth muscle cells: Potential role in pulmonary hypertension

Diebold I., Petry A., Burger M., Hess J., Görlach A.
Experimental Pediatric Cardiology, Dept. of Pediatric Cardiology and Congenital Heart Diseases, German Heart Center Munich at the TU Munich, Munich, Germany

Pulmonary artery vasoconstriction and vascular remodelling are major contributors of pulmonary hypertension (PH). However, the mechanisms linking these events are not well understood.
Human urotensin-II (hU-II) is a newly identified vasoactive peptide which by binding to a G-protein-coupled-receptor, termed GPR-14, acts as the most potent vasoconstrictor known. Although hU-II has been associated with vascular remodelling in PH, its precise role in regulating remodelling processes is unclear. Since degradation of ECM plays a pivotal role in vascular remodelling, we investigated whether hU-II is linked to expression and activity of matrix metalloproteinases (MMP) in pulmonary artery smooth muscle cells (PASMC).
By immunohistochemistry (IHC) we found high levels of GPR-14 and MMP2 in the media and intima of pulmonary vessels in tissue samples from patients with pulmonary vascular disease and PH. In vitro experiments confirmed that hU-II time-dependently enhanced MMP2 mRNA and protein levels in PASMC. Actinomycin D inhibited hU-II-stimulated MMP2 expression indicating the involvement of a transcriptional mechanism. Bioinformatic analysis of the MMP2 promoter revealed the presence of a putative consensus site for Forkhead transcription factor O 3 (FoxO3). FoxO3 belongs to the family of FoxO transcription factors and is implicated in cell differentiation and migration. We could demonstrate by IHC that FoxO3 is present in the media of remodelled pulmonary arteries. Therefore we investigated whether there is a link between hU-II and FoxO3. hU-II increased FoxO3 mRNA and protein in PASMCs. In addition reporter gene assays confirmed that hU-II increases activity of FoxO transcription factors. Using an siRNA approach we found that downregulation of FoxO3 inhibited hU-II-induced MMP2 expression. Conversely, overexpression of wild type or constitutively active FoxO3 increased MMP2 expression. Furthermore overexpression of FoxO3 significantly increased MMP2 promotor activity, whereas mutation of the MMP2-promotor at the FoxO3 binding site completely abolished MMP2-promotor activity by hU-II and FoxO3.
In summary the results show that U-II induces MMP-2 expression and activity via FoxO3. Since GPR14, MMP2 and FoxO3 are highly expressed in the media of remodelled vessels, this pathway may provide a new therapeutic target for treatment of PH.

O9-2
Urotensin-II activates Rac-1 and ROS production in pulmonary artery smooth muscle cells involving Gαi3 proteins

Diebold I., Petry A., Schäfer N., Hess J., Görlach A.
Experimental Pediatric Cardiology, Dept. of Pediatric Cardiology and Congenital Heart Diseases, German Heart Center Munich at the TU Munich, Germany

Human urotensin II (hU-II) has recently been described as a potent vasoactive peptide associated with pulmonary vascular remodelling processes and pulmonary hypertension. hU-II acts via the G-protein coupled receptor GPR14. However, the mechanisms mediating GPR14-signalling remain unclear. ROS have been shown to act as signalling molecules in vascular cells, and NADPH oxidases which are activated by Rac-1 are important sources of ROS generation. Therefore, we investigated whether hU-II and GPR14 can activate Rac-1 and ROS production in pulmonary artery smooth muscle cells (PASMC).
Urotensin-II activated Rac-1 within 15 seconds and this response was abolished by treatment of PASMC with the GPR14 antagonist urantide. In addition, hU-II induced a rapid increase in ROS levels within 5 minutes of application, which was prevented by urantide or transfection of a dominant-negative mutant of Rac-1. Furthermore, in cells overexpressing GPR14 or constitutively active Rac-1, hU-II-stimulated ROS generation was further enhanced. Interestingly, in the presence of Pertussis toxin, hU-II-induced Rac-1 activation and ROS generation were abolished, indicating coupling of GPR14 to Gαi proteins. Indeed, transfection of dominant-negative Gαi3 completely diminished hU-II-stimulated ROS generation and Rac-1 activation in PASMC. Finally we could show that hU-II increased proliferation of PASMC. Similarly expression of active Rac-1 or NOX4 increased proliferation of PASMC whereas dominant-negative Rac-1 or depletion of NOX4 decreased U-II-stimulated proliferation.
In summary, the results show, that hU-II leads to a rapid increase in ROS production by binding to its receptor GPR14 and activation of Gαi3 protein, which then leads to activation of Rac-1 and subsequently to ROS formation by a NOX4-dependent NADPH oxidase. This pathway then results in increased proliferation of PASMC. Since media thickening is a hallmark of pulmonary vascular remodelling, and GPR14 was found in remodelled pulmonary vessels, this novel mechanism may provide a therapeutic target for pulmonary vascular remodelling associated with pulmonary hypertension.

O9-3
Characterisation of circulating multipotent progenitor cells during postnatal human development

Rupp S. (1), Koyanagi M. (2), Iwasaki M. (2), Trauth J. (1), Schütz C. (2), Bushoven P. (2), Schranz D. (1), Zeiher AM. (2), Dimmeler S. (2)
Pediatric Heart Center, University of Giessen and Marburg, Giessen, Germany (1); Department of Molecular Cadiology, Internal Medicine III, J.W. Goethe University, Frankfurt, Germany (2)

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Background: Multipotent stem cells are known to exist during embryonic development and in umbilical cord blood. These cells can differentiate into cardiovascular lineages. In adults, circulating cells which are characterised by the co-expression of hematopoietic stem cell markers and endothelial markers are known to contribute to neovascularization. However, their proliferation and differentiation capacity is reduced compared to embryonic or umbilical cord blood cells. We hypothesised that distinct subsets of circulating progenitor cells might exist during early human postnatal development and during childhood.
Methods and Results: In order to determine the phenotype and the functional properties of circulating progenitor cells during postnatal human development, we isolated circulating progenitor cells from peripheral blood of children aged between 8 days and 2.5 years (n = 8). For that purpose, isolated mononuclear cells were plated on fibronectin-coated dishes in EBM medium supplemented with growth factors. At day 14, isolated cells showed mesenchymal markers CD13 and CD73, endothelial markers CD 105, KDR and VE-cadherin, but were negative for hematopoietic markers CD45 as shown by FACS analysis and RT-PCR. This is in contrast with endothelial progenitor cells from adults which express hematopoietic and endothelial markers but lack the expression of mesenchymal markers. The marker expression profile resembles the profile of vessel associated resident mesoangioblasts (CD44+, CD13+, CD45−, CD31−). Circulating progenitor cells from children show a marked proliferation (27.6+/− 1 passages, 64.8+/−3.6 population doublings) before entering a senescent state. Consistently, these cells display a high telomerase activity (0.58+/−0.03). Cells differentiate into cardiomyocytes, adipocytes, smooth muscle cells, endothelial cells or osteoblasts. Injected cells improved recovery after hind limb ischemia or myocardial infarction in the mouse model. Children derived circulating progenitor cells express several transcription factors such as Isl1 and Nkx2.5, which were recently described as important factors for mesodermal cell determination.
Conclusion: Our study identified novel multipotent mesodermal progenitor cells in the peripheral blood of children, which are easily accessible, can be expanded in vitro to large numbers and are capable to differentiate into all 3 distinct cardiovascular cell lineages in vitro and in vivo. These blood-derived cells may represent a correlate of embryonic dorsal aorta-derived mesoangioblasts.

O9-4
Clinically benign LQTS founder mutation in the Swedish population
Winbo A(1), Stattin E.L.(2) Jensen S.M.(3), Diamant U.B.(3), Rydberg A.(1)
Department of Clinical Sciences(1); Division of Medical and Clinical Genetics, Department of Medical Biosciences(2); Department of Cardiology, Heart Centre(3) Umeå University Hospital, Umeå, Sweden

Introduction: Risk stratification in the congenital long QT syndrome (LQTS) remains a challenge. Mutation-specific effects on clinical expression have been earlier reported. Founder populations may serve as models in which genotype-phenotype correlations can be studied. The aim of this study was to investigate the symptomatology of carriers of the KCNQ1 c.332A>G mutation in the Swedish population.
Methods: While conducting a national inventory of LQTS-causing mutations, all families of index cases with suspected LQTS were subjected to genetic analysis by mutation identification and cascade screening, genealogical studies, analysis of clinical data (medical journals, familial interview and questionnaire) and ECG recordings. The familial interview of the carriers included the analysis of the age at death of family members, end-point sudden cardiac death (SCD).
Results: Forty-one patients (27 adults, age > 40 y, and 14 children, age 1–17y) were identified as carriers of the KCNQ1 c.332A>G mutation. This mutation was previously described by Splawski 2000 in one patient with long QT syndrome. Enrichment of the mutation, genealogy and the geographical clustering were indicating a founder effect in the Swedish population.
Carriers of the c.332A>G mutation had a history of syncope in 34% (12 adults and 2 children) but no life threatening cardiac event or aborted cardiac arrest could be reported in the study group. None of the carriers presented in the familial interview any experience of early sudden cardiac death < 40y within ancestors in the family, apart from one case of drowning.
Conclusions: The c.332.A>G mutation seems to cause a benign phenotype among mutation-carriers in Swedish families, supporting the existence of mutation-specific effects on risk for life threatening cardiac event or aborted cardiac arrest. Mutation-specific risk stratification could enable family-adjusted information and management of selected LQTS families. The national inventory is proceeding and complementary results will be presented.

O9-5
Development of an adjustable device for transcatheter pulmonary artery banding: evaluation in animals
Boudjemline Y.
Necker For Sick Children Hospital, Paris, France

Objective: Pulmonary artery banding (PAB) is the first palliation in infants with complex congenital heart disease and elevated pulmonary blood flow. In older patients with corrected transposition of the great arteries, it may be used to retrain the left ventricle. To date, the only option is surgical. Following our initial report with an endovascular device, we report design and developmental improvements to obtain a two-way adjustable device for transcatheter PAB.
Methods: We intended to implant percutaneously (20-Fr) a newly designed PA reducer between the native pulmonic valve and the pulmonary bifurcation in 8 sheep. This reducer was made of a balloon and a self expandable stent. During delivery, the self-expandable stent is firstly opened and fixed to the pulmonary wall. This is followed by the inflation of a balloon catheter to expand the inner part of the device. Initial inflation of stents was 18-mm. After, insertion we planned to calibrate the banding in both directions: opening and closing using balloons and snares of various diameters. This calibration was repeated after a follow-up of one month.
Results: The reducer was delivered successfully in all. It allowed the PA diameter to be reduced. No para-prosthetic leak was found when injecting contrast dye. Downsizing this diameter to 10-mm (decrement of 2-mm) was possible in all animals but one acutely and in all at one month evaluation. Down to 12-mm, the downsizing was tolerated in all animals but one. At 10 and 12-mm for one animal, the re-opening of the reducer was required. The reducer was thereafter expanded from 12-mm to 22-mm without any problem during the follow-up. Right ventricular pressure gradually increased following reduction of PA diameter up to a certain point (mean 65% of Aortic pressure). After this point, decrease in aortic pressure appeared and prompted us to re-open the reducer.
Conclusion: Implantation of a reducer is possible in animal through a transcatheter approach allowing intravascular PAB. The new developments of this device allowed us to calibrate in both directions the PAB.

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O9-6

Accessory atrioventricular pathways in normal mouse heart development, possible explanation for perinatal atrioventricular reentrant tachycardias

Hahurij N.D. (1), Kolditz D.P. (2), Bökenkamp R. (1), Markwald R.R. (3), Schalij M.J. (2), Poelmann R.E. (4), Gittenberger-de Groot A.C. (4), Blom N.A. (1)
Department of Pediatric Cardiology, Leiden University Medical Center, Leiden, The Netherlands (1); Department of Cardiology, Leiden University Medical Center, Leiden, The Netherlands (2); Department of Cell Biology and Anatomy, Medical University of South Carolina, South Carolina, USA (3); Department of Anatomy and Embryology, Leiden University Medical Center, Leiden, The Netherlands (4)

Introduction: Fetal and neonatal atrioventricular reentrant tachycardias (AVRTs) spontaneously resolve in the majority of cases. Late presence of accessory atrioventricular myocardial pathways (APs) during normal formation of the annulus fibrosis may form the substrate of perinatal AVRTs.
Methods: Electrophysiological recordings of ventricular activation patterns were performed in early (13,5-15,5 dpc; n = 29) and late post-septated (16,5–18,5 dpc; n = 13) stages of mouse heart development by positioning unipolar electrodes on the right atrium, left ventricular apex and left and right ventricular base. Furthermore, all hearts were investigated immunohistochemically using monoclonal antibodies specifically against MLC-2a, Nkx2.5, periostin and Cx43.
Results: In early post-septated hearts a mean heart rate of 115 ± 41 bpm and an AV conduction time of 80 ± 17 ms were recorded. Of these hearts only 38% showed a mature ventricular apex-to-base activation pattern, all others (62%; n = 18) showed a ventricular base-to-apex conduction pattern, indicating the presence of antegrade conducting APs. In late post-septated hearts the mean heart rate and AV conduction time recorded were 95 ± 27 bpm and 81 ± 18 ms, respectively. In 46% (n = 6) of these hearts antegrade conducting APs were still present, mainly at the left side of the atrioventricular junction. Immunohistochemical analysis confirmed the presence of APs, staining positive for MLC-2a and Nkx2.5 and negative for Cx43, indicating an AV myocardial origin. Interestingly, in the developing annulus fibrosis periostin expression was mainly observed adjacent to the APs, suggesting an inductive role of periostin in formation of the isolating fibrous tissue. Longitudinal analysis showed that the APs gradually decreased in number (p = 0.003) and size (p = 0.035) at subsequent stages of development (13.5-18.5 dpc).
Conclusion: Functional APs remain present at late stages of normal mouse heart development. These APs may serve as substrate for perinatal AVRTs.

O10-1

N-terminal-pro Brain Natriuretic Peptide Correlates with Treatment Effects of Bosentan Therapy in Eisenmenger Patients - First Results of the German Competence Network for Congenital Heart Disease

Farahwaschy B. (1), Mebus S. (2), Berger F. (1), Lange P.E. (3), Schulze-Neick I. (4)
Department of Congenital Heart Defects and Pediatric Cardiology, German Heart Institute Berlin, Germany (1); Department of Paediatric Cardiology and Congenital Heart Defects, German Heart Centre Munich, Germany (2); Competence Network for Congenital Heart Disease, Germany (3); Pulmonary Hypertension Team, Cardiac Unit, Great Ormond Street Hospital for Sick Children, London, Great Britain (4)

Introduction: Growing evidence indicates that bosentan is effective in the treatment of patients with Eisenmenger syndrome. Nevertheless, not all patients respond unequivocally and equally to this therapy. Due to the complexity of the disease, the subjective evaluation of the therapeutic outcome by the patient itsself is often difficult, whereas an objective hemodynamic assessment by cardiac catheterisation is linked to considerable risks.
The N-terminal-pro brain natriuretic peptide (NT-pro BNP) is related to right heart morphology and dysfunction in patients with pulmonary hypertension (PH) and possesses prognostic value in various forms of PH. We aimed to investigate its use in evaluating the response to bosentan in Eisenmenger patients by correlation with exercise and hemodynamic parameters.
Methods: 60 adult patients with Eisenmenger physiology received bosentan in an open-label, single-arm, prospective, multicenter study. Physical examination, 6-MWT, cardiopulmonary exercise testing and right heart catheterisation were performed both before and after 24 weeks of treatment. Additionally, blood samples were obtained for assessment of serum levels of NT-pro BNP.
Results: After bosentan treatment, patients showed a significant improvement in exercise capacity being reflected in an increase in 6-MWT (+72 ± 84 m; p < 0.0001) and peak oxygen uptake ((VO_2/kg) +0.82 ± 2.74 ml/min/kg; p = 0.0552) and a decrease in VE/VCO_2 (-4.39 ± 11.51; p = 0.0305). Baseline levels of serum NT-pro BNP were highly above normal with 1821 ± 5047 pg/ml and decreased significantly to 1408 ± 3389 pg/ml (p = 0.0130) under therapy. Values of 6-MWT, VO_2/kg and VE/VCO_2 showed a significant correlation with NT-pro BNP both at baseline and after treatment, whereas correlation with right atrial pressure (RAP) was only significant during baseline conditions.

	Correlation with NT-pro BNP Before Therapy		Correlation with NT-pro BNP After Therapy	
	r	p	r	p
6-MWT [m]	-0.3728	0.0042	-0.3043	0.0210
VO_2/kg [ml/min/kg]	-0.4864	0.0002	-0.4530	0.0007
VE/VCO_2	0.3843	0.0086	0.4620	0.0020
RAP [mmHg]	0.5571	0.0000	0.2308	0.0707
PVRi	0.1615	0.1364	0.0622	0.3477

Conclusions: In our 60 Eisenmenger patients NT-pro BNP correlates significantly with major noninvasive and invasive parameters indicating the effect of 24 weeks treatment with bosentan. Our results show that serum NT-pro BNP levels may be a useful marker for therapy monitoring in these and similar patients, thus avoiding the need for cardiac catheterisation.

O10-2

Clinical and Exercise Effects of a 24 week bosentan treatment of 60 Patients with Eisenmenger's (EM) Physiology- First Results of the German Competence Network for Congenital Heart Disease

Mebus S. (1), Farahwaschy B. (2), Lunze K. (2), Gilbert N. (2), von Bismarck I. (2), Peters B. (3), Berger, F. (2) Lange P.E. (4), Schulze-Neick I. (5)
Department of Paediatric Cardiology and Congenital Heart Defects, German Heart Centre Munich, Germany (1); Department of Congenital Heart Defects and Pediatric Cardiology, German Heart Centre Berlin, Germany (2); Department for biometry and medical informatics, University Magdeburg, Germany (3); Competence Network for Congenital Heart Disease, Germany (4); Pulmonary Hypertension Team, Cardiac Unit, Great Ormond Street Hospital for Sick Children, London, Great Britain (5)

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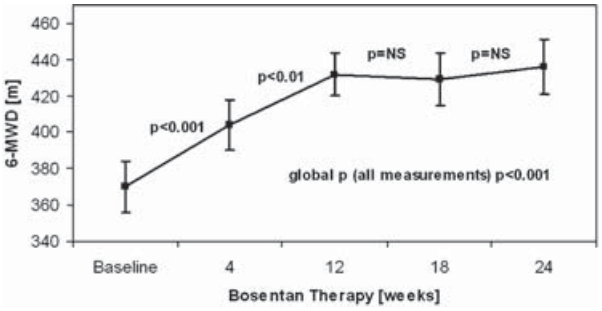
Background: EM physiology causes a complex and multisystem disorder including progressive hypoxemia, dyspnoea, cyanosis, right-sided heart failure and reduced quality of life. Currently, new selective pulmonary vasodilators have proven to be beneficial in other various forms of pulmonary arterial hypertension. Similar data in EM patients while showing promising benefits suffer from incomplete documentation or small patient numbers. We present our experience with a large cohort of EM patients and response to specific anti-pulmonary hypertensive therapy.
Methods: In this open-lable, single-arm, prospective and multicentre study, patients with EM physiology were recruited to receive bosentan (Tracleer®) for 24 weeks. Treatment effects were evaluated by clinical outcome, WHO functional class, transcutaneous saturation (TCS), maximal metabolic exercise capacity and 6-minute walking distance (6-MWD), echocardiographic and magnetic resonance imaging, and cardiac catheterisation. In addition, patients received questionnaires to estimate quality of life (SF-36) and potential treatment side effects during the treatment.
Results: Sixty patients (25 male, 35 female, average age 35, range 16-51 years) were enrolled. 4 patients did not finish the study and this was related to treatment side effects in one patient. 6-MWD increased from 370 ± 101 to 436 ± 108 m (mean of differences 72 m; $p < 0.0001$), together with an improved blood pressure response at peak exercise (RRmax dia 73 ± 15 to 68 ± 13 mmHg, $p = 0.03$; RRmax sys 125 ± 19 to 129 ± 22 mmHg, $p = 0.09$; RR amplitude 52 ± 9 to 61 ± 8 mmHg, $p = 0.02$). Maximum oxygen uptake trended to increase (14.1 ± 4.1 to 14.8 ± 4.1 ml/min*kg, $p = 0.06$), but the VE/VCO2-slope improved (56.4 ± 17.9 to 52.3 ± 11.6 L/L, $p = 0.03$). Clinical WHO function improved at least one class in 15, whereas it worsened in 3 (average WHO class change, 2.64 to 2.38, $p < 0.05$). TCS did not decrease (81.8 ± 6.7 to 82.6 ± 7.2%, p = NS).
Conclusions: In patients with EM physiology, bosentan was safe and effective. The treatment resulted in significant improved ambulatory exercise tolerance and clinical status and a trend in overall metabolic exercise function. These marked responses to a relatively short period of 24 weeks of treatment, considering the long-standing of the disease, underline that patients with Eisenmenger's disease do profit from this therapeutic principle.



O10-3
Prenatal versus Postnatal Diagnosis of Significant Congenital Heart Disease in the Current Era
Andrews R.E., Jones A.M.R., Callaghan N., Miller O.I., Simpson J.M., Sharland G.K.
Evelina Children's Hospital, Guy's and St Thomas' NHS Foundation Trust, London, UK

Objectives: To determine the proportion of babies with significant congenital heart disease (CHD) who are prenatally diagnosed in the current era, and which diagnoses are most likely to go undetected. The majority of CHD detected prenatally is found in low risk pregnancies during routine obstetric anomaly scans.
Methods: Records from our fetal and paediatric cardiac databases were examined to determine the total number of fetuses and infants with significant CHD with expected delivery dates in, or born in, 2006. Significant CHD was defined as that likely to require catheter intervention or surgery before the age of 1 year. Details of individual diagnoses and their timings were noted.
Results: Of 1699 pregnant women with expected delivery dates in 2006 undergoing fetal echocardiography in our unit, 136 fetuses with significant CHD were identified. Of these, there were 9 intrauterine deaths, 51 terminations of pregnancy, 68 livebirths seen postnatally in our unit (2 were born in late 2005), and 8 seen postnatally at other hospitals. Of 653 infants born in 2006 referred to our paediatric cardiac service, 220 had significant CHD. Of these, in addition to the 66 diagnosed prenatally in our unit, 46 were diagnosed prenatally elsewhere, and 108 were diagnosed postnatally. Lesions most likely to be detected prenatally during obstetric scanning included those with very abnormal 4 chamber views (mitral/tricuspid atresia, 12/12 prenatally diagnosed; hypoplastic left heart syndrome, 21/26), and those with complex CHD/multiple abnormalities (33/38). Lesions least likely to be detected included total anomalous pulmonary venous drainage (0/6), ventricular septal defects (5/23), and lesions affecting the outflow tracts (transposition of the great arteries 3/12, aortic stenosis/atresia 3/10, and pulmonary stenosis/atresia with intact septum, 10/29).
Conclusion: Prenatal cardiac screening using the 4 chamber view was introduced over 20 years ago, but a significant number of defects with an abnormal 4 chamber view are still overlooked during obstetric ultrasound screening. Incorporation of outflow tract views increases the range of diagnoses detected prenatally, but this step has not been universally implemented. Further study is needed to determine the effect of screening nationally, and the cost/benefit ratio of incorporating further views into routine practice.

O10-4
Dilated Cardiomyopathy (DCM) in Children: novel strategies for improving outcome
Schranz D., Rupp S., Reinsch H-L., ElMakfi N., Michel-Behnke I., Kreuder J., Thul J., Müller M. Bauer J., Valeske K., Akintuerk H.
Pediatric Heart Center JLU Giessen, Germany

Background: DCM is the most common form of cardiomyopathy and cause for cardiac transplantation (HTX) in children. The incidence of new-onset heart failure due to heart muscle disease in childhood is 0.87/100 000. One third of children die or require ransplantation within 1 year of presentation (Circulation 2008; 117:79).
Objective: Retrospective study to determine the outcome of infants and children with new-onset heart failure due to DCM referred to our Paediatric Heart Transplantation Centre from January 1st 2003 to December 31st 2007.
Methodology: The clinical profile and course of 23 infants and children (median age 3years, range, 1 day to 14years) with DCM were evaluated to detect factors that might predict outcome. Factors studied included age, gender, history, symptoms and clinical signs at presentation. Furthermore, data on serial echocardiographic and histological findings, and treatments were analyzed.
Results: Causes of DCM included idiopathic DCM (n = 11), familial (1), TNT2 gene mutation (1), myocarditis (2), occult arrhythmias (2), anthacycline toxicity (1), metabolic (2), and left ventricular non-compaction (3). During a median follow-up of 23 months (range 2 to 52 months) no patient died despite 20 pts were admitted in NYHA IV, 8 pts after prior resuscitation, and all but 3 with catecholamine therapy at admission. 11 pts received HTX,

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4 after previous use of assist device (Berlin Heart); 4 pts with left ventricular failure were successfully treated by a pulmonary artery banding, 3 pts were treated by intracoronary autologous stem cell therapy. All, but one non-transplanted pts remained on medication, and treated by ß-blockers and ACE-inhibitors. All were discharged home in NYHA I and II, only one with NYHA III. Clinical improvement correlated with a significant decrease of BNP-values (admission-values: median.3046; max 12797; min 76 pg/ml; values at discharge median 491; max 2643, min 55 pg/ml).
Conclusion: Improved survival is shown by utilizing all current therapeutic resources. Novel therapeutic strategies as autologous stem cell therapy and placement of a pulmonary banding might be an adjunctive option in selected patients to avoid or delay HTX.

O10-5
Acute rejection after pediatric heart transplantation in the current era: Far less common, but still a serious problem
Lammers A.E. (1), Roberts P. (1), Brown K. (2), Rees P. (1), Kostolny M. (3), Sebire N. (4), Burch M (1)
(1) Paediatric Cardiology, (2) Paediatric Cardiac Critical Care, (3) Department of Surgery, (4) Department of Pathology, Great Ormond Street Hospital for Children, London, UK

Introduction: We report prevalence and outcome of rejection following heart transplantation in the recent era from a single center.
Methods: Retrospective review of biopsy proven rejection/acute hemodynamic collapse. Cox proportional hazard analysis of predictors of rejection.
Results: Between 2002 and 2007 105 consecutive children (male n = 50) with a mean age of 8.3–5.8 years (0.1–17.9 years) underwent cardiac transplantation for dilated cardiomyopathy (DCM, n = 66), restrictive cardiomyopathy (RCM, n = 12) or congential heart disease (CHD, n = 27).
In 271.9 patient-years we observed 23 episodes of significant rejection in 21 patients, 20% of patients treated for rejection and 0.2 episodes per patient .
Five presented in hemodynamic collapse 1.6-35.9 months after transplant requiring ECMO support, of whom four survived the rejection episode. On 5 occasions children presented with symptoms of cardiac dysfunction, whereas in 13 asymptomatic children significant rejection (>3A) was found on a routine surveillance biopsy. Only two children had evidence for humoral rejection on their endomyocardial biopsy (positive C4d stains). Apart from 1 death from severe sepsis of a patient requiring ECMO, all other children survived the rejection episode.
Cox analysis showed a younger age was associated with a significantly reduced risk for rejection($p < 0.03$). A combination of Tacrolimus and Mycophenolate mofetil (MMF) had a better protective profile and was associated with a significantly lower incidence of rejection ($p = 0.047$), whereas Sirolimus and MMF has a significantly higher risk for rejection ($p < 0.01$).
Children with < 5 HLA mismatches appear to have a lower risk of rejection, however these observations did not reach statistical significance. Body mass index, ethnicity, diagnosis,CMV status of the recipient/-mismatch, number of HLA-A, -B and –DR mismatches, morbidity (post-op ICU stay, hospitalization) were not associated with rejection.
Conclusions: While rejection rates in the current era are substantially lower than registry data 10% of patients experience symptomatic rejection and half of these require mechanical circulatory support. Humoral rejection was uncommon and only age was a predictor of risk. Medication regimes significantly differed in the incidence of rejection.

O10-6
Time-scale for Recovery of Heart Function in Children with Acute Severe Heart Failure
Roche S.L., Crossland D.S, Chaudhari M.P, Kirk R., Hasan A., O'Sullivan J.J.
Freeman Hospital, Newcastle upon Tyne, United Kingdom

Introduction: A ubiquitous challenge exists in the management of children who present with severe heart failure in the absence of congenital heart disease: after what duration of illness is recovery of ventricular function so unlikely that pursuit of recovery with medical therapy should be abandoned in favour of cardiac transplantation? The time course of ventricular recovery has not previously been studied.
Methods: A cohort of 27 children, (< 16yrs), with severe ventricular dysfunction (LV FS < 15%), no evidence of congenital heart disease and symptomatic heart failure in whom full clinical and echocardiographic recovery occurred (LV FS>30%) were identified from the records of a single paediatric cardiac transplantation centre during a 15 year period. Notes were retrieved, clinical data extracted and echocardiograms reviewed.
Time from presentation until the last echocardiogram that demonstrated LV FS < 20% and FS < 30% was recorded. These times were used to describe the earliest possible time taken to recover. Patients were only included if documented to have LV FS > 30% and to be asymptomatic on most recent follow-up.
Results: Mean age at clinical presentation was 15.7 (0.2–72) months. Male:Female = 0.7:1. 21/27 required inotropic support at presentation. Mean support duration = 9.7(1–36) days. Three patients required ECMO for 8, 14 and 68 days respectively. Eight patients were listed for cardiac transplantation and would have been transplanted had an organ become available. Mean duration of listing 260 (10–540) days.
Evidence of viral infection was detected in 4/27 patients. Routine biopsy was not performed. One patient was treated with immunoglobulin, none received steroid.
Mean time to ventricular improvement to FS>20% was 3.6 (SD 4.7 range 0.3–4) months. Mean time to FS>30% was 8.9 (SD 7.7 range.0.7–24) months. There was no correlation between age at presentation and time to recovery to either FS>20% ($r = -0.11$) or FS>30% ($r = -0.26$).
Conclusions: All patients in our population who have recovered from severe heart failure have been younger than 6 years of age at presentation. Although the majority of those who will recover do so early, a small but definite group exist who show ventricular recovery as late as two years after presentation.

PW1-1
Hemodynamic Responses of 60 Eisenmenger Patients Treated 24 Weeks with Bosentan – First Results of the German Competence Network for Congenital Heart Disease
Farahwaschy B. (1), Mebus S. (2), Lunze K. (1), Gilbert N. (1), von Bismarck I. (1), Peters B. (3), Peters B. (1), Miera O. (1), Berger F. (1), Lange P.E. (4), Schulze-Neick I. (5)
Department of Congenital Heart Defects and Pediatric Cardiology, German Heart Institute Berlin, Germany (1); Department of Paediatric Cardiology and Congenital Heart Defects, German Heart Centre Munich, Germany (2); Department for biometry and medical informatics, University Magdeburg, Germany (3); Competence Network for Congenital Heart Disease, Germany (4); Pulmonary Hypertension Team, Cardiac Unit, Great Ormond Street Hospital for Sick Children, London, Great Britain (5)

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Introduction: In Eisenmenger patients, increasing pulmonary blood flow by using targeted pulmonary vasodilative medication to improve exercise tolerance and quality of life is an attractive concept, and is indeed supported by an increasing number of clinical studies. However, unless such a vasodilator is selective for the pulmonary circulation, the concomitant systemic vasodilation may allow increased right-to-left shunting and cyanosis instead. In order to obtain more insight in this promising therapy, we performed a large multicenter national study and examined the clinical and hemodynamic effects in patients with Eisenmenger physiology in response to treatment with bosentan, which competitively blocks the effects of the powerful endogenous vasoconstrictor endothelin.
Methods: 60 patients (median age 35, range 16–51 years) with Eisenmenger physiology were included into this open-label, single-arm, prospective, multicenter study. In addition to extensive clinical and noninasive examinations, cardiac catheterisation was performed both before and after 24 weeks of bosentan (Tracleer®) treatment. Following assessment of oximetry and systemic/pulmonary artery pressures, systemic (SVRi) and pulmonary (PVRi) vascular resistance indices and blood flows (CIpul and CIsys) were calculated according to the Fick principle. Maximum pulmonary vasodilatory reserve was evaluated ("testing"), using an inhaled combination of all three substances oxygen, nitric oxide and nebulised ilomedin.
Results: Clinically, the patients showed improvement with the bosentan treatment (+72 ± 84 m in the 6-MWT, $p < .0001$; average improvement of 0.28 NYHA class as a group, $p < 0.05$). This was associated with a significant change including all contributing variables of both pulmonary and systemic vascular beds (see table). There was also a slight preference for an effect on the pulmonary circulation including an augmentation of maximum pulmonary vasodilatory reserve after treatment.

Variable	Before Therapy		After Therapy		Effect of Therapy	
	Baseline	Testing	Baseline	Testing	% change (base)	p
RAP [mmHg]	5.30	5.40	5.50	5.30	3.8	0.179
MPAP [mmHg]	81.5	79.1	78.1	74.0	4.2	***0.007**
$Sat_{Ao}O2$ [%]	80.3	90.8	82.0	93.7	2.1	0.283
Cl_{pul} [l/min*m^2]	2.10	3.83	2.35	4.76	10.6	***0.040**
Cl_{sys} [l/min*m^2]	2.64	2.61	2.71	2.51	2.6	0.639
PVRi [WU*m^2]	42.5	27.2	36.7	23.5	13.7	***0.001**
--%red by testing	38.0	---	39.4	---	3.5	0.686
SVRi [WU*m^2]	34.8	37.9	31.3	35.6	10.1	***0.030**
PVRi/SVRi ratio	1.39	0.97	1.27	0.71	8.9	0.261
--%red by testing	34.2	---	45.4	---	7.9	0.243

Conclusions: In this largest and carefully studied patient cohort with Eisenmenger physiology, 24 weeks of bosentan treatment leads to a clinical improvement which is accompanied by small albeit significant hemodynamic changes without pulmonary specificity, however. This is a finding also inherent, but not yet explicit adressed, in previous studies. On the basis of our data, the clinical improvement is probably better explained by other mechanisms as opposed to the minimally selective changes of PVRi induced by bosentan treatment.

PW1-2
Novel Genetic Associations with Delayed Cardiomyopathy Onset in Muscular Dystrophy Utilizing MDSTARnet Data

Barber B.J., Andrews J.G., Cunniff C.M., Klewer S.E., Miller T.M., Meaney F.J., and MDSTARnet
University of Arizona, College of Medicine, Tucson, Arizona, U.S.A.

Introduction: Heart failure is one of the leading causes of death in Duchenne and Becker muscular dystrophy (DBMD) patients. DBMD patients exhibit few symptoms or signs of heart failure, yet early detection and treatment of cardiomyopathy (CM) may lead to improved outcomes.
Discovery of a genotypic marker for severity and onset of CM could be of paramount importance in DBMD. A marker might identify a subset of patients in which aggressive monitoring and early treatment could be employed. Recently published evidence has associated dystrophin gene mutations in exons 12 and 14–17 with CM, and those in exons 51-52 with possible protection against CM.
We sought to confirm these findings utilizing data from the Muscular Dystrophy Surveillance, Tracking and Research Network (MDSTARnet), a Centers for Disease Control and Prevention cooperative agreement with five participating sites (Arizona, Colorado, Georgia, Iowa, and western New York) and the first population-based surveillance system for DBMD. One objective of MDSTARnet is to abstract medical records for data on multiple health outcomes in DBMD, including CM.
Methods: Of 471 DBMD subjects, 73 had both the echocardiographic and genetic mutation data to be classified into one of 2 groups: 1) Early CM = CM onset at age < 15 years or 2) Delayed CM = no evidence of CM by age 15. CM was defined via echocardiographic parameters of systolic function: SF < 28% and/or EF < 55%. Each affected dystrophin exon was tested in 2x2 tables relating presence of mutation to CM group using Fisher's Exact Test.
Results: Dystrophin mutations in the following exons were associated with delayed CM: exon 3 ($p = 0.009$), 4 ($p = 0.009$), 7 ($p = 0.037$), 46 ($p = 0.005$), and 47 ($p = 0.009$). No mutations were significantly associated with early CM.
Conclusions: MDSTARnet data revealed a significant association of delayed CM onset in DBMD patients with dystrophin mutations in exons 3, 4, 7, 46 and 47. These data differ from previous reports and suggest that additional dystrophin gene mutations are associated with delayed CM onset in DBMD.

PW1-3
Indications and outcome of radiofrequency ablation in infants: a retrospective UK multicentre study over 15 years

Shebani S.O., Shebani S.O. (1), DeGiovanni J. (2), Macleod K. (3), Morgan J. (4), Nanapragassam J. (4), Peart I. (5), Rosenthal E. (6), Stuart G. (7), Till J. (8), Walsh K. (9), Duke C. (10).
Royal Manchester Children's Hospital, UK (1); Birmingham Children's Hospital, UK (2); Royal Hospital for Sick Children, Glasgow, UK (3); Southampton General Hospital, UK (4); Royal Liverpool Children's Hospital, Alder Hey, Liverpool, UK (5); Evelina Children's Hospital, London, UK (6); Bristol Children's Hospital, UK (7); Royal Brompton Hospital, London, UK (8); Our Lady's Children's Hospital, Dublin, Republic of Ireland (9); Glenfield Hospital, Leicester, UK (10).

Introduction: The decision to perform radiofrequency ablation (RFA) in infancy (< 1 year) is controversial as most tachycardias respond to antiarrhythmic drugs and RFA is perceived as high risk. All UK infant RFA cases over the last 15 years were retrospectively studied to identify the clinical characteristics of patients, indications for RFA and success and complication rates.
Methods: Every UK paediatric electrophysiologist was contacted to identify ablations carried out in infancy at their institution since the inception of RFA. Patient and procedural data was collected by a single investigator visiting each centre to review medical records.

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Results: RFA in infancy was performed in 8/13 UK centres.
20 ablations were performed on 19 infants (age 0–12 months, mean 2.6 months, weight 3.5–10, mean 5.9 kg) 15 had normal anatomy and 4 congenital heart disease. 9 had atrioventricular re-entrant tachycardia, 7 permanent junctional reciprocating tachycardia, 2 ectopic atrial tachycardia and 1 atrial flutter. 14 presented with incessant tachycardia and 4 with frequently recurring tachycardia. 10 had tachycardia related cardiomyopathy and 7 haemodynamic compromise. The indication for RFA in 18 patients was resistance to multiple antiarrhythmic drugs. 1 RFA was elective pre-surgery. In 4 infants antiarrhythmic drugs caused severe adverse effects (poor function, hypotension, bradycardia, collapse). Four required extracorporeal circulatory support (ECMO) and 3 had RFA on ECMO. 19/19 (100%) of procedures were acutely successful, with 5 (26%) recurrences at 7–23 (mean 16) days. One RFA was repeated in infancy. 4 patients (20%) had a major complication (complete heart block, valve perforation, 2 pericardial tamponade) and 3 (15%) a minor complication (thrombus on mitral valve, skin burn. pericardial effusion). There was no procedure related mortality. 9/10 tachymyopathies recovered fully after 5–75 (mean 24) days. There have been no adverse events on 0.5–14 (mean 6) year follow up.
Discussion: UK cardiologists remain conservative, only attempting RFA after failure of multiple drug therapy, even in the setting of tachymyopathy or haemodynamic compromise, both of which may worsen with drug therapy. RFA is acutely successful, but recurrence rates were high. Complication rates (20%) were higher than those reported from the US paediatric ablation registry (7.8%).

PW1-4
Arrhythmias during follow-up in Patients undergoing either surgical or Percuteneous Atrial Septal Defect Closure

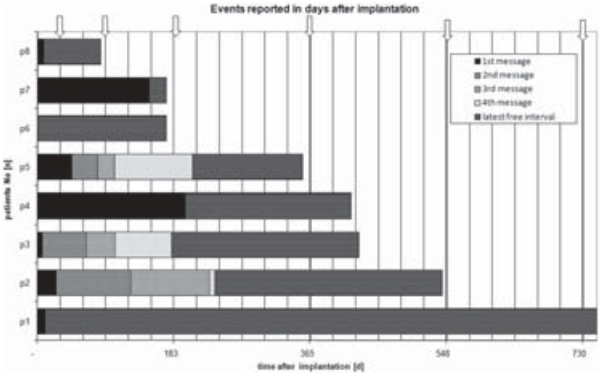
Butera G. (1), Ferrero P. (2), Foresti S. (2), Pittalis M. (2), Chessa M. (1), Carminati M. (1), Frigiola A. (1), Giamberti A (1), Abella R. (1), Pomè G. (1), Cappato R. (2)
Pediatric Cardiology and GUCH unit (1); Rhythmology and Electrophysiolgy unit (2); Policlinico San Donato IRCCS, San Donato Milanese - Italy

Introduction: ASD closure impact positively on patients arrhythmic history only if performed early, usually before 40 year of age. Surgical and percutaneus closure results during follow-up have never been compared so far. We aimed at comparing the arrhythmic outcome, in particular as far as atrial fibrillation (AF) is concerned, in a large cohort of patients.
Methods: Between January 1998 and January 2004, 929 patients who underwent ostium secundum ASD closure either surgically (340 pts) or percutaneously (589 pts). Symptoms and documented AF have been taken into account. Follow up of patients with documented recurrent episodes has been picked up.
Results: There were no differences in NYHA class distribution and comorbidities between the two groups. Patients treated surgically were older (22 ± 20 vs. 28 ± 19 years; $p < 0.01$) and had a longer follow-up (5 ± 2.3 vs. 5.48 ± 1.6; $p = 0.001$)
No differences were found between the two groups according to the documented AF either before (1,7% vs. 2,8%; $p = 0.4$) or after closure (4,1% vs. 2,7%; $p = 0.3$). New onset events of AF occurred equally in both groups (2.3% vs. 1,1%; $p = 0.2$). There were no differences for the occurrence of symptoms either before (27.5% vs. 27.6%;$p = 0,1$) or after closure (12.3% vs. 15.4%; $p = 0.3$).
Finally, in both groups, symptomatic patients were significantly lower after the procedure but those with documented episodes were not ($p = 0.2$).
AF recurrences free survival were plotted against time (months). Patients who underwent percutaneous closure display a trend of a better outlook not achieving the statistical significance, as showed by Log Rank test ($p = 0.13$). Episodes occurring within the first month have been ignored.
Conclusions: ASD closure affects positively symptoms perception. Even fulfilling the requirement of an early closure, as recommended by former papers, we couldn't demonstrate a clear benefit on documented AF episodes. Furthermore we observed a not negligible proportion of new onset AF equally represented in both groups, being likely related to a substrate remodelling independently from the procedure.

PW1-5
Telemonitoring in young patients with a congenital heart disease and the need for pacemakers or defibrillators

Zartner P., Handke R., Kallenberg R., Schneider M.B.E.
German Paediatric Heart Centre, Department of Cardiology, Sankt Augustin, Germany

Subject: Children and young adults with a congenital heart disease (CHD) show a wide spectrum of cardiac arrhythmias. With the indication for pacemaker or defibrillator (ICD) implantation, decision has to be made whether an online monitoring system will reliably improve further therapy.



Method: 8 patients (age 4.1 to 37.6 years, mean 15.5 years) with a congenital heart disease received an implantable device with an integrated Home Monitoring facility (Biotronik, Berlin). Follow-up time ranges between 168 and 896 days (mean 457 days, 10 patient years).
Results: In 7 of 8 patients specific event reports were received independently from follow up visits (figure) or individual perception of any clinically felt symptoms, which led to modification of antiarrhythmic medication, exercise tolerance, electrophysiological catheter ablation or system revision. Data tracking covered 94% of all days during follow up and gives more coherent information than any other system available. More than 1000 episodes were reported and evaluated. In three patients the reported events were of critical nature (ventricular fibrillation with shock not realised by the patient, lead failure, ventricular tachycardia in a Fontan patient).
Conclusion: Patients with a CHD and progressive arrhythmia or antiarrhythmic medication as well as patients with an ICD constitute a high risk group and should be followed as closely as possible. The integrated monitoring system reported decisive data on a daily basis, which led to early modifications of the actual therapy. Additionally it

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serves to control proper system function, analysing lead impedances, sensing and threshold parameters. For the patient's protection the safety and efficacy of the implanted systems are thus controllably improved. The online information system extends medical care especially in our young and active patient population.

PW1-6
Catheter Ablations In Children With Ventricular Tachycardia

Pokushalov E., Turov A., Shugaev P., Artemenko S., Romanov A.
Research Institute of Circulation Pathology,
Novosibirsk, Russia

Introduction: Evaluation of the effectiveness of catheter ablations of ventricular tachycardia.
Methods: 167 children with mean age of 13.8 ± 2.9 who underwent catheter ablation for different ventricular tachycardia were observed. The structure of the arrhythmias was as following – re-entry VT (n = 22; 13.2%), ectopic VT accompanied with frequent ventricular extra bit (n = 145; 86.8%; daily average number of extra bits being 16140 ± 930). VT had random paroxysm character in 71 patients (42.5%), constant paroxysm in 45 patients (26.9%) and constant-repeating in 51 patients (30.6%). Catheter ablations were necessary due to syncopal conditions (n = 39; 23.4%), arrhythmic heart failure (n = 55; 32.9%), refusal to take antiarrhythmical medication (n = 73; 43.7%).
Results: Re-entry VT in all cases had a fascicular character and was coming out of anterior (n = 8), posterior left bundle branch (n = 12) and right bundle branch of His (n = 2). Ectopic focuses were revealed in right ventricular outflow tract (n = 34), anterior wall (n = 13) and basal parts of the right ventricular (n = 14), aortic sinus (n = 38), in Purkinje's fibers of the left bundle branch of His area (n = 27), free wall of the left ventricle (n = 19). Ablation through pericardium was performed on three children with subepicardial localization. In 15 cases of septal focuses for enduring effect application on interventricular septum had to be done. In 110 cases ablation was performed with electroanatomical method (65.9%) in the rest cases (34.1%) with conventional method. 3 patients had complications in the form of a complete AV-block which required implantation of pacemaker (n = 1) and prolonged PR interval (n = 2). The defects of fascicular conduction in fascicular VT were not considered complications because they were the purpose of ablation. In two cases ablation was impossible because the focus was close to coronary artery (n = 1) or to bundle of His (n = 1). During RF applications there was always a distance of more than 12 mm to the coronary artery. During follow-up of 22.1 ± 6.5 months (up to 76 months) – relapses of VT appeared in 7 patients (4.1%); the effectiveness of repeated ablations was 100%.
Conclusion: Catheter ablation is a highly effective and safe method of treatment of VT in children.

PW1-7
Linkage of Left Ventricular Outflow Tract Obstruction to XQ28 in 3 French-Canadian pedigrees

Chetaille Ph. (1), Provost S. (2), Gendron R. (3), Thibeault M. (3), Bureau N. (3), Riopel K. (3), Siles A. (3), Mercier J. (3), Houde C. (1), Bigras JL. (3), Dubé MP. (2), Richter A. (3), Andelfinger G. (3)
Centre Mère-Enfant du Centre Hospitalier Universitaire de Québec, Québec, canada (1); Research Center, Montreal Heart Institute, Montréal, Canada (2); Cardiovascular Genetics, Sainte Justine Hospital, Montréal, Canada (3)

Introduction: Statistical genetic analysis of cohorts with left ventricular outflow tract obstruction (LVOTO) shows high heritability of such lesions. We have identified four possibly X-linked French-Canadian pedigrees with multiple cases with LVOTO and septal defects.
The aim of this study was to characterize the family structure and affected members in all four families and to evaluate three out of four pedigrees with LVOTO and septal defects for genetic linkage to the X chromosome.
Method: Detailed family history, physical exam, ECG, echocardiography and chart review was performed. Informed consent was obtained from all participants. X chromosome markers were genotyped on genomic DNA isolated from peripheral blood. We used a dominant genetic model and performed analyses with MLINK and MERLIN.
Results: A total of 109 members were enrolled out of which 39 were affected (29 males, 10 females). Seven members have an unspecified cardiac defect (two died under the age of 1, two at 7 and 11 and one at 43). Predominant lesions were atrial (n = 5) and ventricular (n = 6) septal defects, coarctation (n = 4) and aortic lesions (stenosis (n = 10), bicuspid/abnormal aortic valve (n = 12)), as well as mitral valve lesions (n = 4). Males had a high incidence of supraventricular arrhythmias postoperatively (4 with atrial flutter/fibrillation, one with ICD implantation). For families included in the parametric two-point linkage analysis, we obtained LOD scores above 2 for 3 markers in the Xq28 region. Multipoint non-parametric linkage analysis confirmed these results with a NPL score of 9.1 ($P < .00001$) over a STR marker in intron 13 of the F8C gene. Haplotype analysis allowed the definition of a candidate interval flanked by marker DXS8069 spanning to the telomeric end of Xq. No mutations were detected in the coding sequence of filamin A gene, a prime candidate within the critical region.
Conclusions: By analysing 3 multi-generation French-Canadian pedigrees, we mapped a syndrome of LVOTO and septal defects to the Xq28 region. The characterization of a fourth family mainly affected by aortic lesions is ongoing. Further work will refine our candidate region with dense SNP coverage to search for a shared haplotype identical by descent between all French Canadian families.

PW1-8
New observations on the structure/function coupling of the inter-ventricular septum

Corno A.F. (1), Kocica M.J. (2), Chappory L.A. (3), Moore S.A. (3), Sutherland H. (3), Jarvis J.C. (3).
Alder Hey Hospital, Liverpool, England (1), Institute for Cardiovascular Diseases, Belgrade, Serbia (2), Human Anatomy & Cell Biology, University of Liverpool, Liverpool, England (3)

Introduction: The inter-ventricular septum has been investigated to define the histological relationships between myocardial cells and connective tissue, and to quantify the distribution of gap-junctions (= intercalated disks) involved in propagation of the electrical excitation.
Methods: Pig hearts have been studied using 2 techniques: immune-staining with antibodies against Desmoplakins to delineate gap-junctions (= intercalated disks), and Picro-Sirius red-collagen-staining of frozen sections to delineate the connective tissue septa.
By overlaying serial sections 10 microns apart, the distribution of 1,010 gap junctions in 11 base-to-apex fields has been counted and scored in relationship to the connective tissue septa between groups of myocytes.
Results:
a) The inter-ventricular septum is a single anatomical entity, not separated into anatomical parts by major connective tissue septa

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b) There are perimysial connective tissue septa running base-to-apex within the inter-ventricular septum for considerable distances following spiral patterns
c) The myocytes are consistently grouped by these septa into sheets 5 or 6 myocytes thick
d) The gap junctions (= intercalated disks) are completely absent from the connective tissue septa larger than twice the myocyte diameter
e) Only 8.36% of the total number of gap junctions (= intercalated disks) were co-localized with connective tissue by overlaying sections stained for connective tissue or Desmoplakins. Co-localisation reduced to just 3.00% when counts were made in immune-stained sections with haematoxylin counter-stain, guided for connective tissue localization by comparison with Picro-Sirius red-collagen-stained sections.
Conclusions:
a) The reported ultrasound reflection from mid septum seems to be caused by a change in direction of myocytes rather than presence of a connective tissue sheet.
b) In the inter-ventricular septum the spiral pattern of the myocardial and connective tissue structures is confirmed at histological level.
c) The presence of few gap junctions (= intercalated disks) in the perimysial connective tissue septa of the inter-ventricular septum confirms the myocardial predisposition for longitudinal propagation of the electrical excitation.
d) The perimysial connective tissue septa, not directly involved in impulse transmission between the myocardial cells, modulate the ventricular propagation of the electrical excitation.
e) The structure/electrophysiological relationships within the inter-ventricular septum have important consequences in understanding the ventricular function and dysfunction.

PW1-9

Characterization of long term endogenous cardiac repair in children after heart transplantation

Rupp S. (1), Koyanagi M. (2), Iwasaki M. (2), Bauer J. (1), von Gerlach S. (3) , Schranz D. (1), Zeiher AM. (2), Dimmeler S. (2)
Pediatric Heart Center, University of Giessen and Marburg, Giessen, Germany (1); Department of Molecular Cadiology, Internal Medicine III, J.W. Goethe University, Frankfurt, Germany (2); Department of Pathology, University of Giessen and Marburg, Giessen, Germany (3)

Background: Circulating cells repopulate the heart at a very low rate in adult humans. However, the knowledge about time-dependent cardiac regeneration is very limited and the contribution of circulating cells to cardiomyocytes or vascular cells in children is unknown. This study investigates the endogenous repair capacity and the long-term incorporation of circulating cells in heart transplanted children.
Methods: Cardiac and endothelial chimerisms were detected in endomyocardial biopsies of 9 children (age 1 months-14 years) with sex-mismatched heart transplantation by fluorescence in situ hybridization. Cardiomyocytes were detected by alpha-sarcomeric actinin and troponin I staining. Endothelial cells were stained with CD31. Time from transplantation to biopsy ranged from 1 month up to 10 years.
Results: The number of Y-chromosome+/alpha-sarcomeric actinin+ cells in male children transplanted with female hearts was 2.39+1.54 % (range: 0–4.2%). The extent of cardiomyocyte chimerism significantly correlated with the time from transplantation to biopsy sampling ($r^2 = 0.69$, $p = 0.006$; $n = 9$). The calculated contribution of male cardiomyocytes in the female heart per year was 0.36+0.09%. Results were confirmed with troponin I staining ($n = 6$). In biopsies with documented rejection ($n = 3$), cardiomyocyte chimerism was significantly higher.
Consistent with previous reports in adult patients, the incorporation of Y-chromosome+/CD31+cells was higher compared to cardiomyocytes (14.4+4.17%), but did not correlate time dependently.
Conclusion: Circulating cells contribute to the development of cardiomyocytes and endothelial cells in children after heart transplantation. The incidence of repopulating cardiomyocytes continuously increases in a time dependent manner (approximately 0.4% Y-chromomsome+cardiomyocytes/year) and resembles cardiac regeneration activity observed in adults.

PW1-10

Blood pressure independent structural abnormalities of retinal arteries in patients after coarctation repair

Hager A. (1), Preßler A. (2), Esefeld K. (2), Kotliar K. (2,3), Lanzl I. (3), Halle M. (2), Kaemmerer H. (1), Schmidt-Trucksäss A (2).
Department of Pediatric Cardiology and Congenital Heart Disease, Deutsches Herzzentrum München, Technische Universität München (1); Department of Preventive and Rehabilitative Sports Medicine, Technische Universität München (2); Department of Ophthalmology, Klinikum rechts der Isar, Technische Universität München (3)

Coarctation of the aorta is not only a simple local defect but includes structural and functional changes in other vessels. Studies have shown an impaired endothelial function in peripheral vessels, a key event in beginning atherosclerosis. We performed static and dynamic retinal vessel analysis to investigate whether there are structural and functional abnormalities in cerebral vessels as well.
Methods: 34 patients after repair of aortic coarctation (22 male; 23–58 years old, 0–32 years after surgical repair) and 34 age and sex matched controls underwent retinal vessel analysis with the Dynamic Vessel Analyzer (Imedos, Jena, Germany). By static analysis, the arteriolar-to-venular-ratio (AVR) was calculated. The average arterial diameter was summarized as central retinal arteriolar equivalent (CRAE). For dynamic analysis, the dilatation of retinal vessels in response to flicker light stimulation was studied. Blood pressure was measured ambulatory, defining mean values > 135/85 mmHg during daytime or > 120/75 mmHg at night or antihypertensive drug treatment as hypertensive.
Results: In static analysis, patients after coarctation repair showed a significant reduction of AVR compared to the control subjects (mean $0.80 \pm$ SD 0.06 vs. 0.87 ± 0.04; $p < 0.001$). This was caused by a reduced diameter of retinal arteries represented by CRAE (184 ± 14 µm vs. 199 ± 12; $p < 0.001$), whereas the venules showed no significant changes ($p = 0.42$). By subdividing the patients into a hypertensive (HT) and non-hypertensive (NT) group, no influence of blood pressure on AVR ($p = 0.91$) and CRAE ($p = 0.47$) was detected. Also, there was no correlation between AVR or CRAE and the age at intervention. In dynamic analysis, there were no significant differences between patients and controls (mean maximum dilatation to flicker related to baseline; $p = 0.44$) and between sub-groups (HT vs. NT: $p = 0.98$).
Conclusions: Retinal artery diameter, a inverse marker for endothelial dysfunction and cardiovascular mortality, is significantly reduced in patients after coarctation repair independent of persisting high blood pressure and age at intervention. Therefore, the results give further evidence for the systemic character of aortic coarctation including cerebral vessels and may contribute to explain the increased rate of cerebrovascular events in affected patients.

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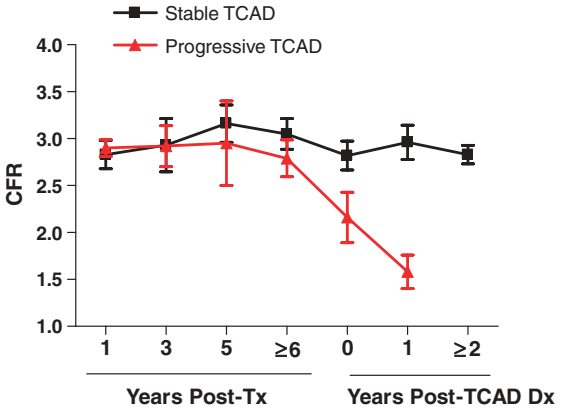
PW2-1

Coronary flow reserve can detect progressive transplantcoronary artery disease in pediatric heart transplant recipients

Tissot C. (1), Pietra B.A. (1), Boucek M.M. (2), Gilbert D.J. (1), Campbell D.N. (1), Mitchell M.B. (1), Ivy D.D. (1), Miyamoto S.D. (1)
(1) The Children's Hospital, Denver, Colorado, US (2) Joe DiMaggio Children's Hospital, Hollywood, Florida, US

Introduction: Transplant coronary artery disease (TCAD) represents the largest cause of late graft loss and the most frequent indication for retransplantation in pediatric recipients. TCAD is often asymptomatic and can present with sudden death. Coronary flow reserve (CFR) measures the functional status and microcirculation of epicardial coronary arteries.
Methods: We retrospectively reviewed pediatric heart transplant recipients who had invasive CFR measurements recorded at the time of routine surveillance catheterizations. CFR is defined as the ratio of hyperemic (following intracoronary adenosine) to basal peak Doppler flow velocity (normal ≥2.5). Patients with and without TCAD were compared. Patients with TCAD were separated into two groups: (1) stable TCAD- no change in angiography or IVUS severity after serial evaluation and (2) progressive TCAD- worsening disease or death as a result of TCAD.
Results: 486 measures of CFR were obtained in 176 patients. Forty-six (26%) were diagnosed with TCAD. At the time of diagnosis, CFR was significantly lower but still within the normal range in patients with TCAD compared with no TCAD (2.7 ± 0.6 vs. 3.2 ± 0.6, p≤0.01). Thirty-six patients (72%) had stable and 14 (28%) had progressive TCAD. There was no difference between groups in time from transplant to diagnosis of TCAD (5.8 ± 2.8, stable vs. 6.9 ± 4 years, progressive), total ischemic time, gender, age at transplant, weight ratio or number of rejections. CFR in the progressive group was lower at the time of diagnosis (2.2 ± 0.6 vs. 2.8 ± 0.8, p = ns) and showed a progressive decline compared to the stable group (1 year post-TCAD diagnosis, 1.6 ± 0.4 vs. 3.0 ± 0.8, p ≤ 0.01, cf graph). No patient in the stable group died or required retransplantation as a consequence of TCAD, while all patients in the progressive group died (60%) or required retransplantation (40%). The survival time after diagnosis of TCAD was significantly lower in the progressive group (1 ± 1.7 vs. 6.2 ± 3.8 years, p ≤ 0.01).
Conclusion: CFR remains within the normal range in pediatric recipients with stable TCAD but shows a steady decline over time in patients with progressive TCAD. Increased surveillance and a lower threshold to proceed with retransplantation in patients with an abnormal or decreasing CFR could improve survival in this subset of patients.



PW2-2

Reduced aortic elasticity and dilatation are associated with aortic regurgitation and LV systolic dysfunction in Tetralogy of Fallot after pulmonary valve replacement.

Grotenhuis H.B. (1,2), Ottenkamp J. (1,2,3), Bruijn de L. (1), Westenberg J.J.M. (1), Bax J.J. (1), Kroft L.J.M. (1), Roos de A. (1)
Leiden University Medical Center, Leiden, The Netherlands (1), Emma Children's Hospital / AMC, Amsterdam, The Netherlands (2), VU Medical Center, Amsterdam, The Netherlands (3)

Introduction: Intrinsic pathology of the aortic wall is a possible explanation for reduced aortic elasticity and aortic dilatation in patients with Tetralogy of Fallot (TOF). The relationship between aortic wall elasticity, aortic dimensions, aortic valve competence and LV function in patients with TOF after pulmonary valve replacement (PVR) has not previously been studied with magnetic resonance imaging (MRI).
Methods: MRI was performed in 16 patients with TOF after PVR (mean ± SD age (yrs.): 31.2 ± 15.5) and 16 matched controls.
Results: TOF patients showed reduced aortic elasticity as indicated by increased pulse wave velocity (PWV) in the aortic arch (5.5m/s ± 1.2 vs. 4.6m/s ± 0.9, P = 0.04) and reduced aortic root distensibility (1.4*10-3mmHg-1 ± 1.7 vs. 5.9*10-3mmHg-1 ± 3.6, P < 0.01). In addition, TOF patients showed aortic root dilatation as compared to controls (mean difference 7.8–8.8mm, P≤0.01 at all 4 predefined levels). Minor degrees of aortic regurgitation (AR) were present in 7 patients (AR fraction 6.0% ± 2.1 vs. 1.2% ± 1.6, P < 0.01). LV ejection fraction was reduced (51% ± 8 vs. 58% ± 7, P = 0.01), whereas RV ejection fraction was within normal limits (47% ± 9 vs. 52% ± 7, P = 0.06). Dilatation at all levels of the aortic root (r = 0.39–0.49, P < 0.05) and reduced aortic root distensibility (r = 0.44, P = 0.02) correlated with AR fraction. Reduced LV ejection fraction was correlated with degree of AR and RV ejection fraction (r = 0.41, P = 0.02, and r = 0.49, P < 0.01, respectively).
Conclusions: Reduced aortic elasticity and aortic root dilatation were frequently present in patients with TOF, related to minor degrees of AR. In addition, reduced LV systolic function was present in patients clinically doing well after PVR, being associated with degree of AR and RV ejection fraction. Monitoring of aortic elasticity and aortic dimensions, in conjunction with aortic valve competence and LV function seems therefore indicated in the long-term follow-up of TOF patients, and should be added to routine RV assessment.

PW2-3

Routine clinical magnetic resonance imaging of patients with congenital heart disease in 2007– a single-center experience

Fratz S., Schuhbäck A., Buchner C., Hendrich E., Martinoff S., Hess J., Stern H.
Deutsches Herzzentrum München an der TUM, Munich, Germany

Background: Magnetic resonance imaging (MRI) of patients with congenital heart disease (CHD) has become routine practice during the last few years. However, virtually all cardiac MRI protocols used worldwide focus on adult patients with ischemic heart disease and no specific protocols for patients with CHD exist. Additionally, it is not clearly defined which patients with which indications should be studied and what type of information should be gathered during routine clinical imaging. Specifically, it is unknown what type of sequences are used in routine clinical MRI of patients with CHD. Therefore the aim of this study was

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to analyse all routine clinical MRIs of patients with CHD during 2007 at our center with specific emphasis on the clinical value.
Methods: The reports of all 362 patients that recieved routine clinical MRI during 2007 at the Department of Pediatric Cardiology and Congenital Heart Disease at the Deutsches Herzzentrum München were reviewed. The following parameters were studied: diagnosis, demografical data, need of sedation/intubation, scan-duration, amount of contrast agent, used sequences and succesfull answering of the question asked.
The sequences used were grouped by the question to be answered: (1) ventricular volumes, (2) flow, (3) unknown anatomy, (4) specific individual morphology of known anatomy, (5) myocardial scar, (6) stress-induced myocardial perfusion defect.
Results: The underlying diagnosis was in 33% Fallot´s tetralogy, 15% aortic coarctation, 8% Morbus Ebstein, 4% Fontan/TCPC circulation, 4% Marfan's syndrome, 4% s/p Mustard/Senning operation, and 32% others. Median age was 26 years (7 days–75 years). Ventricular volumes were assessed in 65% of the patients; flow in 74%; unknown anatomy only in 8%; specific individual morphology of known anatomy in 81%; myocardial scar in 8%; stress-induced myocardial perfusion defects in 1%. Only in 3% of the cases the question could not be fully answered. The main reasons were artefacts caused by coils and arrhythmia.
Conclusions: In contrary to common belief routine clinical MRI of patients with CHD does not address global anatomical questions, but specific individual morphology and function of known anatomy. The protocols used differ strongly from cardiac MRI protocols focussing on adults with ischemic heart disease.

PW2-4

3-dimensional plastic heart models for planning of surgical procedure in complex congenital heart disease

Riesenkampff E. (1), Rietdorf U. (2), Wolf I. (2), Schnackenburg B. (3), Hübler M. (4), Alexi-Meskishvili V. (4), Berger F. (1), Kühne T. (1)
German Heart Institute Berlin, Department of Pediatric Cardiology and Congenital Heart Disease, (1); German Cancer Research Institute Heidelberg, Department of Medical and Biological Informatics (2); Philips Medical Systems, Hamburg, Germany (3); German Heart Center Berlin, Department of Cardiothoracic Surgery (4)

Background: Planning of the optimum surgical treatment strategy is complicated in some congenital heart diseases due to very complex anatomy.
Standard imaging methods, including echocardiography, x-ray angiography, CT or MRI do not always provide the means for satisfying decision making. However, three-dimensional (3D) cast models of the heart, that are made on the basis of CT- or MRI-data, provide detailed information about intra- and extracardiac anatomy and can help to ease surgical decision making. We now present our first clinical experiences with these heart models.
Methods: In 7 patients (age 5–27 years) with complex congenital heart disease, consensus about the optimum surgical strategy was not reached using standard diagnostic tools as mentioned above. Surgical questions were directed towards uni- or biventricular/palliative or corrective surgery (n = 6) and in patch-closure of a swiss cheese VSDs (n = 1).
In these patients, cast models of the heart were made, whereas intracardiac anatomy could be inspected through "viewing windows". The MRI-protocol included 3D, high-resolution free breathing navigator-triggered whole-heart (3DWH) and/or breathhold multiphase-multislice-cine sequences.
Results: Technical aspects: 3DWH-sequences gave good contrast in soft tissue anatomy in patients with regular breathing (e.g. sedated patients) and a heart-rate < 90/min. Cine MRI with breathhold showed advances in patients with irregular breathing and higher heart rates and thus short diastolic phase.
Clinical aspects: In the surgical consensus conference, the cast models were judged helpful by the responsible participants. The following decisions were made, using the cast models in conjunction to standard diagnostic tools: implantation of homograft in one patient with congenital corrected TGA, VSD and pulmonary atresie; patch-closure of swiss-cheese-VSDs from the left ventricle; total cavo-pulmonary anastomosis in 2 patients where there was no option for biventricular corrective surgery. In 3 patients, no surgical option was seen and palliative therapy was favoured, for example in one patient with double outlet right ventricle, atrio-ventricular septal defect and multiple VSDs.
Conclusions: In patients with complex congenital heart disease, realistic 3D cast models of the heart provide invaluable information that can be of crucial importance in surgical decision making.

PW2-5

Pulmonary artery growth after the Norwood procedure. Does the right ventricle to pulmonary artery conduit make any difference?

Maschietto N. (1), Cacace AE. (1); Reffo E. (1); Vida V.L. (2); Padalino M.A. (2); Speggiorin S. (2); Stellin G. (2); Milanesi O. (1)
Department of Pediatrics, Cardiology Unit, University of Padua-Medical School, Italy (1); Institute of Cardiovascular Surgery, Pediatric Cardiac Surgery Unit, University of Padua-Medical School, Italy (2)

Background: The so called Sano modification (right ventricle to pulmonary artery conduit) of the Norwood procedure has dramatically changed the prognosis of newborns with hypoplastic left hear syndrome (HLHS) at least at the time of the first palliative surgery. We tried to investigate how a pulsatile flow could influence the growth of the pulmonary artery and if there were any differences in the growth of the pulmonary arteries between a classical and a modified Stage 1 operation.
Methods: In our Institution, between January 1994 and February 2007, 78 patients (pts) underwent a surgical palliation for HLHS. We retrospectively reviewed all the angiographic data obtained by cardiac catheterization before the cavo-pulmonary anastomosis. We measured the proximal (P) and distal (D) diameter of the right and left pulmonary arteries (RPA and LPA) and of the descending aorta (DAo) at the level of the diaphragm. The McGoon indexes were then calculated.
Results: 78 pts underwent a Norwood procedure for HLHS. 51 had a classical Norwood Stage 1 (group 1) and 27 had a Sano modified Norwood (group 2). 19/51 pts in group 1 and 20/27 pts in group 2 underwent a cardiac catheterization just before the second palliative surgery. The two groups were not statistically different in terms of age, body surface area and gender distribution. The McGoon index calculated at the level of the peripheral pulmonary arteries was comparable between the two groups. Calculated at the level of the distal pulmonary arteries (DPA) resulted higher in group 2 (p 0.0039). The ratio between PRPA and DAo and between PLPA and DAo were comparable in the two groups. Both the ratio between DRPA and DAo and between DLPA and DAo were higher in group 2 (respectively p 0.049 and p 0.0024). In group 2 there were no differences in the growth of the two DPA whereas in group 1 the DRPA showed bigger diameters than the DLPA (p 0.0012).
Conclusion: The pulsatile flow of the Sano modification seems to be able to promote a better and a more symmetrical growth of the pulmonary artery.

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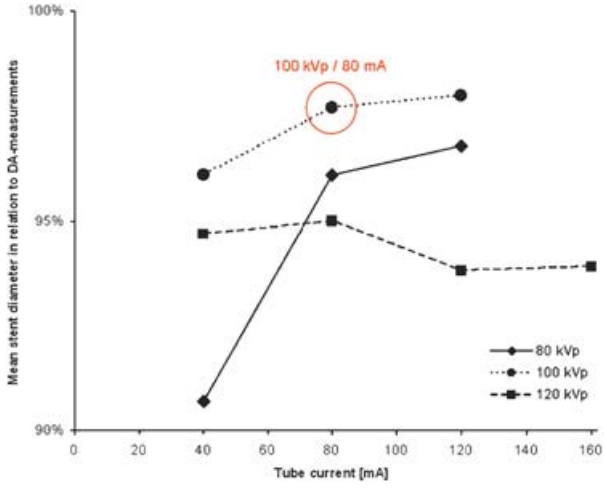
PW2-6

CT imaging of pediatric vascular stents used to treat congenital heart disease

Eichhorn J.G. (1, 2), Raman S.V. (3), Ulmer H.E. (1), Long F.R. (4), Cheatham J.P. (5)
University Children's Hospital, Pediatric Cardiology, Heidelberg, Germany (1); Ohio State University College of Medicine, Radiology (2) and Cardiology (3); Columbus Children's Hospital, Children's Radiological Institute (4) and Pediatric Cardiology (5) Columbus, Ohio, United States

Objective: To assess the visibility of lumen narrowing of pediatric vascular stents using various CT dose parameters in an in vitro model.
Materials and Methods: Ten steel stents of varying design and size commonly used in the treatment of congenital heart disease were implanted in PVC-tubes and were partially obstructed with wax by 25% (mild) to 60% (moderate) of the lumen and filled with contrast material. The stents were scanned on a 64-slice multi-detector CT at tube voltages (kVp) of 80, 100, 120 and tube currents (mA) of 40, 80, 120 and 160. CT-measurements of inner-stent diameter, strut thickness, and percent lumen (in-stent) stenoses were compared to biplane fluoroscopy of digital angiography (DA).
Results: The stent diameter and percent stenosis on all CT images were consistently smaller than measured on DA but were highly correlated ($r = 0.97$, $P < .0001$) with improvement as stent diameter increased (93% agreement with DA for 4 mm- up to 99% for 25 mm-stent; $P = .001$). Moderate stenosis could be assessed better than mild stenosis (99% versus 91% agreement with DA; $P = .003$). Increasing exposure settings improved CT correlations of all measurements for mA up to 120 and kVp up to 100 (98.1% agreement). Higher settings did not improve accuracy (93.9% for 160mA/120kVp; $P = .03$).



Conclusion: CT is feasible to assess lumen narrowing of pediatric vascular stents at a wide range of tube settings. The study suggests that is possible to lower the radiation exposure settings without loss in image quality and accuracy detecting in-stent stenosis.

PW2-7

Intraoperative stenting of pulmonary arteries in complex congenital heart disease

Haas N.A. (1), Blanz U. (2), Schaeffler R. (1), Schulz U. (2), Matthies W. (1), Sarikouch S. (1), Laser T. (1), Kececioglu D. (1)
Department for Congenital Heart Defects (1), Cardiothoracic Surgery (2) Heart and Diabetes-Centre Nordrhein Westfalen, Ruhr-University Bochum, Bad Oeynhausen, Germany

Introduction: Complex congenital heart disease often requires extensive surgical reconstruction and remodelling of pulmonary arteries, especially in patients with additional arch reconstruction. In patients with defects like TOF, PA and VSD or TAC and similar lesions, hypoplastic pulmonary arteries may exist necessitating patch augmentation or other reconstructive efforts. We present our experience of intraoperative stenting to improve perioperative anatomy and outcome.
Methods: During the past 4 years intraoperative stenting of 23 pulmonary arteries was performed in 21 patients. The preoperative diagnoses were univentricular physiology and stenting of 10 pulmonary arteries during Glenn or Fontan procedures. Mostly the relief of compression or secondary stenosis after initial reconstruction after extensive arch reconstruction was required. In 11 patients stenting was performed to secure perfusion of severely hypoplastic or stenotic pulmonary arteries in patients with TOF, PA and VSD with or without unifocalisation or TAC. In most of the patients previous reconstructive surgery was performed.
After preparation according to catheter, ultrasound and MRI data additional direct intraoperative measurement was performed to optimize stent length and inflated diameter. The stents were introduced under direct vision, inflated and the proximal ends secured by surgical fixation. In the last 7 patients, manual flaring of the stent was performed to facilitate easier access for future stent redilatation. Initially we used Palmaz stents, but in the past 2 years (14 patients) CP stents were used solely.
Results: Catheter re-examination was performed at the first post-OP day after Glenn procedures. In one patient a second stent had to be inserted due to distal stent migration; in this patient surgical fixation has not been performed intraoperatively. In the other patients, catheter examination was performed before discharge and showed good results.
Conclusion: We believe that intraoperative stenting of pulmonary arteries is feasible and a very helpful tool to reduce the surgical procedure time and improve outcome in these complex patients. We would like to encourage others to use this strategy more frequently. We emphasize the close collaboration of the surgical and interventional team to facilitate rapid decision finding and good outcome.

PW2-8

Stent fractures after percutaneous pulmonary valve implantation (PPVI): impact of pre-stenting of the right ventricular outflow tract

Nordmeyer J., Lurz P., Coats L., Khambadkone S., Taylor A.M., Bonhoeffer P.
UCL Institute of Child Health and Great Ormond Street Hospital for Children, London, UK

Introduction: Stent fractures are the most common complication after percutaneous pulmonary valve implantation (PPVI), with an incidence of 20%. A previous risk factor analysis demonstrated that calcifications in the right ventricular outflow tract (RVOT), as a surrogate for increased RVOT rigidity, prevented from this complication. We sought to analyze, whether RVOT stent placement prior to PPVI reduced the incidence of stent fractures.
Methods: We retrospectively reviewed the outcomes of 194 patients who underwent PPVI between September 2000 and January 2008 as treatment for RVOT dysfunction. All patients were assessed before and after PPVI according to a detailed protocol that included clinical assessment, antero-posterior and lateral chest X-ray and trans-thoracic echocardiography (VIVID 7, GE, Medical Systems, Milwaukee, Wis.). Fisher's exact test and Kaplan-Meier analysis were used and a P-value of <0.05 was considered statistically significant.
Results: 49/194 (25%) of the patients had RVOT stent placement prior to PPVI. The incidence of stent fractures was lower in this group

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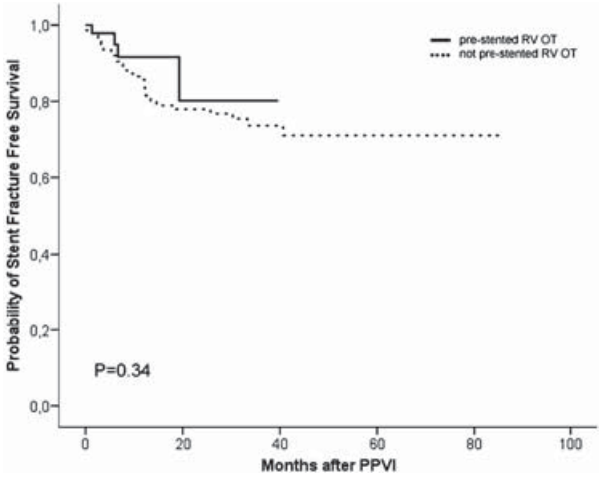
compared to the group without previous RVOT stent placement (8% vs. 22%, P < 0.05). On Kaplan-Meier analysis, there was a trend towards higher probabilities for stent fracture free survival, although this was not statistically significant (at 2 years: 91.6% vs. 85.7%, P = 0.34, Figure). This is likely due to both, the imbalanced number of patients in the two groups and significant differences in mean follow-up (with pre-stenting: 10.7 ± 1.3 vs. without pre-stenting: 25.3 ± 1.6 months, P < 0.0001).



Conclusion: The incidence of stent fractures after PPVI is lower in patients who had previous stent placement in the RVOT. Therefore, elective pre-stenting with bare-metal stents may be a beneficial strategy to reduce the occurrence of stent fractures after PPVI.

PW2-9

Cutting balloon angioplasty to optimize stenting of stenotic MAPCAs

Haas N.A. (1), Schaeffler R. (1), Blanz U. (2), Matthies W. (1) Sarikouch S. (1), Laser T. (1), Kececioglu D. (1)
Department for Congenital Heart Defects (1), Cardiothoracic Surgery (2) Heart and Diabetes-Centre Nordrhein Westfalen, Ruhr-University Bochum, Bad Oeynhausen, Germany

Introduction: Interventional treatment of stenotic MAPCA´s may facilitate subsequent corrective surgery or optimize oxygenation in patients with complex cyanotic heart disease. Successful stenting of MAPCA´s may be impaired by rigid stenosis resistant to high pressure balloon treatment. We report our experience with cutting balloon angioplasty to facilitate MAPCA stenting after failed balloon angioplasty.
Methods: In 4 patients with pulmonary atresia, VSD and MAPCA-dependant pulmonary perfusion interventional treatment was considered as a part of a planned multistage approach to improve oxygenation. The mean age was 12.7 years, the mean weight 34.3 kg and the transcutaneous saturation at rest was 76%. Initial balloon dilatation was performed but showed rigid stenosis resistant to high pressure balloons. In preparation of stent deployment, initial balloon angioplasty was performed followed by rapid subsequent stent implantation. The size of the cutting balloon was about 80% of the diameter of the stenosis when measured with the high pressure balloon in place. In one patient a slightly oversized diameter was used (110%).
Results: The procedure was successful without complications in the 4 patients where slightly undersized balloons were used. In the last patient, the oversized balloon led to an immediate and significant intramural dissection of the vessel followed by subsequent complete vessel obstruction distal to the implanted stent. In this patient surgery with unifocalisation of the distal vessel was possible and performed successfully. We implanted 3 self-expanding stents and 2 balloon expandable stents in our patients. The saturation increased to 82% and the clinical status as well as the exercise capacity increased dramatically in all patients.
Conclusion: Cutting balloon angioplasty is feasible and a helpful tool to facilitate stenting of rigid stenosis in MAPCA's. A slightly undersized balloon diameter seems favourable to minimize complications especially intramural haematoma. We recommend stenting thereafter to maintain vessel patency.

PW2-10

Impact of induced cardiac arrest in balloon dilatation of aortic valve stenosis

Michel-Behnke, I. (1), Hagel K.-J. (1), Stahnke M. (1), Akintuerk H. (1), Awaad R. (2), Schranz D. (1)
Pediatric Heart Centre, Justus-Liebig University Giessen, Germany (1); Caritas Childrens Hospital Bethlehem (2)

Introduction: Balloon valvotomy is widely used for acute management of congenital aortic valve stenosis; however occurrence of aortic regurgitation (AR) limits its acceptance for long term palliation in childhood. Induced bradycardia or functional cardiac arrest by rapid ventricular pacing (RVP) is described as useful for prevention of AR. We report on patients treated by RVP during aortic balloon valvotomy in comparison to a group with adenosin induced bradycardia and a historical group with continuous beating heart during the procedure.
Patients and Methods: From Jan 1999 – Dec 2007 a total number of 126 patients where treated for aortic valve stenosis (AS) by balloon dilatation in 148 procedures. Out of these all complex AS where excluded and the remaining 128 analyzed in 3 groups: A: normal beating heart, B: Adenosin administration, C: RVP. Balloon dilatation was performed retrogradely in almost all pts. All pts were investigated in conscious sedation, heparinized and with IE-prophylaxis. RVP was performed by a transvenously placed catheter in the right ventricle at rates of 260–320 bpm. Adenosin was administered with a starting dose of 0.1 mg/kg. Balloon inflation was started at decreasing heart rate in group B and after 10 beats of rapid ventricular rate in group C.
Results: In the total cohort BVP was effective in 30/34 in group C; 15 pts of group B and 18 pts of group A were redilated or operated. The pressure gradient could be reduced from 62mmHg to 25 mmHg in group A, from 75mmHg to 31 mmHg in group B, and from 68 mmHg to 33 mmHg in group C. There was no higher incidence of arrhythmia in the RVP-group. With respect to AR, the results were superior in the RVP-Group (see table 1).

Table 1:

	Group A n = 53	Group B n = 41	Group C n = 34
Age	1dd- 23.4 y, m = 2.3 y	4dd-16.5 y, m = 5.9y	2dd-19.6y, m = 6.0y
Z-score aortic valve	2.6 ± 2.14	0,74 ± 2.30	0,82 ± 2.41
Balloon /valve ratio	0.91 (0.70-1.33)	0.9 (0.57-1.27)	0.98 (0.78-1.33)
AR post cath	Grade 0: n = 34.0% Grade 1: n = 47.2% Grade 2: n = 13.2%	Grade 0: n = 19.5% Grade 1: n = 46.3% Grade 2: n = 29.3%	Grade 0: n = 35.3% Grade 1: n = 50.0% Grade 2: n = 2.9%
Follow up	2.3 y (2dd - 6.8 y)	2.3 y (27dd – 6.6 y)	0.81 y (18 dd – 2.3 y)
Surgery for AR	N = 5	N = 3	N = 0

Conclusion: Rapid ventricular pacing during percutaneous aortic valvotomy leads to decreased AR early after the procedure and the same seems to be true in mean term follow up.

PW3-1

Antegrade Selective Cerebral Perfusion: adequate flow rate at deep hypothermia: an intravital-microscopy study on mini-piglets

Ben Mime L. (1), Arnhold S. (2), Fischer J. (3), Addicks K. (2), Wahlers T. (1), Bennink G. (1)
Departments of Cardiothoracic Surgery (1), Anatomy (2) and Experimental Medicine (3) University of Cologne, Germany

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Background: Cerebral flow/metabolism coupling and pressure/flow autoregulation which maintain a constant Capillary Blood Flow (CBF) are lost at temperatures below 22°C. We investigated the effect of DHCA with various ASCP flow rates at capillary level using live-monitoring intravital-microscopy and post-mortem brain histology.
Methods: 4 groups of 6 mini-piglets each underwent DHCA at 15°C on CPB. They received ASCP at 10 ml/kg/min in group 1, 20 ml/Kg/min in group 2, and 40 ml/kg/min in group 3. Group 4 served as control. Indices of cerebral perfusion such as CBF, functional capillary density and NADH were monitored using fluorescence IVM. Cerebral tissue changes were investigated using histology and electron microscopy after brain fixation and isolation.
Results: During cooling toward 22°C there was an intact pressure/flow autoregulation. Cerebral oxygenation was flow dependent. During cooling below 22°C CBF showed the highest velocity in group 3 ($P < 0.01$ group 3 versus group 1; $P = 0.02$ group 2 versus group 1). During rewarming group 1 showed a significantly reduced FCD, and CBF was heterogeneous and significantly reduced. Group 3 showed a cerebral hyperperfusion, which induced capillary leakage by the end of rewarming. Group 2 showed the best balance between CBF and tissue oxygenation. Histological brain mapping of cortex and hippocampus revealed a heterogeneous histological figure with endothelial damage of various severity degrees. A histological score showed best endothelial preservation in group 2. At cellular and nuclear level, the EM was favorable in group 2.
Conclusions: IVM and histological findings demonstrate an adequate cerebral oxygenation and superior brain tissue preservation with an ASCP flow rate of 20 ml/kg/min, which therefore should be the clinical approach of choice.

PW3-2
Early primary repair of Tetralogy of Fallot in neonates and infants less than four months of age
Tamesberger M., Lechner E., Mair R., Hofer A., Sames-Dolzer E., Tulzer G.
Children's Heart Centre Linz, Austria

Introduction: The ideal age of correction of tetralogy of Fallot (TOF) is still under discussion. Early primary repair avoids prolonged right ventricular hypertrophy and hypoxemia as well as the risks associated with systemic to pulmonary artery shunting versus the potential disadvantage of a higher operative risk. The aim of this study was to analyze morbidity and mortality in patients less than 4 months of age undergoing primary repair of TOF, and to assess whether neonates, who needed early repair within the first 3 weeks of life, face an increased risk.
Methods: From 1995 to 2006 90 consecutive patients with TOF and pulmonary stenosis underwent early primary repair at our institution. The patients were analyzed retrospectively in two groups: group A consisted of 17 neonates less than 21 days old, who needed early operation due to duct-dependent pulmonary circulation or severe hypoxemia, group B consisted of 73 infants less than 4 months of age who underwent elective repair.
Results: There was no 30 day mortality; late mortality was 2% (2/90). Both deaths occurred in group B and were not associated to the operation. After a median follow up time of 4.7 years overall survival rate was 98% (88/90). So far 7/88 patients (8%) needed re-operation and 12/88 patients (14%) needed re-intervention, most commonly due to severe left pulmonary artery stenosis. Group A and B did not differ significantly in terms of ICU stay, days of ventilations, overall hospital stay, major or minor complications, or re-operation. Significant differences were found in a shorter aortic cross clamp time ($p = 0.02$), a more frequent use of a transannular patch ($p = 0.045$) and more re-interventions ($p = 0.046$) in group A.
Conclusion: Early primary repair of TOF can be performed safely and effectively in infants younger than four months of age and even in neonates younger than 21 days with duct-dependent pulmonary circulation or severe hypoxemia.

PW3-3
Interventricular Dyssynchrony and RV Dysfunction in Adolescents and Adults with Tetralogy of Fallot (TOF) after Surgical Repair and Right Bundle-Branch Block – Evaluation with 2D-Strain-Imaging
Müller M. (1), Hötzer K. (1), Rentzsch A. (1), Rädle-Hurst T. (1), Böttler P. (2), Stiller B. (2), Vogel M. (3), Vogt M. (3), Lemmer J. (4), Abdul-Khaliq H. (1)
Department of Paediatric Cardiology, University Hospital Saarland, Homburg/Saar, Germany (1); University Hospital Freiburg, Germany (2); German Heart Institute Munich, Germany (3); German Heart Institute Berlin, Germany (4)

Introduction: Patients after surgical repaired TOF frequently have right bundle-branch block with prolonged QRS duration, which contributes to right ventricular dysfunction. The aim of this study was to investigate the presence of cardiac dyssynchrony and to asses the influence on the regional and global myocardial function.
Methods: 51 Patients with TOF were included in this study with a median age of 19,63 years (±7,1) and were compared to an age-matched healthy control group ($n = 36$, 19.44 ± 6.1 J). Myocardial analysis was performed 16.59 years after surgical repair and deformation was assessed by Speckle-tracking-derived Strain (2D-Strain). The interventricular septum, the RV and LV lateral wall were examined in an apical four-chamber view. From each myocardial segment, the time interval between the onset of the QRS-complex and the peak of maximal deformation (strain) was assessed. The delay was quantified by measuring the time difference between the intervals.
Results: Myocardial strain was significantly reduced in patients after TOF repair compared to the normal control group in the RV lateral wall and the IVS ($p < 0.001$). 62.7% of the patients examined showed interventricular dyssynchrony with a delay between RV and LV above the normal range of the control group (mean ± 2SD), which was mainly due to the significant delay of deformation in the RV lateral wall rather than in the LV lateral wall (86.09 ± 26.8 ms). RV dyssynchrony was detected in 50.9% of the patients with a delayed deformation of the RV lateral wall to the septum (87.1 ± 19.76 ms). The correlation of the interventricular delay to the strain was highly significant for the RV lateral wall ($R = 0.595$, $p < 0.001$) and the IVS ($R = 0.425$, $p < 0.001$. The ejection fraction of the RV was significantly reduced in patients with interventricular dyssynchrony ($p = 0.003$). The strain of the IVS correlated significantly to the RVEF ($R = 0.547$, $p < 0.001$) and the LVEF ($R = 0.342$, $p = 0.014$).
Conclusions: A significant cardiac dyssynchrony can be assessed using 2D-Strain-Imaging in patients after TOF repair and right bundle-branch block Delayed deformation of the RV lateral wall is associated with reduced global and regional RV function. Myocardial deformation of the IVS had a significant influence on global systolic function of the RV and the LV.

PW3-4
Pulse oximetry screening (POS) in newborns can reduce the diagnostic gap in critical congenital heart disease (cCHD) – First results of a prospective multicenter trial
Riede F.T. (1), Dähnert I. (1), Wörner C. (1), Kostelka M. (2), Schneider P. (1)
Department of Paediatric Cardiology (1), Department of Paediatric Cardiac Surgery (2), Heart Center, University of Leipzig, Germany

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Introduction: Neonates with cCHD may be free of symptoms for a short period after birth leading to a diagnostic gap. The first clinical manifestation may be cardiac collapse with the risk of death or adverse neurological long term outcome even after successful resuscitation. In some recent single center studies, POS has been proposed as an effective, non invasive, inexpensive tool allowing earlier diagnosis of cCHD.
Aim of the study: To test the hypothesis that POS can reduce the diagnostic gap in cCHD in daily clinical routine in the setting of tertiary, secondary and primary care centers.
Methods: Prospective multicenter trial in Saxony, Germany. POS was performed in healthy term newborns at the age of 24–72 hours. Newborns with a prenatal diagnosis or postnatal clinical signs of CHD have been excluded. An oxygen saturation (SpO2) of ≥96% on lower extremities was defined as normal. If a SpO2 of ≤95% was measured and confirmed after 1 hour, complete clinical examination and echocardiography were performed.
Results: From July 2006 until June 2007 19668 newborns from 26 institutions have been included in the study. 33 children were excluded due to prenatal diagnosis (n = 22) or clinical signs of cCHD (n = 11) before POS. 406 newborns not receive POS, mainly due to early discharge after birth (n = 358, 88%). In 19262 newborns POS was performed. POS was true positive in 7 (TAPVD n = 4, HLHS n = 1, TGA-VSD n = 1, Taussig-Bing-Syndrome n = 1), false positive in 10 (healthy n = 5, PPHN n = 5), true negative in 19242 and false negative in 3 (CoA-VSD n = 1, CoA n = 2) children. Sensitivity, specificity, positive and negative predictive value were 70%, 99.95%, 41.18% and 99.98%, respectively.
Conclusions: POS can reduce the postnatal diagnostic gap in cCHD. POS can also detect children with PPHN. False positive results leading to unnecessary evaluation of healthy newborns are rare. POS should be proposed as a routine procedure in postnatal care.

PW3-5
Maternal methylation status, ageing and the risk of congenital heart defects

van Driel L.M.J.W. (1,2), de Jonge R. (3), Helbing W.A. (2), van Zelst B.D. (3), Ottenkamp J. (4), Steegers E.A.P. (1), Steegers-Theunissen R.P.M. (1,2,5,6)
Departments of Obstetrics and Gynaecology/Division of Obstetrics and Prenatal Medicine, Erasmus MC, University Medical Centre, Rotterdam, The Netherlands. (1), Paediatrics/Division of Paediatric Cardiology, Erasmus MC, University Medical Centre, Rotterdam, The Netherlands (2), Clinical Chemistry, Erasmus MC, University Medical Centre, Rotterdam, The Netherlands (3), Department of Paediatric Cardiology CAHAL-Centre for congenital Anomalies of the Heart Amsterdam/Leiden, Leiden University Medical Centre, Leiden, The Netherlands (4), Clinical Genetics, Erasmus MC, University Medical Centre, Rotterdam, The Netherlands (5), Epidemiology, Erasmus MC, University Medical Centre, Rotterdam, The Netherlands (6)

Objectives: Maternal hyperhomocysteinemia is a risk factor for congenital heart defects (CHDs). It is not clear whether homocysteine or the accompanying hypomethylation, reflected by low S-adenosylmethionine (SAM) and high S-adenosylhomocysteine (SAH), is detrimental. Therefore, we investigated the association between these biomarkers of the maternal methylation status and CHD offspring.
Methods: As part of a case-control study, we studied 231 case mothers of a child with a complex CHD, and 315 control mothers of a nonmalformed child at 15 months after the index-pregnancy. The case group was analyzed and stratified into isolated (n = 180) and non-isolated CHDs (n = 51). The latter subgroup was further subdivided into non-syndromic (n = 20), Down syndrome (n = 19) and other syndromes (n = 12). Homocysteine, SAM, SAH and SAM/SAH ratio were determined in maternal blood. A multivariate General Linear Model was used to test for differences between cases and controls. Odds ratios (OR) and 95% confidence intervals (CI) were computed using logistic regression analysis.
Results: Homocysteine was significantly different between the total case group (median (range) 10.3 (4.0,43.8) p-value = 0.026), non-isolated cases (11.1 (5.5,43.8) p-value = 0.006) and controls (10.0 (5.3-42.0)). The results of the subgroup analysis of Down syndrome are shown in the table. The significant risk estimate for the SAM/SAH ratio disappeared after adjusting for maternal age.
Conclusions: Maternal hyperhomocysteinemia is a strong risk factor for having a child with CHD. Moreover, maternal hypomethylation may be associated with offspring with both CHD and Down syndrome. Because of confounding by maternal age, hypomethylation can be considered as feature of ageing.

Concentrations and risk estimates of biomarkers

	Down syndrome (n = 19)	Controls (n = 315)†	P-Value‡	OR(95% CI)‡	P-value	OR(95% CI)#	P-value
SAM(nmol/L)	79.9(60.6,150.2)	80.3(41.7,121.2)	n.s.	1.02(0.99-1.06)	n.s.	1.01(0.97-1.05)	n.s.
SAH(nmol/L)	16.0(10.6,95.8)	14.0(7.2,34.2)	.001	1.10(0.99-1.23)	n.s	1.08(0.97-1.21)	n.s.
SAM/SAH ratio	5.6(0.6,6.7)	5.9(2.2,11.6)	.007	.69(.48-.996)	.047	.75(.52-1.08)	n.s.
Homocysteine (μmol/L)	11.1(6.7,43.8)	10.0(5.3,42.0)	.034	1.08(1.01-1.16)	.037	1.11(1.03-1.20)	.009

Values are median(range).
†Homocysteine: Controls (n = 310).
‡Adjusted for maternal B vitamin use at the study moment.
#Adjusted for maternal B vitamin use and age at the study moment.

PW3-6
Effects of Long-Term Bosentan in Children with Pulmonary Arterial Hypertension

Ivy D.D. (1), Barst R.J. (2), Doran A.K. (1), Lemarie J.-C. (3), Brand M. (4), Rosenberg D. (4), Rosenzweig E.B. (2)
University of Colorado/Children's Hospital, Denver CO, United States (1); New York Presbyterian Hospital, New York NY, United States (2); Effi-stat, Paris, France (3); Actelion Pharmaceuticals Ltd, Allschwil, Switzerland (4)

Objectives: In adults with pulmonary arterial hypertension (PAH), bosentan improves long-term outcome; limited data are available in children. In a previous analysis of a retrospective cohort study of paediatric PAH patients (Rosenzweig et al, 2005), one and two year survival estimates (n patients at risk) were 98% (63) and 91% (10), respectively; in this analysis, we describe disease progression, long-term outcome and safety after an extended follow-up period.
Methods: Paediatric patients started first-time bosentan with or without chronic epoprostenol/treprostinil (epo/trep) between May 2001 and April 2003, and were followed for up to 5 years until August 2006. Demographic, clinical and laboratory data were abstracted from the medical chart.
Results: Overall, 86 patients started bosentan: 49 (57%) females; mean (± SD) age at bosentan start 11 ± 5 years; mean (± SD) age at diagnosis 5 ± 5 years. PAH was idiopathic (36 patients, 42%), related to congenital heart disease (48 patients, 56%) or connective tissue disease (2 patients, 2%). WHO functional class (FC) I, II, III and IV at bosentan start was 7%, 44%, 41%, 7%, respectively. 42 patients (49%) started bosentan as monotherapy, and 44 patients (51%) added bosentan to epo/trep. Median follow-up was 30 months (range 2 to 60 months); median exposure to bosentan was 24 months (range 2 to 60 months).
At end of follow-up, 25 (29%) patients continued bosentan. Five (6%) patients discontinued bosentan due to liver function test abnormalities. Kaplan-Meier (K-M) estimate of disease progression (defined as death, atrial septostomy, transplantation, increase in WHO FC, 15% decrease in 6MWD, addition of PAH-specific therapy for deterioration, or medical judgement to discontinue) was 53% at 3 years (20 patients at risk). K-M estimates of survival

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in patients on bosentan (n patients at risk) at 1, 2, 3 and 4 years were 98% (69), 88% (42), 82% (27) and 82% (16), respectively.
Conclusions: Extended follow-up of paediatric PAH patients treated with bosentan as part of current treatment regimens, indicates absence of disease progression at 3 years in ~50% of patients with favourable estimates of survival, as previously observed.

PW3-7
Comparison of Six-minute Walk Test Distance and Cardiopulmonary Exercise Test Performance in Children with Pulmonary Hypertension
Lammers A.E., Diller G.P. (2), Odendaal D. (1), Taylor S. (1), Derrick G. (1), Haworth S.G. (1)
(1) Great Ormond Street Hospital for Children, Pulmonary Hypertension Service for Children, London, UK (2) National Heart and Lung Institute, London, UK

Introduction: Six minute walk test distance (6MWT) is an established measure of exercise capacity and related to prognosis in patients with pulmonary hypertension. Cardiopulmonary exercise testing (CPET) with metabolic monitoring is considered by many to be the gold standard to assess physical status. Correlations between 6MWT distance and parameters of CPET in children with pulmonary arterial hypertension (PAH) are unknown.
Patients and Methods: 41 exercise studies were included, 15 exercise tests in children with idiopathic pulmonary PAH (mean age 13.0 ± 3.0 years; 9 female), 18 in children with pulmonary hypertension associated with congenital heart disease (mean age 14.8 ± 2.8 years; 7 female) and 8 in children with Eisenmenger syndrome (mean age 11.8 ± 2.9 years; 4 female). All patients underwent a CPET and a 6MWT.
Results: Overall peak oxygen consumption (pVO_2) was reduced in pulmonary hypertension patients to 31.5 ± 12.2% of the predicted value ($p < 0.0001$). PeakVO_2 and oxygen consumption at anaerobic threshold (AT) correlated with 6MWT distance ($r = 0.49$; $p = 0.001$ and $r = 0.40$, $p = 0.01$, respectively) while an inverse correlation betweenVE/VCO_2 ratio at anaerobic threshold and 6MWT distance was found ($r = -0.43$; $p = 0.005$). While we could not find evidence of a 6MWT distance "ceiling-effect" in patients with PAH, peak heart rate was higher and oxygen saturation lower during CPET compared to the 6MWT supporting the concept that the 6MWT is indeed a sub-maximal test in patients with PAH (Figure 1).

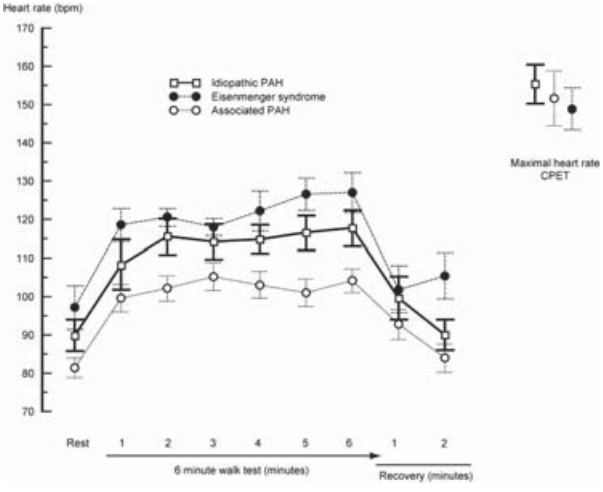
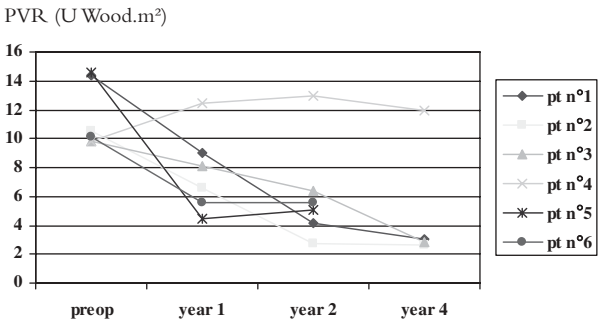


Figure 1 – Heart rate profile and maximal heart rate in children with PAH during 6MWT (left) and CPET (right), showing lower peak HR during 6MWT compared to CPET suggestive of submaximal effort during 6MWT.

Conclusions: Performing a CPET is feasible and safe in older children with pulmonary hypertension. While the 6MWT is easy to perform and can be repeated at low cost, CPET with metabolic monitoring requires expensive equipment, technical expertise and training of the subject. However, CPET provides additional objective measures of exercise capacity, ventilatory efficiency and surrogate parameters of pulmonary blood flow. Further prospective studies are required to assess whether CPET is superior to 6MWT in guiding therapy and assessing prognosis in children in PAH.

PW3-8
Atrial Septal Defect with recent and severe pulmonary hypertension : evolution of pulmonary artery pressure and resistances after closure
Petit J., Houyel L., Capderou A., Roussin R., Belli E., Serraf A.
Marie Lannelongue Hospital, Le Plessis Robinson, France

Introduction: Pulmonary hypertension (PHT) remains the most severe complication of atrial septal defect (ASD). To close or not to close in this case is still a difficult decision. Moreover, the evolution of pulmonary artery pressure (PAP) or, better, pulmonary vascular resistances (PVR) after suppression of the left-to-right (LR) shunt is not clearly known. The aim of this study was to clarify this natural evolution, using iterative measurement of PVR with Fick method and oxygen consumption measurement.
Methods: From 01/01/98 to 31/12/2004, 861 consecutive ASD were closed in Marie Lannelongue Hospital, 488 percutaneously and 373 surgically. Among them, 6 patients (3 ostium secundum and 3 sinus venosus) with severe PHT (mean PAP>40 mmHg) and recent dyspnea (< 6 months) underwent surgical (4) or percutaneous (2) closure of their ASD. Criteria for operability were determined on cardiac catheterization: PVR < 15 U Wood.m2 and persistence of significant LR shunt (QP/QS>2.0). Age at ASD closure was 19–66 years (m = 34). Mean PAP was 41–54 mmHg (m = 46), PVR was 9.8–14.6 U Wood.m² (m = 11.5). ASD closure was uneventful in all. None received medical therapy for PHT either before nor after closure. PVR were measured at post-closure year 1, 2 and 4.
Results: All but one patients felt dramatic improvement of dyspnea during the months following ASD closure. Mean PAP decreased from 46 to 22.7 mmHg. The evolution of PVR is described on Table 1: all but 1 experienced progressive decrease of PVR. Pregnancy was authorized in Pat n°1 four years after closure, with a normal course and persistence of normal PAP 6 months after delivery. Pat n°4 was 42-yrs-old at ASD closure and the beginning of the dyspnea was less clear-cut, letting suppose that PHT could have been not so recent.
Conclusions: In selected cases, ASD closure is possible even if mean PAP is > 40 mmHg. Decrease of PAP after closure is not only due to suppression of the LR shunt but also to the decrease of PVR. This decrease is progressive with time, without any specific medical treatment. Pregnancy can be authorized after normalization of PVR.



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PW3-9

Improvement in long-term survival but not in freedom from reoperations after the change from atrial to arterial switch for transposition of the great arteries

Hörer J. (1), Cleuziou J. (1), Schreiber C. (1), Prodan Z. (1), Vogt M. (2), Holper K. (1), Lange R. (1)
Department of Cardiovascular Surgery (1) and Department of Pediatric Cardiology and Congenital Heart Disease (2), German Heart Center Munich at the Technical University, Munich, Germany

Introduction: Since 1958, patients presenting with transposition of the great arteries (TGA) with intact ventricular septum (IVS), or ventricular septal defect (VSD) were treated by means of an atrial switch operation, like the Mustard (MO) or the Senning operation (SO). In the 1980ies these procedures were abandoned in favour of the arterial switch operation (ASO). From 1969 until today, the Rastelli operation (RO) is applied for patients with additional left ventricular outflow tract obstruction (LVOTO).
Methods: Data of 968 patients operated for TGA between 1974 and 2006 were analyzed. The MO was performed in 88, SO in 329, ASO in 512, and the RO in 39 patients, respectively.
Results:

	MO	SO	ASO	RO
follow-up time	22.6 ± 8.1	18.2 ± 5.7	9.5 ± 5.7	9.9 ± 6.5
hospital mortality	8.0%	4.6%	6.4%	0%
survival at 20 years	75.8 ± 4.6%	88.4 ± 1.8%	93.5 ± 1.1%	57.5 ± 15.1%
freedom from reoperation at 20 years	70.6 ± 5.4%	88.7 ± 1.9%	75.0 ± 6.4%	32.6 ± 10.1%

Stratified by type of operation, the lowest hospital mortality was observed after the SO (4.6%), and the best overall survival at 20 years was observed after the ASO (93.5 ± 1.1%). Stratified by diagnostic groups, the best overall survival at 20 years was observed in patients presenting with TGA+IVS (92.7 ± 1.2%; n = 582), and the lowest in patients presenting with TGA+VSD+LVOTO (69.1 ± 8.2%; n = 75; p < 0.001). Among morphology, prior operations, age, and type of correction, presence of a VSD at the time of operation (HR = 2.8, 95% CI = 1.8–4.2, p < 0.001), the MO (HR = 3.2, 95% CI = 1.8–5.5, p < 0.001), and older age at the time of operation (HR = 1.3, 95% CI = 1.1–1.6, p = 0.004) emerged as independent risk factors for overall mortality.
Stratified by type of operation, the best freedom from reoperation at 20 years was observed after the SO (88.7 ± 1.9%), and the lowest after the RO (32.6 ± 10.1). Stratified by diagnostic groups, the best freedom from reoperation at 20 years was observed in patients presenting with TGA+IVS (87.2 ± 1.9%), and the lowest in patients presenting with TGA+VSD+LVOTO (50.8 ± 7.2%; p < 0.001).
Conclusions: The change in surgical strategy from atrial to arterial switch led to improved long-term survival but not lower incidence of reoperations. Outcome in terms of survival and freedom from reoperation is determined by morphology. The RO yields high reoperation rates.

PW3-10

Aerosolized Iloprost for Pulmonary Vasoreactivity Testing in Children with Long-Standing Pulmonary Hypertension Related to Congenital Heart Disease

Limsuwam A., Wanichkul S., Khosithseth A., Khowsathit P.
Division of Pediatric Cardiology, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand

In children with congenital heart disease and significantly increased pulmonary blood flow, the vascular structure of pulmonary vessels is progressively changed. Changes in the vascular structure may lead to an irreversible pulmonary artery hypertension associated with a poor prognosis. Measuring the capacity of pulmonary vasodilation allows a risk/benefit assessment of surgical intervention. Therefore, assessment of the actual vasodilator capacity of the pulmonary vascular bed is of utmost importance for risk/benefit assessment of surgical intervention in children with congenital heart disease scheduled for surgical correction.
Objective: To investigate whether aerosolized iloprost could be used to access the vasodilator capacity of the pulmonary vascular bed in a selected group of children with a long-standing left-to-right shunt and elevated pulmonary vascular resistance.
Method: Children with long-standing pulmonary hypertension secondary to congenital cardiac anomalies were included in this study. The children were sedated during the cardiac catheterization. Various hemodynamic parameters including pulmonary and systemic blood pressure and arteriovenous differences in oxygen content over systemic and pulmonary vascular beds were measured before and after Iloprost inhalation and vascular resistance and central shunts volumes were calculated according to standard equations. Iloprost was delivery during either controlled ventilation or spontaneously breathing via jet nebulizer (Delphenius™, Italy). The iloprost dosage was calculated to be 0.5 mcg/kg administrated over 10 minutes. A simultaneous decrease in both pulmonary vascular resistance and in Rp/Rs of > 10 % was considered to indicate selective reactivity of pulmonary vascular bed.
Result: Eighteen consecutive children aged 7 months to 13 years, mean age 5.7 years, were enrolled. Twelve children had a positive response, resulting in a mean ± SD decrease in pulmonary vascular resistance of 9.3 ± 4.8 to 4.6 ± 2.9WoodU/m2 (p = 0.001), and in Rp/Rs of 0.54 ± 0.39 to 0.25 ± 0.15 (p < 0.05). Eight of the 12 iloprost responder underwent surgical correction of their congenital heart anomaly.
Conclusion: The capacity of pulmonary vasodilatation as measured by using aerosolized iloprost varies among children suffering from pulmonary hypertension and increased pulmonary vascular resistance. The results of this study suggest, that aerosolized iloprost is useful for a preoperative risk/benefit assessment of surgical intervention in children with long-standing PAH secondary to congenital heart disease.

PW4-1

Safety and efficacy of intravenous amiodarone as therapy for early postoperative tachycardias in children after congenital heart surgery

Trip H. (1), Bokenkamp R. (1), Hogenbirk K. (2), Geukers V. (2), Clur S.A. (1), Hazekamp M.G. (3), Blom N.A. (1)
Departments of Pediatric Cardiology (1) Pediatric Intensive Care (2), Cardiothoracic Surgery (3), Center for Congenital Heart Disease Amsterdam-Leiden, The Netherlands

Postoperative tachycardias after pediatric cardiac surgery can be life threatening and difficult to control. Intravenous (iv) amiodarone therapy is often used as first line therapy.
Objectives: to evaluate efficacy and adverse effects of iv amiodarone in relation to loading and maintenance dose and loading time.
Methods: 57 consecutive children receiving iv amiodarone therapy for postoperative arrhythmias were retrospectively studied during a 5-year period. Iv amiodarone loading dose was titrated individually with 1 mg/kg every 10 minutes up to maximal 10 mg/kg according to the institutional protocol.

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Results: Median age at operation was 3.0 months (range 0.1–12 yrs), median weight 4.8 kg (range 2.7–27). The majority of pts had tetralogy of Fallot/DORV repair (n = 22.38%) or VSD closure (n = 16, 27%). Postoperative tachycardias included junctional ectopic tachycardias (79%), AV reentrant tachycardia (12%), atrial flutter (5%), ventricular arrhythmia (4%). Median onset of tachycardia was 4 hrs (range 0–454), mean maximal heart rate was 223 ± 27 bpm. Amiodarone loading dose was effective in 56/57 pts (98%); median loading dose 4,1 mg/kg (1–15), median loading time 1.5 hrs (0.2–6.5), median time from start of therapy to rate control (< 180 bpm) or conversion to sinus rhythm 3 hrs (0.5–20). The median maintenance dose of amiodarone was 10 mg/kg/day (0–30), median duration of therapy 77 hrs (0.8–324). Recurrences occurred in 34 pts (60%) and were effectively treated with an additional loading dose in 28 pts (median 5 mg/kg). Adverse effects occurred in 19 pts (33%), hypotension in 13 pts (23%) and bradycardia in 8 pts (13%), including transient AV block in 1 pt. Severe adverse effects occurred in 3 pts, including cardiac arrest in 2 pts and torsade des pointes in 1 pt. Rescuscitation was unsuccessful in 1 pt. Three other deaths were unrelated to the use of amiodarone or underlying arrhythmia. In this retrospective analysis success rate or adverse effects were not related to the iv amiodarone loading dose or time.
Conclusions: iv amiodarone is an effective therapy for postoperative arrhythmias after pediatric cardiac surgery but adverse effects frequently occur. Careful titration of iv amiodarone is essential in this group of critically ill children.

PW4-2
VT Ablation in Children – a Single Center Experience
Will J.C., Opgen-Rhein B., Franzbach B., Krings G., Berger F.
Pediatric Cardiology, Charite University Hospital Berlin, Germany

Introduction: Although RFA and cryoablation in children with SVT in common, still there is little published data about VT ablation in this patient age group.
Objectives/Methods: Retrospective analysis of all children and congenital heart disease patients [pts] with attempted VT ablation in our institution with respect to patient demographics, procedural data, outcome and mid term follow up.
Results: From 01.0.5.2001 to 03.01.2008 36 invasive EPS with attempted VT ablations (12.7% of our overall ablation procedures) were performed in 25 pts (male = 13). 28% had structural anomalies /CHD (DORV/TOF = 3, ASD = 1, ARVC = 1, Myocarditis = 2). Mean age was 16.5 yrs (range 0.7–35.9), mean weight 57.2 kg (range 5.0–93). There were 33 VTs in 25 pts (focal RVOT-VT = 14 (1 in aortic cusp), scar related RVOT-VT = 5, fascicular LV-VT = 4, focal non-RVOT-VT/micro-reentry VT = 8, BB-Reentry VT = 1). 44% of the pts showed CHF, one pt was ablated on ECMO. 1 pt with 2 attempted ablations and LV-foci and s/p biopsy-proven myocarditis was later found to be diagnosed Andersen syndrome. 3D mapping systems were used in 28% [CARTO/CARTO merge:9, ENSITE:1]. 28% of the pts had more than 1 VT. 88.9% of the VT were ablated with RFA (mean RF time: 736 secs), 13.9% with cryoenergy ± RFA. Mean fluoroscopy time was 30.5 min, total area dose was 3960 cGycm2. Overall success rate per procedure was 80.6%. There were 9 recurrences in 6 pts., 4 of these pts were ablated successfully during further procedures. There were no procedure related complications. Mid term F/U showed overall success rate of 87.5% (mean F/U time 6.1 months, range 0.1–65).
Conclusion: T ablation in children in young adults with CHD accounts for a substantial group in our pediatric ablation population. Acute success rate and mid term F/U do show a success rate of > 80%, although in 28% of the pts more than 1 ablation procedure had to be performed. In our series VT ablation seems to be a safe and highly effective therapeutic option.

PW4-3
Identification and distribution of cardiac stem cells in the fetal human heart
Quaini F. (1), Graiani G. (1), Boni A. (1), Ferraro F. (1), Delucchi F. (1), Agnetti A. (1), Osmi S. (1), Giordano G. (2), Urbanek K. (3)
Cardiac Stem Cell Center-University of Parma, Italy (1) Dept. of Pathology- University of Parma, Italy (2) Brigham & Women's Hospital, Harvard-Boston USA(3)

Mitotic and apoptotic myocytes are present in the human heart constituting a cell turnover which may be regulated by resident primitive cells. The hypothesis was advanced that during fetal life this cell population and its progeny may actively dictate myocardial development allowing the identification of stem cells and their niches. For this purpose 20 fetal human hearts at 20–22 weeks of gestation were longitudinally sectioned, fixed and processed for the detection by confocal microscopy and fluorescent probes of the different cardiogenic cell populations. Cardiac Stem Cells (CSCs) were defined as a small-undifferentiated cell showing surface stem cell markers (c-kitpos) and absence of neuronal, skeletal muscle and hematopoietic lineage commitment (Linneg). Cardiac Progenitors (CPCs) showed the combination of c-kit with nuclear cardiogenic transcription factors (GATA-4, MEF2C). Cardiac precursors consisted of small cells, lacking stem cell surface antigens, expressing nuclear cardiogenic transcription factors and early muscular or vascular filaments (nestin). Young cycling (Ki67pos, MCM5pos) and mitotic (Ph-H3pos) small cardiomyocytes were considered as transient amplifying precursors. Antibodies against Flk1, Factor VIII, CD34 and CD31 were used to identify vascular progenitors and endothelial cell differentiation. CD45, CD45RO, CD4, CD8, CD20 and glycophorin A excluded myeloid, lymphoid and erythroid cells, respectively. All these different cell myogenic and vasculogenic compartments were detected in the atria and ventricles of the fetal heart. Accumulation of CSCs and CPCs was observed in the atrio-ventricular junction. CPCs and area of intense myocyte proliferation were prominent in the epicardial layers of both side of the heart. However, undifferentiated cells nested in niches, where Notch-1 and its ligand Jagged-1 was operative, were more numerous in the apical aspects of fetal ventricles. Notch intracellular domain (NICD) was also involved in myocyte, vascular and valvular formation documenting that cell fate decision was an essential component of the fetal human heart. Thus, the developing human myocardium contains a resident population of primitive cells provided by phenotypic, functional and anatomical characteristics fulfilling the properties of stem cells. The distribution of all cardiogenic phenotypes strongly suggests that both tissue supply and mechanical factors contribute to the regulation of human cardiac growth.

PW4-4
Presence of PA to RV Regurgitation Minimally Affects Postoperative Hemodynamics of Norwood Procedure: A Theoretical Analysis with Valved RV-PA Conduit
Shimizu S. (1), Shishido T. (1), Kawada T. (1), Sano S. (2), Sugimachi M. (1)
Department of Cardiovascular Dynamics, National Cardiovascular Center Research Institute, Osaka, Japan (1); Department of Cardiovascular Surgery, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama, Japan (2)

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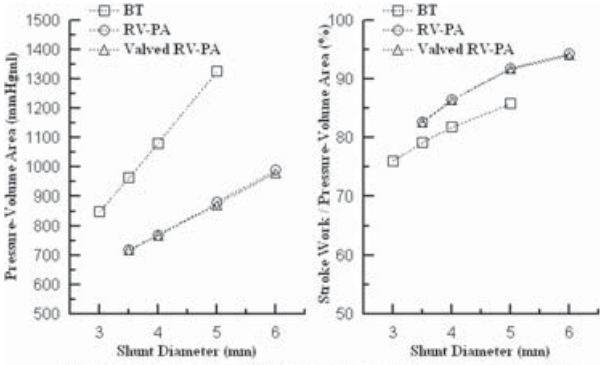
Introduction: For 20 years, a shunt originated from right ventricle rather than Blalock-Taussig shunt (BT) for pulmonary circulation has caused the clinical dramatic improvement in Norwood procedure. To support this, we have already reported that Norwood procedure with right ventricle-pulmonary artery shunt (RV-PA), in spite of the use of non-valved conduit, dramatically improved ventricular energetics compared to BT in a theoretical analysis (ESC Congress '07). But diastolic PA to RV regurgitation may increase RV preload and be deleterious when non-valved RV-PA conduit is used for Norwood procedure. We clarified the impact of the regurgitation from PA to RV by a theoretical analysis using computer models examining the effect of valved conduit.
Methods: Computer models of postoperative cardiovascular dynamics of Norwood procedure with BT, valved and non-valved RV-PA were developed with a time-varying elastance chamber model and modified three-element Windkessel vascular model. Pressure drop across the shunt (ΔP) was described as a non-linear function of instantaneous volume flow (Q) rate and shunt diameter (D) (ΔP = {aQ+bQ^2}/D^4). We estimated systemic and pulmonary blood flow (Qs, Qp), systemic and pulmonary arterial pressure (SAP, PAP), saturation, right ventricular end-diastolic volume (EDV), stroke work (SW), and pressure-volume area (PVA) for various shunt diameters.
Results: Excessive Qp in BT (1.34l/min for 4mm diameter conduit) was ameliorated in both non-valved (1.02 l/min) and valved RV-PA (1.06 l/min). Regurgitation in non-valved RV-PA was 0.05 l/min. RV preload (RVEDV) was smaller in both non-valved (14.8%) and valved RV-PA (15.0%) than that in BT. In both non-valved and valved RV-PA, compared to BT, decreased Qp contributed to the decreased PVA (−28.8% and −28.8%) and increased mechanical efficiency (SW/PVA), irrespective of the presence of small PA to RV regurgitation. The differences between non-valved and valved RV-PA were very small.
Conclusions: Though valved RV-PA improved pulmonary blood supply and it resulted in the increase in Qp, PAP and saturation compared to non-valved RV-PA, the impact on systemic circulation was small. In conclusion, presence of PA to RV regurgitation minimally affects postoperative hemodynamics from the viewpoint of ventricular energetics.



BT, Blalock-Taussig Shunt; RV-PA, Right Ventricle to Pulmonary Artery Shunt

PW4-5

Single-centre experience of evaluation and treatment of 34 consecutive children with idiopathic pulmonary arterial hypertension (iPAH)

Zimmermann R., Ketelhodt G., Kreuder J., Schranz D.
Pediatric Heart Center, University of Giessen, Germany

Introduction: IPAH (idiopathic pulmonary arterial hypertension) is a progressive disease with no cure. Treatment options and prognosis have improved in the last decade, but are not satisfactory. Treatment algorithms are subject to ongoing change with experience and new drugs. We present our single-centre experience with 34 children treated for IPAH between 1999-2007.
Methods: Retrospective review of 34 consecutive IPAH patients. All received right heart catheterisation (RHC) for global vasodilator testing and assessment of endothelium dependant pulmonary flow reserve with acetylcholine (intraluminal flow-wire). Treatment decisions were individually based on primarily testing results.

Drug	n =
CCB (calcium channel blocker) mono	7
CCB including comb. therapies	17
Sildenafil mono	4
Sildenafil including comb. therapies	19
Bosentan mono	0
Bosentan including comb. therapies	13
Prostcyclin total (never mono)	14
inhaled / i.v.	9 /8
Monotherapy	10
Combination of 2 drugs	14
Combination of 3 drugs	9
Combination of 4 drugs	1

Results: Age range at presentation: 2 months–16 years. Survival: 0.5 month–16 years, median observation period 46 month. 10 patients died, 3 underwent heart-lung transplantation (HLTX); 2 received fenestrated ASD closure. 7 underwent atrial septostomy/PFO stenting. At last visit (or before death/HLTX), specific drug therapy for PH was as shown in the table.
At first assessment, baseline PAP/SAP (range): 0.46–1.56; best PAP/SAP during vasodilator testing 0.23–1.41. 10/34 patients were responders (PAP/SAP < 0.4). At last available evaluation, baseline PAP/SAP: 0.41–1.51; best PAP/SAP: 0.24–1.51, responders n = 9. In 66% of patients baseline PAP/SAP before treatment initiation stayed equal or improved compared to PAP/SAP in the latest RHC. 1 non-responder showed response 48 months after treatment initiation. NYHA class improved in 7,unchanged in 13 and worsened in 4 of the survivors.
Conclusions: Our data confirm the high responder rate in children with IPAH, therefore, RHC remains essential to identify children suitable for CCB. With new treatments for IPAH, individualised therapeutic strategies are needed to warrant the best possible outcomes. Significantly improved hemodynamics, as seen in some of our children, are rarely reported in adults and other forms of PAH.

PW4-6

Clinical Features, Etiology and Outcome of Fetal Cardiomyopathies (CM)

Weber R. (1), Kantor P. (2), Chitayat D. (3), Jaeggi E.T. (2)
Department of Cardiology, University Children's Hospital, Zurich, Switzerland (1); Division of Cardiology, The Hospital for Sick Children, Toronto, Canada (2); Division of Clinical Genetics, The Hospital for Sick Children, Toronto, Canada (3);

Background: CM account for a small but clinically relevant portion of prenatally diagnosed cardiac abnormalities. Consequently, information on the disease spectrum and outcome is scarce.
Objectives: To determine disease patterns, mechanisms, hemodynamic findings, and outcome of fetal CM.

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Methods: Review of all cases presenting with suspected myocardial disease at our centre since 2000. Excluded were subjects with major structural heart disease and with reversible CM causes (twin-twin syndrome; maternal diabetes; high output state). Echocardiograms, reviewed by two independent observers, and if available autopsies were used for classification into dilated (D), hypertrophic (H) or restrictive (R) CM and to assess myocardial function. The value of different markers in predicting adverse outcome was tested.
Results: 45 of 8309 pregnancy referrals were diagnosed with fetal CM at a median age of 23 (range: 18–41) weeks. Twenty cases had DCM, including 4 with chromosomal anomalies, 10 with metabolic/genetic disorders and 6 idiopathic cases. Fourteen cases had HCM, including 5 cases with genetic disorders and 9 idiopathic cases. Eleven cases had RCM, including 4 with maternal anti-Ro antibody exposure, 1 with atrial myocardial degeneration; 1 post CMV infection and 4 idiopathic cases. Maternal antibody-induced RCM was treated with steroids and IVIG. Overall, 6 (13%) pregnancies were terminated, 17 (37%) cases died in-utero and 6 (13%) as neonates. Fetal non-survivors were earlier diagnosed (22.5 ± 4.8 vs. 27.0 ± 5.7 weeks; $p < 0.02$), more likely hydropic (8/12 cases vs. 7/27 without hydrops; $p = 0.03$), and had shorter heart-rate corrected AV valve inflow durations (0.36 ± 0.04 vs. 0.42 ± 0.08, $p < 0.02$) than survivors. At a median follow-up of 212 (range: 0.2-2207) days, only 12 (27%) cases remain alive (5 DCM: 24%; 3 HCM: 27%; 4 RCM: 36%), including 2 cases after heart transplantation and 1 case after bone marrow transplantation.
Conclusions: Fetal CM has a wide spectrum of predominantly lethal etiologies. Intrauterine demise, affecting almost 50% (17/39) of ongoing pregnancies with fetal CM, is predicted by the presence of earlier diagnosis, fetal hydrops, and, compatible with severely impaired ventricular filling, by a significantly shortened AV inflow duration at the time of the initial echocardiography.

PW4-7

Premature fetal closure of the arterial duct may result in very different clinical presentations and outcome !

Gewillig M., De Catte L., Boshoff D.E., Van Schoubroeck D., Cossey V., Debeer A., IWitters I., Devlieger R., Lewi L., Delhaas T, Mertens L., Eyskens B.
Fetal & Pediatric Cardiology, Neonatology, Leuven, Belgium.

Background: premature constriction/closure of the duct most frequently occurs subclinically. However severe forms do occasionally present. Clinical presentation and final outcome still needs to be defined.
Methods: retrospective study; fetal and neonatal echo databases were searched for pathology due to or associated with premature closure of duct between 1998-2007. All clinical data and imaging studies were reviewed.
Results: 12patients were identified. 8patients were referred prenatally because of an abnormal fetal 4 chamber view (gestational age: mean 28w, range 20–37.5 w; incidence since 1998: $8/602 = 1.3\%$ abnormal scans); 4patients presented at birth with significant cyanosis; no patient had a patent duct. 4 mothers had taken NSAID during pregnancy. Patients had right ventricular hypertrophy (12), with evolution to bipartite right ventricle (8); significant tricuspid regurgitation (12) with right atrial congestion, hydropericardium (1) and hydrops (1); pulmonary valve stenosis (2) and regurgitation (12) ranging from mild to « agenesis » of pulmonary valve (3); dilation of pulmonary trunk (5) and branch pulmonary arteries, compression of airways with "fluid-trapping" and microcystic malformation of lungs (1); fetal suprasystemic pulmonary hypertension (2); 1 pt had a thrombosed aneurysm of the duct. In 5 patients premature delivery (34–36 w) was chosen to avoid further intra-uterine damage of the right heart & lungs. Neonatal treatment varied from observation (7), ventilation with pulmonary vasodilators (5) and ECMO (1); resection of a thrombosed aneurysmal duct as the thrombus was occluding the left pulmonary artery (1). 3 patients died in the neonatal period because of respiratory insufficiency. Late treatment in the 2 patients with cardiovascular residuae consisted of balloon dilation of PS at 7 months and later closure of ASD a 9 years (1), and homograft reconstruction with PA plasty at 4 years for late compression of coronary artery (1). 2 pts have mild psychomotor delayed development; 1 pt developed non-compaction cardiomyopathy.
Conclusions: Fetal premature closure of the arterial duct can stress at different fetal ages many different levels of the right heart and pulmonary circulation, resulting in a very wide range of secondary pathology. The clinical outcome ranges from normal to death; neonatal death was due to lung damage. Premature delivery might be indicated in selected patients.

PW4-8

Fetal Diagnosis of Congenital Heart Disease by Telemedicine

McCrossan B.A., Sands A.J., Casey F.A.
Department of Paediatric Cardiology, Royal Belfast Hospital for Sick Children, UK.

Introduction: There remains a disparity between, and within, regions in terms of access to fetal cardiology and detection rates of congenital heart disease (CHD). This study aimed to evaluate the feasibility and accuracy of performing remote fetal echocardiograms (FE).
Methods: This was a prospective study over a 16 month period. An initial FE was performed by a radiographer in the district general hospital (DGH) (D1) followed by a second FE transmitted to the regional centre, in real time, via a telemedicine link (384 Kbps) with live guidance by a fetal cardiologist (D2). A FE was performed later at the regional centre (D3, gold standard). The paediatric cardiologist awarded a subjective mark (/10) for each transmitted FE and a checklist was completed detailing 10 components necessary for a comprehensive FE.
Results: 64 remote FE have been performed: 53 normal hearts and 11 with CHD (3 duct dependent) confirmed by D3. D2 was accurate in 97% cases compared with D3 (kappa = 0.90) indicating excellent agreement. No studies were terminated as a result of technical difficulties. All transmissions connected at the first attempt. The mean number of components adequately seen per transmitted FE was 9.6/10. The mean subjective score for overall quality of transmitted FE was 7.4/10 (s.d. = 1.1).
Conclusions: To date this is the largest study of its kind. FE transmitted in real-time with live guidance is feasible and reliably provides the paediatric cardiologist with adequate images on which to make a diagnosis. CHD can be confidently diagnosed and excluded by remote FE. This application of telemedicine could improve access to fetal cardiology, support radiographers screening for CHD and reduce the burden on the limited capacity of tertiary centres.

PW4-9

Developmental outcome after three stage palliation for hypoplastic left heart syndrome: comparison with and without deep hypothermic circulatory arrest

Furck A.K., Schulze V., Drabkin A., Geiger F., Scheewe J., Kramer H.H.
Children Heart Center University of Schleswig-Holstein, Campus Kiel, Kiel, Germany

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Introduction: The treatment of children with hypoplastic left heart syndrome (HLHS) holds the risk of impaired psychomotor development. Antegrade selective cerebral perfusion (ASCP) at the Norwood operation contributed significantly to avoid long duration of deep hypothermic cardiac arrest (DHCA). We evaluated the effect on the neurodevelopment.
Patients and Methods: 53 between 1996 and 2003 born HLHS children were evaluated after their 3 staged palliation with the help of standardized tests for their cognitive (K-ABC) and visual motor integration (VMI) developmental stage. The group without (n = 22) and with (n = 31) ASCP were compared among each other and also with a control group of healthy children. At the same time we used a parent questionnaire to get information about the quality of life (Kindl test) and behaviour problems (CBCL).Possible influence factors such as cardiopulmonary bypass time, circulatory arrest time, complications were checked of significant correlations with the test data.
Results: Both patient groups showed in the "scale of intellectual abilities" and "scale of proficiency" of K-ABC as well as the VMI significantly ($p < 0.01$) lower scores then the control group (see table). In comparison among each other the ASCP group was doing significantly better in all categories.

	SIF	FS	VMI
HLHS DHCA	74 ± 17	71 ± 18	69 ± 25
HLHS ASCP	85 ± 12	82 ± 17	89 ± 14
Control group	103 ± 9	104 ± 10	105 ± 10

Particularly the influence factor cardiac arrest time showed a negative significance for both areas.
All groups had equal good quality of life and no behaviour problems.
Conclusion: Children with HLHS show a cognitive and visual motor integration delay which is significantly less since the implementation of ASCP and the avoidance of circulatory arrest time during the Hemifontan and Fontan operation. Further steps for improving neuroprotection are important goals in treating this complex heart defect.

PW4-10

Exercise Capacity Relates to the c.46A>G Genomic Polymorphism of the Beta2-Adrenoreceptor in Patients after Coarctation Repair

Bildau J. (1), Hager A. (2), Kreuder J. (1), Kaemmerer H. (2), Hess J. (2)
(1) Department of Pediatric Cardiology, Justus-Liebig-Universität, Giessen, Germany; (2) Department of Pediatric Cardiology and Congenital Heart Disease, Deutsches Herzzentrum München, Technische Universität München, Germany

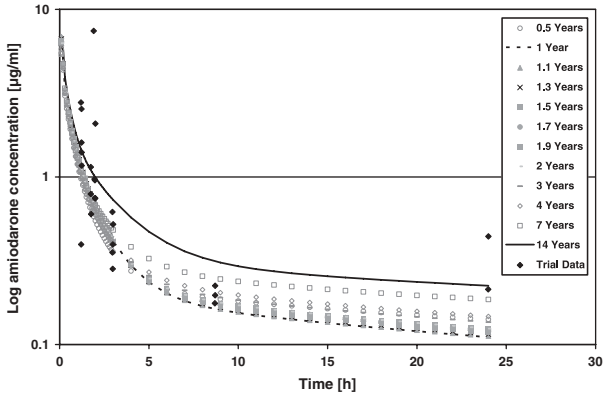
Background: Even after repair of aortic coarctation without restenosis there are limitations in exercise capacity at long-term follow-up independent of the surgical approach and grade of restenosis. This study was performed to assess the contribution of inherited gene polymorphisms to exercise capacity in those patients.
Patients and Methods: 122 patients aged 17–72 years, 46 female, 76 male, 2–27 years after repair of aortic coarctation with a residual brachial-ankle-gradient ≤20 mmHg were investigated. Genomic polymorphism of angiotensin converting enzyme (ACE, intron 16 insertion/deletion), angiotensinogen (AGT, c.704T>C), angiotensin II - receptor type 1 (ATR1, c.1166A>C), aldosterone synthetase (Cyp11B2, c.-344C>T), endothelin 1 (END, ET1 IVS4 c.8002G>A), G protein (GNB3, c.825C>T), fibillin (FIB1, variable nucleotide tandem repeats) and two polymorphisms each of the ß1 adrenoreceptor (ß1AR, c.145A>G, c.1165C>G), ß2 adrenoreceptor (ß2AR, c.46A>G and c.79C>G), and eNOS (ENOS, c.894G>T and ENOS4, intron 4 insertion/deletion) were determined by PCR amplification and fragment length analysis. Exercise capacity was determined by an upright bicycle exercise test and expressed in Watt. The impact of the genotypes was tested in a multiple regression analysis after correcting for body weight, length and age.
Results: Only the c.46A>G polymorphism of the ß2AR showed a significant relation to exercise capacity ($p = 0.028$). Patients with the A/G and especially the G/G genotype showed a significant higher exercise performance.
Conclusions: In contrast to a previous study in heart failure patients, adults after coarctation repair showed a better exercise capacity with the G allele of the ß2 adrenoreceptor c.46A>G polymorphism. Therefore, their exercise capacity profits from the better function of the receptor, whereas patients in heart failure profit from a worse function of their beta receptors.

P-1

Simulation of a virtual paediatric population as a basis for an amiodarone dosing regimen

Ramusovic S., Vogt W., Frobel A. K., Läer S.
Clinical Pharmacy and Pharmacotherapy, University of Duesseldorf, Germany

Introduction and Objectives: Amiodarone still is the most effective antiarrhythmic agent available, especially in children who do not respond to betareceptor blockers, class I antiarrhythmics or sotalol or have specific types of supraventricular tachycardia. There is, however, a severe gap of knowledge on paediatric dosing of amiodarone in various age groups. A novel technique called physiologically based pharmacokinetics (PBPK) may enable us to create a virtual paediatric population and to simulate paediatric drug concentrations based on adult data in a physiological manner. The objective of this investigation was to establish a PBPK-model as a basis for age appropriate intravenous and oral amiodarone dosing regimens in children.
Methods: First, PBPK-based simulations of amiodarone concentration time profiles were conducted using the PK-SIM™ 3.0 software for adult patients. The results were compared with intravenous and oral amiodarone data from literature. After sufficient representation of adult datasets, a scaling step was conducted considering age depending aspects of children's growth. To validate the model it was compared with intravenous amiodarone data from children (n = 20) aged 0.5 to 14 years.
Results: Based upon the adult PBPK-model with fit parameters of logP: 8.5, unbound fraction: $3*10^{-7}$, hepatic clearance 2 ml/kg/min paediatric plasma concentration time profiles were simulated. The diagram shows plasma concentration time profiles of the simulated paediatric population after administration of amiodarone (5 mg/kg,10 min, symbols) and trial data (black diamonds; Saul P.



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et al 2005, Circulation 112, 3470-7). Fast distribution of amiodarone is observed followed by a much slower elimination phase. One year old individuals (dashed line) have an enhanced amiodarone elimination (CLtot: 4.27 ml/kg/min) compared to the other groups. Fourteen year-old children (solid line) show the slowest elimination (CLtot: 2.04 ml/kg/min).
Conclusion: A PBPK-model of amiodarone was developed which reflects the measured plasma concentrations of adult and children datasets. Based on this an age appropriate dosing regimen for children can be established which then needs to be validated.

P-2

Twenty-five years of single centre experience with permanent pacemaker therapy in children

Bhole V., Stickley J., Chikermane A., Ofoe V., Desai T., Mills K., Atkins G., Timmins J., Dhillon R., DeGiovanni J.G.
Birmingham Children's Hospital NHS Foundation Trust, Birmingham, U.K.

Introduction: Pacing in the paediatric population is complicated by small patient size, complex congenital heart disease and need to maintain vascular patency over a lifetime. We share our experience of pacing in a paediatric population over the last 25 years.
Materials and Methods: We retrospectively reviewed pacemaker records of all the patients who had undergone permanent pacemaker implantation at our institute from January 1982 to December 2007.
Results: We evaluated the pacemaker records of all the patients who had permanent pacemakers over the last 25 years. A total of 345 children underwent 541 pacemaker implantations. 664 leads were implanted over this period.
The commonest indication for pacemaker implantation was an end of life of the old system (25%). This was followed by postoperative heart block (22%), congenital heart block (14%) and heart block with congenital heart disease (7%). The remainder were paced for other reasons (19%) including 3 patients who had cardiac resynchronization therapy.
Pacemaker leads were endocardial in 71.6% and epicardial in 28.2% of implants. Of these 205 (37%) were explanted of which 5 were at the time of cardiac transplant. Currently 117 (20%) systems are followed up at our institute and 152 (27%) have been transferred elsewhere.
There were 51(9.2%) deaths during this period. There was no death related to pacemaker implantation. There were 2 deaths attributed to epicardial lead fracture.
There were 14 episodes of pacemaker infection in 13 children. The overall incidence of infection was low (2.5%). Since 1997 we have not encountered any pacemaker infection needing explantation.
On statistical analysis endocardial systems were superior to epicardial systems when assessed by the length of life of the generator ($p < 0.05$ Log rank test).
A total of 125 (18%) leads were explanted. Only one patient who had a transvenous ventricular lead explanted had damage to tricuspid valve requiring surgery.
Conclusion: Endocardial systems are superior to epicardial systems in terms of generator longevity. The risk of infection can be minimized by good technique. Lead extraction in children can be performed safely. The availability of smaller diameter leads should encourage early change to an endocardial system where appropriate.

P-3

Early Intervention Reduces Histological Changes of the Pulmonary Root Under Pressure Overload

Maschietto N. (1), Vida V.L. (2), Stellin G. (2), Milanesi O. (1), Thiene G. (3), Basso C. (3)
Department of Pediatrics, Pediatric Cardiology Unit, University of Padua Medical School - Italy (1); Pediatric and Congenital Cardiac Surgery Unit, Centro Gallucci, University of Padua Medical School - Italy (2); Department of Medical-Diagnostic Sciences and Special Therapies, University of Padua Medical School - Italy (3)

Background: In the adult population, aortic regurgitation due to dilatation of the autograft is the main reason for reintervention after a Ross Procedure.
The aim of our research was to study the histological changes of the pulmonary root (PR) in an animal model of adult and neonatal rats, under physiological conditions and under pressure overload, to infer whether an early intervention can prevent the subsequent dilatation of the pulmonary root subjected to systemic pressure.
Material and Methods: Ten neonatal and ten adult Sprague Dawley rats underwent pulmonary artery banding tightened as to establish systemic pressure in the right ventricle and in the PR. Thirty days later they were sacrificed, the PR explanted and histologically analyzed. The following histological parameters were evaluated: thickness of the wall, number of elastic lamellae (EL), apoptotic rate of the smoot muscles cells (SMC), fibrosis, EL disarray and fragmentation and mucoid degeneration. The operated rats (OR) were compared to a group of same age shame operated (SO) rats.
Results: In the neonatal group, the OR had increased fibrosis (p 0.007), mucoid degeneration (p 0.012) and reduced rates of apoptosis ($p = 0.0099$) than the SO.
The adults OR, besides fibrosis (p 0,0002) and mucoid degeneration (p 0.043), showed an increased EL fragmentation (p 0.0054) and disarray (p 0.0005) but no differences in the apoptotic rate.
Comparing the OR, the adults showed higher rate of disarray (p 0.0058).
The SO neonates had higher rate of apoptosis ($p < 0,0001$) and disarray (p 0,0057) than the adults.
Conclusions: As shown in the SO, an involution of the PR, consisting in apoptosis of SMC and disarray of the EL, seems to be a physiological process in the neonates.
When subjected to systemic pressure, the PR shows an increase in fibrosis and mucoid degeneration both in neonates and adults, while pronounced EL disarray and fragmentation are present only in the adults.
Our results suggest that the dilatation of the autograft is due to major alterations of PR subjected to systemic pressure. However these seem to be more pronounced in the adults inferring better long term results after a neonatal Ross operation.

P-4

Fetal 3D echocardiography in congenital cardiac and extra-cardiac defects

Reffo E., Santagati C., Zabadneh N., Milanesi O., Biffanti R., Cerutti A., Maschietto N., Grison A.
Department of Pediatric Cardiology, Pediatric Clinique of University of Padua, Italy.

Background: Dismorphic appearance and extra cardiac malformations are quite commonly associated with CHD (25%) and suggest the presence of a genetic or chromosomal syndrome. Prenatal diagnosis enables us to formulate an accurate prognosis and to indicate an appropriate management plan.
The aim of this study was to explore which information 3-D echocardiography can add to the prenatal diagnosis of cardiac and extra cardiac anomalies and in which percentage of foetuses.
Methods: Foetuses with CHD diagnosed by 2D US at our Institution between October and February 2006 were prospectively

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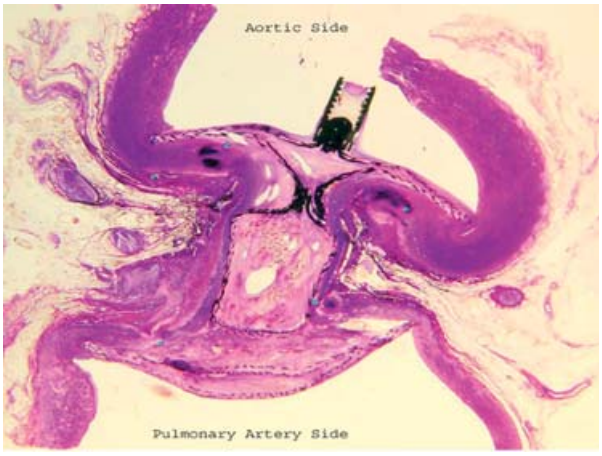
included in our study. Three-D images of skull, face, lips, hands, superior and inferior limbs, genitalia and heart, were acquired using a 2-4 MHz phased matrix array 3-D transducer. All 3-D volumes were off-line evaluated by one paediatric cardiologist, unaware of the 2-D diagnosis. The accuracy of the 3-D diagnosis was confirmed by correlation with post-natal or post-abortion findings.
Results: A total of 30 consecutive foetuses with cardiac defects were included in the study. After birth or interruption of the pregnancy, chromosomal or genetic malformations were present in 12 (trisomy 18 and 21, Cornelia De Lange syndrome, unbalanced translocation t4: 15, DiGeorge syndrome and heterotaxy syndrome), and 22 extra cardiac malformations were diagnosed. Three-D US enabled us to detect 17/22 extra cardiac defects with good imaging quality of skull, face and lip. Concerning the 12 syndromes, it was diagnostic in 7 cases; well showing micrognathia, brachiocephaly, lip cleft and left superior arm agenesis. Inverse correlation was found between gestational age and possibility to obtain diagnostic images of the face, but not of the limbs and genitalia. We didn't have any false positive. Three-D foetal heart examinations consented a very accurate definition of AV valves, ventricular, septal and outflow tracts anatomy. Pulmonary veins and aortic arch were poorly depicted in all cases no matter the gestational age. Three-D foetal heart evaluations provided additional diagnostic information in 1/3 of cases.
Conclusion. Our study suggests that the foetal 3-D echocardiography, mainly when performed before 24GW, can be a useful adjunction to 2D, allowing to identify syndromic foetuses.

P-5
Preclinical Evaluation of the Amplatzer Ductus Occluder-II
Gu X. (1), Qian Z. (1), Zhao C. (1), Oslund J. (1), Bass J. (2), Gruenstein D. (2)
AGA Medical Corporation, Plymouth, MN, UDA (1); University of Minnesota Children's Hospital, Minneapolis, MN, USA (2)

Purpose: Small Patent Ductus Arteriosus (PDA) is predominately treated with coils which are associated with risk of migration, especially in tubular type PDA. This study was to determine safety and effectiveness of Amplatzer Ductus Occluder-II (ADO-II) in animal models.
Materials and Methods: The newly designed ADO-II was constructed of multi-layer nitinol wire braid with symmetric retention disks and an articulating connecting center waist (3-6 mm in diameter). The devices were implanted via 4-5F catheters either transarterially (n = 7) or transvenously (n = 1) in 8 dogs with PDAs surgically created with autologous carotid grafts. Angiograms and pressure gradients were obtained in both the pulmonary arteries and aortas. The animals were followed-up for 90 days and afterwards euthanized for pathology.
Results: Technical success was achieved in all animals. All PDAs with the smallest diameter 1.4–3.44mm and length 3.05–10.2 mm were occluded within 8 minutes. All subjects were followed and examined at day 7, 30, 60 and 90 post implant; PDAs remained occluded in all subjects at 90 days. Post-implant pressure gradients were statistically insignificant ($p > 0.05$). Recapturability and repositionability were successfully demonstrated in all attempted 4 cases. Pathology found complete endothelial coverage on all aortic and pulmonary disks. Cross-sectional microscopic examinations show the retention disks are well apposed to the native vessel wall. The disks are incorporated into vessel wall and covered by a thin neointima. The PDA along with the device body is occupied with organized thrombus with 90–100% tissue ingrowth, showing effective occlusion of the PDA by the device (Figure).



Conclusion: The ADO-II appears to be safe and effective for transcatheter occlusion of PDA in the animal models. The device can easily be recaptured and repositioned before released offering a high degree of forgiveness. In addition to quick occlusion, mechanical stability, and ease of use, its unique double-disk and multi-layer design without fabric offers the favorable features of low profile and the ability to deliver from either arterial or venous approach. The ADO-II is expected to be a preferred alternative for clinicians to treat small PDAs defined as < 4mm in diameter.

P-6
Pulmonary Regurgitation: The Impact on Right Ventricle and the Effect of Percutaneous Pulmonary Valve Replacement
Søndergaard L. (1), Vejlstrup N.G. (1), Boudjemline Y. (2), Bonhoeffer P. (3)
Department of Cardiology, Rigshospitalet, Copenhagen, Denmark (1); Service de Cardiologie, Pédiatrique, Hôpital Necker Enfants Malades, Paris, France (2); Department of Paediatric Cardiology, Great Ormond Street Hospital, London, England (3)

Background: Despite successful repair of tetralogy of Fallot, the life expectancy for these patients remains lower than in the general population. This is mainly due to free pulmonary regurgitation (PR) in patients where the surgical correction involved a trans-annular patch. The importance of maintaining a competent valve has been emphasized, but the optimal timing for intervention remains to be established.
The purposes of this study were in a pig model to investigate the consequences of short-, medium- and long-term free PR on RV, and the reversibility of these changes after percutaneous pulmonary valve replacement.
Methods: 36 farm pigs were divided into four groups of nine pigs. At baseline group A, B and C had a percutaneous CP stent inserted into the pulmonary annulus to create free PR. After four, eight and twelve weeks, respectively, group A, B and C underwent percutaneous valve replacement. In order to allow remodelling of RV, the pigs were observed for four weeks after valve insertion before euthanasia. To examine RV function, MRI was performed before stent insertion, before valve replacement, and before euthanasia. Group D served as controls and underwent five MRI examinations: at baseline as well as after four, eight, twelve and sixteen weeks.
Results: Comparing the control group (D) with the three test groups (A, B and C) with different time of free PR, end-diastolic volume (RVEDV) and end-systolic volume (RVESV) were found to gradually increase over time. Furthermore, these volumes normalised after valve replacement in group A and B, but remains increased in the group with long-term free PR. Similar, ejection fraction (RVEF) was normal after valve replacement in group A and B, but impaired in the group the long-term free PR.

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Conclusion: This study examined the impact of free PR on RV in a pig model. The study showed that RV tolerates the volume overload from free PR for a certain time, but too late intervention will cause irreversible deterioration of the ventricular function. Thus, the model may serve to identify predictors, e.g. tissue-Doppler measurements and natriuretic peptides, for RV function after valve replacement and thereby optimise the timing of intervention.

P-7
Preliminary experience with hybrid palliation for patients with hypoplastic left heart syndrome

Knirsch W. (1), Waldvogel K. (2), Balmer C. (1), Dodge-Khatami A. (3), Pretre R. (3), Kretschmar O. (1)
Divisions of Pediatric Cardiology (1), Pediatric Intensive Care (2), Pediatric Cardiac Surgery (3), University Children's Hospital Zurich, Switzerland

Objectives: Hypoplastic left heart syndrome (HLHS) may be treated as the first step alternatively to Norwood procedure by a hybrid transcatheter-surgical palliation (bilateral pulmonary artery banding and ductal stenting).
Methods: Retrospective analysis of patients with HLHS (n = 7) and other single ventricle hemodynamics (n = 2) treated by hybrid palliation between 04/2006 and 12/2007 in our institution.
Results:

	Pat. I	Pat. II	Pat. III	Pat. IV	Pat. V
Diagnosis	HLHS	HLHS	HLHS	HLHS	HLHS
Restrictive ASD/PFO	+	-	+	-	+
Coronary perfusion, antegrade	no	no	no	yes	yes
PDA Stent type, size [mm]	2xSR (8x20)	1xSR (8x20)	SR(9x20)+ PG(8x24)	2xSR (10x20+ 9x20)	3xSR (8x20, 9x20,10x20)
ASD dilation at birth	yes	no	yes	yes	no
Stent CoA at birth	no	no	yes	yes	no
Total number of caths	3	2	3	3	2
Follow up [months]	20	2	17	12	8
Further interventions during follow up	Rebanding + ASD stent	-	Stent for PDA restenosis and for Re-CoA, ASD redilatation	Dilatation of the banding	Rescue septostomy after birth
Outcome	good	died interstage	good	good	good

	Pat. VI	Pat. VII	Pat. VIII	Pat. IX
Diagnosis	HLHS	LV-Tumor	DILV, TGA	HLHS
Restrictive ASD/PFO	-	-	-	-
Coronary perfusion, antegrade	yes	yes	yes	yes
PDA Stent type, size [mm]	2xSR (8x20)+ SR(9x20)	2xSR (8x20)	1xPG (8x24)	2xSR (9x20)
ASD dilation at birth	no	yes	no	no
Stent CoA at birth	no	no	no	yes
Total number of caths	3	2	3	1
Follow up [months]	8	4	6	1
Further interventions during follow up	ASD stent, LPA stent after Norwood	Stent for PDA restenosis	LPA stent after Norwood	Rebanding necessary
Outcome	good	died at Norwood	good	good

ASD = atrial septal defect; CoA = coarctation of the aortic arch; DILV = double inlet left ventricle; HLHS = Hypoplastic left heart syndrome; LPA = left pulmonary artery; PDA = patent ductus arteriosus; PFO = patent foramen ovale; PG = Palmaz Genesis; SR = Sinus Repo stent; TGA = Transposition of the great arteries

Conclusion: Hybrid transcatheter-surgical palliation for neonates with HLHS is feasible with good immediate results. During mid term follow re-interventions, i.e. ASD stenting resp. redilatation, rebanding, stenting of PDA restenosis and aortic arch restenosis before resp. LPA stenting after Norwood, might be necessary. This can only partly be explained as a learning curve. The patients have to be followed closely to reduce interstage morbidity and mortality. The superiority of the approach with regard to the neurological outcome still has to be proven.

P-8
Preoperative HDL-to-total cholesterol ratio predicts hemodynamic instability in patients after cardiopulmonary bypass surgery

Liuba P. (1), Gudbjartsson T. (2), Johansson S. (3)
Paediatric (1), Cardiology and (2), Cardiac Surgery, Lund University Hospital, Lund, Sweden; Cardiothoracic Surgery, Landspitali University Hospital, Reykjavik, Iceland (3)

Background: The increased morbidity after cardiopulmonary bypass (CPB) surgery is though to arise in part from the hemodynamic instability (HI) occurring during the early phase of the postoperative period. We assessed the ability of pre-operative inflammatory, lipid, and arterial endothelial phenotypes to predict HI following CPB surgery.
Methods: Plasma levels of C-reactive protein (CRP) and fibrinogen, high-density lipoprotein (HDL) and total cholesterol (TC), troponin T, and cGMP were measured preoperatively, 4-6 and 48 hours after CPB surgery of 22 patients (age: 9 to 67 years; mean: 28) with congenital heart disease. Systemic arterial endothelial-dependent and independent vasomotor function was assessed preoperatively and 4-6 hours after surgery by measuring flow-mediated dilatation (FMD) and nitroglycerine-induced dilatation of the brachial artery. After the first 12 postoperative hours, hemodynamic instability (HI) was defined by the need of single or repeated administration of vasoactive, inotropic or antiarrhythmic drugs until discharge.
Results: Preoperative HDL-to-TC ratio was significantly lower in patients who developed postoperative HI (n = 9) than in the remaining patients (p = 0.003). After adjustment for age, gender, CRP and fibrinogen, the odds ratio for this lipid index was 4.3 (95% confidence interval: 4.1 to 4.6; p = 0.04). Postoperative (48 hours) but not preoperative (p = 0.14) CRP was elevated in patients with HI (p = 0.03 vs. remaining patients). Although FMD decreased after surgery (from 7.9% preoperatively to 4.1%, p = 0.01) and did not significantly vary in relation to HI, the postoperative value correlated weakly with the preoperative (r = 0.5, p = 0.07) but not postoperative HDL-to-TC ratio. Troponin levels were similar in both groups (p = 0.4).
Conclusion: In this small-scale study, HDL-to-TC ratio preoperatively predicted the risk of HI after CPB surgery independent of patients` inflammatory and demographic characteristics, and, to a lesser extent, the postsurgical drop in systemic arterial vasomotor function. The findings lend support to the concept that HDL could be an additional therapeutic target in order to lower cardiovascular morbidity after CPB surgery.

P-9
Retro- and prospective analysis of cardiovascular risk factors in rehabilitative intervention programs for obese children

Böhm B. (1), Kallweit C. (2), Hartmann K. (2), Bauer C. P. (2), Oberhoffer R. (1)
Institute of Public Health Research, Faculty of Sport Science, University of Technology Munich, Germany (1); Department of Pediatrics and Children's Hospital Medical Center, University of Technology Munich, Germany (2)

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Introduction: Obese children tend to become obese adults with an increased risk to cardiovascular and metabolic diseases. Little is known, however, about the overt or subclinical incidences in paediatric population and the impact of interventions on them. In paediatric cardiovascular health prevention studies, non-invasive diagnostic parameters become of importance. An excellent marker of subclinical atherosclerosis is the carotid intima-media thickness (IMT). The objectives were a retrospective analysis of the intervention focussing on BMI, serum lipids and glucose, plus prospectively to investigate, if a hospitalized rehabilitation program has an effect on cardiovascular risk factors, including vascular changes.

Methods: Retrospective analysis of 2162 obese boys and girls aged 4 to 17 years (13±2), before and after a 4-weeks hospitalized intervention, including the evaluation of weight, BMI, blood pressure as well as serum lipids and glucose. Prospective analysis of 60 obese children pre and post intervention with respect on the same measures plus the sonographic measurement of IMT.

Results: Retrospective study: Average BMI of 31.72 kg/m2 (±5.73) at the beginning and of 28.56 kg/m2 (±5.20) at discharge, equivalent to an average reduction of 10% body weight. Systolic blood pressure in 25.1% of the patients above the 95th percentile, total-cholesterol in 69.9% above age related norm values, LDL-cholesterol in 73.4% increased, glucose in 4.1%. Strong positive association of the pathologies with weight classification. Significant reduction of all parameters during the intervention, especially in extremely obese patients. Prospective study: IMT with a mean of 0.517±0.049mm was above age related healthy peers without significant reduction at discharge.

Conclusion: High incidence of subclinical and evident cardiovascular and metabolic disease. The alterations of the analyzed cardiovascular risk factors were found to be significantly related to the loss of weight throughout the intervention program. Increased IMT is present in the obese paediatric population. In advantage to invasive diagnostics, the ultrasound measurement can demonstrate the effect of the intervention program clearly on the level of the vascular system since it enables an insight to the health of the blood vessel, but obviously pathological changes need longer time to be reduced.

P-10
Changes in left ventricular mass in the acute state and with refeeding in anorexia nervosa compared with ghrelin, leptin and adiponectin.

Docx M. K. F. (1), Eyskens F. (2), Vandenberghe Ph. (1), Gewillig M. (3), Mertens L. (3)
Department of Paediatrics and Paediatric Cardiology Koningin Paola Kinderziekenhuis, Antwerp, Belgium (1); Center of Metabolic Diseases Koningin Paola Kinderziekenhuis, Antwerp, Belgium (2); Paediatric Cardiology University Hospital Gasthuisberg, Leuven, Belgium (3)

Objective: To find differences in left ventriculair mass (LVM) and LVM/height 2,7 heart rate and blood pressure before and after refeeding and to identify which appetite stimulators or depressors are associated with these ventricular mass differences.

Patients and methods: 25 anorexic girls (11,46-17,9 year) who fulfilled the DMS-IV criteria for AN were evaluated at admission and after 9 months refeeding (mean weight gain 0,5 kg/weekly till there minimal healthy weight). Before and at 9 months refeeding BMI,heart rate, blood pressure LVM, LVM height 2,7, ghrelin, leptin and adiponectin were measured. Analysis:Ghrelin (Ghrelin human EIA);Leptin (Leptin(Sandwich) ELISA);(Adiponectin (mediagnost ELISA for adiponectin E09).

Results:

Parameters	Admission	9 months	p-value
Number	25	25	
Age (y)	14.26±1.36	15.00±1.36	0.062
BMI (kg/m²)	15.11±1.35	17.52±1.61	<0.001
Weight loss (%)	21.18±6.67		
HR (bpm)	59.24±18.26	71.44±12.08	0.008
SBP (mm Hg)	94.28±13.46	106.56±11.93	0.001
DBP (mm Hg)	58.28±11.66	64.04±5.66	0.031
LVM (g)	81.64±22.93	100.54±19.46	0.003
LVM/height 2,7 (g/m2,7)	27.60±7.22	33.46±5.95	0.003
Ghrelin (ng/ml)	20.07±8.07	26.26±9.89	0.023
Leptin (ng/ml)	1.53±1.66	7.32±6.01	<0.001
Adiponectin (µg/ml)	23.96±12.66	27.10±14.06	0.42

Conclusions: We demonstrated that, LVM, LVM/height 2,7, heart rate and systolic and diastolic blood pressure ($p < 0.05$) significantly increase after refeeding. This is associated with a rise in leptin and decrease in ghrelin. Ghrelin reduces the cardiac afterload and increases the cardiac output. It has also an anti-apoptotic effect and inhibits sympathetic nerve activity. Leptin has been shown to influence natriuresis with chronic sympathetic activation especially to the kidney and may lead to systemic vasoconstriction and blood pressure elevation. There a no significant changes in adiponectin levels. The change in blood pressure and cardiac parameters like mass index might be related to changes in the endocrine 'appetizer' system.

P-11
Hypertension following pediatric heart transplantation is primarily associated with immunosuppresive regimen

Roche S.L., Kaufman J., Dipchand A.I., Kantor P.F.
The Hospital For Sick Children, Toronto, Canada

Introduction: Although hypertension is recognized as prevalent in paediatric heart transplant recipients, risk factors for this important complication, and in particular the role played by immunosuppressive therapy, have not been studied in children.

Methods: 24-hour ambulatory blood pressure (24ABP) results of children, (<18 years), followed in our clinic and surviving > 1 year after cardiac transplantation were analysed. Subjects were divided into 2 groups:

Normotensive - if mean systolic and diastolic, daytime and night time BPs all < 95th centile

Hypertensive - if mean BP > 95th centile in one or more domains.

Clinical data contemporaneous with 24ABP were compared between the groups and 24ABP compared with clinic measurements made within one month. The effectiveness of BP control was considered by examining the records of hypertensive patients with repeated 24ABP monitoring.

Results: Fifty-one children (25 male) met inclusion criteria. Hypertension was detected in 25/51 children, 64% of these were only hypertensive at night. Referenced to 24ABP, the sensitivity and specificity of casual clinic BP measurements to detect hypertension were 10% and 92%.

Hypertensive and normotensive patients were similar as regards sex, age at transplantation, time between transplantation and 24hr-ABP and choice of calcineurin inhibitor. In contrast, hypertensive patients were receiving a significantly greater number of immuno-

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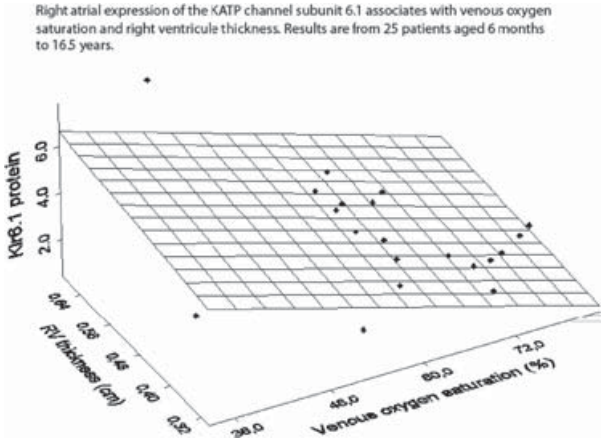
suppressive agents (2.92 vs. 2.12 $p < 0.01$), had higher tacrolimus levels (10 vs. 8.1mcg/L $p = 0.03$) and were more likely to be on maintenance prednisone therapy (64% vs. 23% $p < 0.01$) or regimens including sirolimus (40% vs. 12% $p = 0.03$). Multiple regression analysis showed that after controlling for tacrolimus level a combination of prednisone and sirolimus together were more strongly associated with hypertension than either agent alone (OR 7.3 CI 1.5–36.1 vs. OR 4.1 CI 0.85–26.3). Glomerular filtration rate and echocardiographic data did not differ between the groups.
21/25 hypertensive patients underwent repeated 24hr-ABP monitoring. Despite increasing antihypertensive medication adequate blood pressure control was achieved in only 38% by the time of their second 24hr-ABP a median of 6 months later.
Conclusion: Hypertension after pediatric cardiac transplantation is frequently nocturnal and best detected by 24ABP. It is primarily associated with immunosuppressive regimen and proves resistant to control with current therapeutic strategies.

P-12
Safety of maximal exercise testing in children with pulmonary hypertension
Smith G., Reyes J., Russell J., Humpl T.
Heart Center, Hospital for Sick Children, Toronto, Canada

Introduction: Maximal cardiopulmonary exercise testing is widely regarded as a valuable means to quantify functional capacity in patients with cardiovascular disease. Although several studies have examined exercise testing in adult pulmonary hypertension patients, information regarding pediatric patients is limited, possibly related to safety concerns during exercise. The purpose of this study was to examine the safety of maximal cardiopulmonary exercise testing in pediatric pulmonary hypertension patients.
Methods: Exercise and echocardiography data was obtained retrospectively from pulmonary hypertension patients referred for exercise testing at the Hospital for Sick Children between 01/2001 and 09/2007. Patients with a six minute walk distance < 300m were excluded from maximal cardiopulmonary testing, which was conducted using a cycle ergometer or treadmill until volitional fatigue. Test results were grouped according to: ischemic ECG changes, presence of arrhythmia, and oxygen saturation at peak exercise, and graded as "mild", "moderate" or "severe".
Results: Data was gathered from 32 patients (5 idiopathic, 27 secondary) aged 12.5 years (range 6.9 to 18.8) who participated in 66 maximal cardiopulmonary tests with a resting echocardiography RVsp of 79.5 mmHg (33 to 126). Peak VO2 was 23.2 ml/kg/min (11.4 to 38), with a decrease in SaO2 to 85% (47 to 100) at peak exercise. Thirty six (55%) patients were on standard pulmonary hypertension therapy at time of testing. There was a frequency of 27% for mild and 2% for moderate arrhythmia. Notable ST depression was graded as mild (13%), moderate (1%), and severe (2%) in all tests. Twenty five (38%) patients had a decrease in O2 saturations to < 85% or a 10% absolute decrease from baseline. There were no significant events as syncope, chest pain, or dizziness. Tests were stopped for fatigue (53%), leg fatigue (23%) dyspnea (21%) or miscellaneous reasons (3%).
Conclusion: The results of this study suggest that maximal cardiopulmonary testing can be performed safely in pediatric pulmonary hypertension patients. Although the number of patients in this study was limited, the data suggests that the absence of significant patient symptoms, low incidence of arrhythmia, and no significant ST depression make this a safe choice for measuring functional capacity in this patient population.

P-13
Expression of right atrial KATP channel pore in children with congenital heart disease: an adaptive mechanism during hypoxia?
Dauvé V. (1), Philip-Couderc P. (1), Roatti A. (1), Habre W. (2), Sierra J. (3), Kalangos A. (3), Beghetti M. (4), Baertschi A. J. (1)
Departments of Neuroscience (1), Anaesthesia (2), Surgery (3), and Pediatrics (4), Centre Médical Universitaire, Geneva, Switzerland

Introduction: Children with congenital heart disease (CHD) may be subjected to severe hypoxia, but the mechanisms for their survival are still unknown. We tested whether ATP-sensitive potassium channels (KATP) could be involved. KATP channels are composed of 4 pore-forming potassium channel (Kir) subunits 6.x, and 4 regulatory sulfonylurea receptors (SURy.z). In rodents, KATP channels provide vital adaptation during heavy exercise, but little is known in humans, and nothing at all in children with CHD.
Methods: After obtaining the informed consent form, 25 children undergoing surgical repair of their CHD (Tetralogy of Fallot, ventricular or atrial septal defects, transposition of great vessels, truncus arteriosus, Anomaly of Ebstein B) were included in the study. We measured Kir6.1 and Kir6.2 subunit proteins in extracts of right atrial tissue obtained during the surgery. Histology showed that at least 34% of the tissue sample consisted of cardiomyocytes and < 1% of vascular smooth muscle. Subunit expression levels were related to presurgical blood gases, blood chemistry, cardiac echographic and ECG parameters, age, sex, height and weight (altogether 39 parameters).



Right atrial expression of the KATP channel subunit 6.1 associates with venous oxygen saturation and right ventricule thickness. Results are from 25 patients aged 6 months to 16.5 years.

Results: In optimized step-wise multivariate regressions, KIR6.1 correlated inversely with venous O2 saturation and positively with thickness of right ventricle (model $p < 0.001$), and KIR6.2 inversely with venous O2 saturation and positively with perimeter of left ventricle ($p < 0.008$).
Conclusion: Results strongly suggest that low venous O2 saturation and hypoxia-induced mechanical stress are the dominant variables that determine increased atrial tissue expression of Kir6.1 and Kir6.2. The plasticity of the KATP channel pore may provide new insight about survival and therapy of the stressed human heart
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P-14
Persistent pleural effusions after the Fontan procedure
Stumper O., Ofoe V., Reinhardt Z., Bhole V., Stickley J., Jones T., Barron D.J., Brawn W.J.
The Heart Unit, Birmingham Children's Hospital – NHS Foundation Trust, Birmingham, UK

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Objective: To define the incidence of pleural effusions after completion of a Fontan procedure dependant on season and type of operation and pre- and post operative variables.
Background: Previous studies suggested that avoidance of surgery during winter months and creation of Fontan fenestrations reduce the incidence and duration of pleural effusions. No studies have examined whether the type of Fontan or ventricular morphology has an impact on the incidence and duration of pleural effusions.
Patients and Methods: Between 1/1988 and 7/2004 406 patients underwent a modified Fontan procedure at a median age of 4.7 yrs (IQR 3.8–7.1). A morphologically right ventricle was present in 163 pts (40%). Fenestrations were performed in 216pts (53%). An extracardiac conduit was used in 194 (48%). Early mortality was 4.4% (n = 18), 4 patients required takedown (1%). Late mortality was 5.4% (21/388) , late takedown 1 , Htx 3 patients. Actuarial survival was 90 ± 2%, 86 ± 2% and 82 ± 3% (5, 10, and 15 years). Freedom from re-intervention was 83 ± 4%, 76 ± 4% and 74 ± 8% (5, 10, and 15 y) .
Results: Preoperative and bypass remained stable throughout the observation period.
Intercostal drains were required for a mean of 8(SD12) days and hospital discharge was at 13(14) days.
There was no difference in the duration of pleural effusions between summer and winter periods [8 (10.5) vs. 9(14) days; $p = 0.27$].
Duration of pleural effusions was not significantly correlated to either early post operative pulmonary artery pressure, transpulmonary gradient or creation of a surgical fenestration.
Pleural effusions persisted significantly longer in patients with right vs. left ventricular morphology [14.8 (13.4 vs. 9.7 (10.0) days; $p < 0.01$] and after extracardiac conduit vs. atriopulmonary or lateral tunnel Fontan [15.3 (12.4) vs. 8.2 (9.8) days; $p < 0.01$].
Conclusion: The duration of pleural effusions after the Fontan procedure is prolonged after an extracardiac Fontan procedure and in the context of right ventricular morphology. The creation of a surgical fenestration or the timing of surgery during the year does not impact on the duration of pleural effusions.

P-15
Long term behavioural outcome after modified Fontan repair (Total cavopulmonary connection, TCPC) of functional single ventricle

Vahsen N., Toussaint-Götz N., Schneider M.
Deutsches Kinderherzzentrum Sankt Augustin, Germany

Introduction: Total cavopulmonary connection (TCPC) is the current surgical standard approach for definite palliation of functionally single ventricle hearts. Most studies about the neurodevelopmental outcome after a modified Fontan operation focus mainly on different cognitive and motor skills. Only a few consider potential behavioural problems in the long term due to numerous hospital stays, parental overprotection and/or neurological complications. The purpose of this study was to assess the parent reported behavioural outcome of patients with a congenital single ventricle physiology after a total cavopulmonary connection.
Methods: Seventy-seven parents evaluated their children's behaviour (4 to 17 years) by completing the Child Behaviour Checklist (CBCL/4-18), a psychological inventory designed to identify behavioural problems on eight syndrome subscales (withdrawn behaviour, somatic complaints, anxious/depressed behaviour, social problems, schizoid/compulsive behaviour, attention problems, delinquent behaviour, aggressive behaviour) and three global scales (internalising behaviour, externalising behaviour, total behavioural problems).
Results: Compared to healthy children, patients – as reported by their parents – showed significantly ($p < 0.05$) more behavioural problems on all syndrome subscales and global scales except the subscales "withdrawn behaviour", "schizoid/compulsive behaviour" and "delinquent behaviour". Eighteen percent of all children showed "somatic complaints" in the clinical range, 12% showed "attention problems", 9% showed "social problems", 6% showed "aggressive behaviour" and 5% showed "anxious/depressed behaviour" in the clinical range. Evaluating the global scales, 29% of the children fell within the clinical range for "internalising behaviour", 18% fell within the clinical range for "externalising behaviour" and 23% of the children scored in the clinical range on the "total behavioural problems" scale.
Conclusion: According to their parents, almost one fourth of all patients show substantial internalizing, externalising and/or total behavioural problems that affect their daily living (e.g. school performance, social life). The reasons still remain unclear and have to be focused on in the future. If the child's behavioural problems are severe, careful psychological and psychiatric assessment and therapy is indicated.

P-16
Pulmonary Arterial Hypertension (PAH) predicts outcome of patients with Congenital Diaphragmatic Hernia (CDH)

Charisopoulou D., Hislop A., Moledina S., Foster H., Flynn H., Schulze-Neick I., Haworth S.G.
Great Ormond Street Hospital for Sick Children NHS Trust, London, UK

Introduction: PAH as a consequence of hypoplastic lung vasculature due to CDH may in turn serve as a factor to predict outcome. In our patient group, we correlated the short- and long-term outcome after corrective surgery with a series of pre- and postoperative echocardiographic studies with a focus on PAH.
Methods and Patients: All patients who underwent repair for isolated CDH in the period of 2003 to 2006 (n = 55, 26 female, 7 right, 48 left, no bilateral CDH) were examined in a retrospective study. 3 died prior to surgery and 2 in the immediate postoperative period. 13 (24%) received preoperative ECMO. We analysed retrospectively the echocardiographic examinations that were performed at admission and prior to CDH repair, at the time of discharge from postoperative intensive care, and at standardized follow up at 4 weeks after surgery.
Results: The overall survival of our patient group was 76%. All are well with no evidence of PAH with the youngest being 1 year old and the eldest 3.5 years. Patients were divided in groups each based on echocardiography preoperatively or postoperatively at 4 weeks, and PAH was classified as either being suprasystemic (right ventricular to systemic arterial peak pressure (RVSP to SAP) ratio > 1) or being more than half-systemic (RVSP to SAP ratio > 0.5). PAH occurred in 49% patients preoperatively and 18% postoperatively, and was associated with worse early and late outcome (see table).

Group	n	early	late	Overall mortality
Preop, RVSP/SAP ratio < 1	34/55	0/34	1/34	3%
Preop, RVSP/SAP ratio > 1	21/55	5/21	2/21	38% $p < 0.001$
Postop, RVSP/SAP ratio < .5	41/50	n/a	1/41	2%
Postop, RVSP/SAP ratio > .5	9/50	n/a	4/9	45% $p < 0.05$

Conclusions: Our findings suggest that the presence and severity of PAH during pre- or postoperative echocardiography in pts with CDH is associated with a worse outcome. Thus, the evaluation for PAH in these patients both pre and post operatively may be used as a predictive factor for early and long-term survival.

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P-17

High incidence of acquired von Willebrand syndrome in patients with congenital heart defects

Kidszun A. (1), Rentzsch A. (1), Borchers J. (2), Eichler H. (2), Abdul-Khaliq H. (1)
Department of Paediatric Cardiology (1) and Institute of Clinical Haematology and Transfusion Medicine (2), Saarland University Hospital, Homburg/Saar, Germany

Introduction: Since its first description in 1983 acquired von Willebrand syndrome (AvWD) has been found to be associated with various underlying diseases, i.e. myeloproliferative disorders or aortic stenosis in adults. Information on the incidence of AvWD in patients with congenital heart disease is rare. Because of increased risk of bleeding and occurrence of different coagulation disorders in association with cardiopulmonary bypass further assessment of platelet function is warranted. The aim of this study is the prospective screening and characterization of AvWD in children and adolescents with congenital heart defects (CHD) undergoing surgical or interventional procedures.
Methods: 80 consecutive patients with CHD admitted to our paediatric cardiology department for surgery or cardiac catheterisation were included in this study. In addition to routine coagulation tests, levels of ristocetin cofactor, von Willebrand factor antigen, and factor VIII activity were measured. In addition, cardiac morphological and haemodynamic evaluation was simultaneously performed. Furthermore all patients and their parents were requested to complete a questionnaire to assess a possible increased risk of bleeding.
Results: AvWD was detected in 11 of 80 patients (13.8%) with different forms of CHD. Three children (3.8%) with diagnosed AvWD showed prolonged bleeding after catheterisation or clinically apparent tendency of bleeding. In one patient with surgically corrected tetralogy of Fallot and significant pulmonary artery homograft stenosis (pressure gradient 70 mmHg), who underwent further surgical correction, decreased ristocetin-cofactor activity completely normalized one week following homograft replacement and elimination of the stenosis. Generally no significant relationship between type of heart failure or defined haemodynamic conditions (gradient in the LVOT/RVOT) and AvWD could be observed. Likewise AvWD was not associated with serious bleeding complications.
Conclusion: Patients with CHD have an increased risk to develop AvWD. This disorder should be considered in all patients with prolonged bleeding after cardiac surgery. Although specific causes of AvWD due to congenital heart defects remain unclear, it may be improved by surgical correction of a haemodynamic abnormality. To determine the relevance of AvWD for clinical practice and to provide a better insight into its pathogenesis further investigation is in progress.

P-18

Daclizumab as induction therapy in pediatric heart transplantation

Fuchs A., Diterich J., Beier G., Dalla-Pozza R., Urschel S., Kozlik-Feldmann R., Netz H.
Department of Pediatric Cardiology, University Medical School Grosshadern, Munich

Daclizumab, a humanized monoclonal antibody, is a new immunosuppressive drug which binds with high affinity to the Tac subunit of the Il-2 receptor complex. Effective immunosuppression with Daclizumab in adult patients encouraged the initiation of the administration of Daclizumab as induction therapy in pediatric heart transplantation.
Sixteen patients (9 boys, 7 girls, age 8 7 yrs, BMI 1.75 0.52 m2), received Daclizumab as induction therapy in a dosis of 1 mg/kg intravenously perioperatively and on day 7 and 21 after orthotopic heart transplantation. Additional immunosuppression was cyclosporine (CsA, n = 14) or tacrolimus (TAC, n = 2), mycophenolate mofetil (MMF) and prednisolone. Prednisolone was tapered rapidly in the first six months after heart transplantation.
The administration of Daclizumab was not associated with any side effect. Owing to the blockade of the IL-2-receptor the dosage of calcineurin inhibitors could be reduced leading to less renal and hepatic toxicity. Instead of aiming at CsA trough levels of 350-400 ng/ml/TAC trough levels of 12-15 ng/ml in the first weeks after transplantation we reduced to 250 in the CsA group and to 10 in the TAC group. CD25+ T-lymphocytes began to be re-expressed after 2-3 months after administration of Daclizumab.
In a mean follow-up time of 26 11.5 months no acute or chronic episode of rejection could be experienced. The incidence of opportunistic infections was not elevated (5 bacterial, 4 viral and 3 fungal infections which responded well to adequate treatment). No de novo malignancies, especially no lymphoproliferative disease (PTLD) was noticed. Actually patient and graft survival is 100%.
Our results show that immunoprophylaxis with Daclizumab induction therapy in pediatric heart transplantation is safe, effective and well tolerated and does not lead to increased opportunistic infections or malignancies. The reduction of calcineurin inhibitors led to less calcineurin related side effects and raised the quality of life of transplanted patients.

P-19

Potential coronary complications after arterial switch operation.

Pesonen E. (1), Munkhammar P. (1), Hochbergs P. (2), Jögi P. (1)
Children's Hospital, University Hospital, Lund, Sweden (1);
Department of Radiology, University Hospital, Lund, Sweden (2)

Introduction: Starting from 1980 we have operated 282 patients with arterial switch technique. At present we systematically control coronary angiograms in all our patients with transposition of the great arteries. Since 1996 the operative technique has not been modified essentially.
Methods: Selective coronary angiograms were done on 76 patients.
Results: Before 1996 we had five coronary complications. All these five patients had heart failure early postoperatively and myocardial infarction was diagnosed. The distribution of the coronary tree was normal in three of them but in one patient the circumflex coronary artery aroused from the right coronary artery. In a patient with a single right coronary artery the conus branch was totally obstructed and coronary filling took place trough collaterals. Ostioplasty of the left coronary artery has been done at the age of 16 years in one patient. He still needs ß-blocking medication for ventricular extrasystoles. One patient with ostial stenosis was asymptomatic but was treated at 10 years of age with a coronary drug-eluting stent.
From 1996 two out of 36 asymptomatic investigated patients showed a coronary problem. The left coronary artery was treated with balloon dilatation which led to a clinically insignificant dissection of the artery. He was later at the age of 8 years treated with a drug-eluting stent. An 18-year-old athletic boy with total obstruction of his left coronary artery is still waiting for his treatment decision.
Patients with coronary problems have usually a difficult postoperative recovery. However, our last two patients had a smooth postoperative course and were asymptomatic. Earlier reports indicate that patients operated with the arterial switch technique and having ostial stenosis are at risk of sudden death. We have considered stenting of our patient with almost pinpointed stenosis to be indicated.
Conclusion: The finding of coronary obstruction in totally asymptomatic patients with an uneventful postoperative course indicates the necessity

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of coronary angiography in all patients with arterial switch operation. The examination should probably be carried out before puberty when the physical activities usually increase significantly.

P-20

Outcome of children surviving Persistent Pulmonary Hypertension of the Newborn

Roofthooft M.T.R. (1), Elema A. (1) , Bergman K.A. (2), Berger R.M.F. (1)
Department of Pediatric Cardiology (1) and Department of Neonatology (2), Beatrix Childrens Hospital, University Medical Centre Groningen, University of Groningen, The Netherlands

Objective: To asses the impact of Persistent Pulmonary Hypertension of the Newborn (PPHN) on health-related quality of life and subjective health status and to determine their influencing factors.
Methods: This is an observational study of 145 consecutive newborns with PPHN, admitted to a tertiary cardiac and neonatal intensive care unit, over a 11-year period. Clinical PPHN confirmed by echocardiography was used as inclusion criteria. Parents of surviving patients were asked to complete the Child Behavior Checklist, as well as a Effects of PPHN-Questionnaire.
Results: In the study group 92 patients survived (63.4%, 92/145), with nowadays a mean age of 5,6 jaar (SD = 2.97), ranging from 0 to 11 years. Two families were untraceable. 55 of the 90 parents responded (61.1% response-rate) A high morbidity in the PPHN patients was seen. Problems of the respiratory tract (31.4%), delayed motor development (17.6%), visual impairment (13.7%), congenital heart disease (13.7%) and hearing impairment (11.8%). The rate of rehospitalization (54.9%), the use of medication (29.4%) and health care in general (27.5%) as well as the need for physiotherapy (29.2%) and speech therapy (30%) were increased. Our study suggests a positive correlation between the severity of PPHN and the visual impairment. Moreoever we observed a higher incidence of delayed mental and motor development in boys with PPHN.
Conclusion: PPHN is a serious and often fatal condition, associated with a high morbidity and mortality (31.7%).

P-21

Relation of arterial stiffness with intrauterine growth retardation (IUGR)

Levent E. (1), Atik T. (2), Darcan S. (2), Ulger Z. (1), Goksen D. (2), Ozyurek R. (1)
Ege University, Faculty of Medicine, Department of Pediatric Cardiology (1) and Endocrinology (2), Izmir, Turkey

Much epidemiological evidence has linked low birth weight with late cardiovascular risk. Intrauterine growth retardation (IUGR) is associated with the increased risk of cardiovascular disease in adult life; it is unclear whether the relationship is present at younger ages. We evaluated whether the abdominal aortic stiffness was altered in the patients with IUGR (born at term with birth weight small for gestational age) in younger ages.
Thirty-two (24 females and 8 males) IUGR patients aged 8.77 ± 2.05 years were enrolled into the study. The birth weight was traced from the medical records. Their gestational ages were 38.9 ± 0.85 weeks and birth weight 2130 ± 198 g respectively. Healthy 31 subjects who have normal gestational age and birth weight, matched for age and sex were recruited as a control. Aortic strain (S), pressure elastic modulus (Ep), and normalized Ep (Ep*) and aortic distensibility (DIS) were measured by a sphygmomanometer and transthoracic echocardiography in all subjects from abdominal aorta.
There was no statistically significant difference between the study and the control groups in sex, mean age, body mass index, lipid profile, leptin, IGF-1 and IGF-BP3. In IUGR patients S (0.201 ± 0.027 vs. 0.254 ± 0.031, P < 0.001) and DIS (1.08 ± 0.19 vs. 1.42 ± 0.24, P < 0.001) were significantly lower compared with the control group. However Ep (188 ± 36.2 vs. 146 ± 27.1, P < 0.001) and Ep* (2.97 ± 0.40 vs. 2.1 ± 0.39, P < 0.001) were significantly higher in IUGR patients.
In conclusion, this study demonstrates that abdominal aorta stiffness increased in IUGR patients. These data suggest that prenatal events could predispose to cardiovascular risk in later life.

P-22

Normal values, range and upper limits, of NT-pro B-type natriuretic peptide in infants and children. Analysis of combined data from 4 studies

Nir A. (1), Lindinger A. (2), Rauh M. (3), Bar-Oz B. (4), Laer S. (5), Schwachtgen L. (2), Koch A. (3), Falkenberg J. (6), Mir T.S. (6)
Pediatric Cardiology Unit, Shaare Zedek Medical Center, Jerusalem, Israel (1), Pediatric Cardiology, University Clinic Saarland, Homburg/Saar, Germany (2), Pediatric Cardiology, University Clinic Erlangen, Erlangen, Germany (3), Department of Neonatology, Hadassah Medical Center, Jerusalem, Israel (4), Institute for Clinical Pharmacology, University Duesseldorf, Duesseldorf, Germany (5), Pediatric Cardiology, University Heart center University Hamburg, Germany (6)

Introduction: B-type natriuretic peptide (BNP) and the amino terminal segment of its prohormone (NT-proBNP) are markers for heart disease and can be measured on commercial laboratory platforms. The peptide levels are age and assay dependent. Normal value range and its upper limits are essential in order to facilitate the use of these markers in the pediatric population. This study is a summation of four studies that measured NT-proBNP levels in normal infants and children using electrochemiluminescence immunoassay (Roche Diagnostics, Mannheim, Germany). It is the largest published to date.
Methods: Age intervals for upper limits of normal were chosen for intervals where there was no age dependent peptide level change. Statistics were performed on log-transformed data.
Results: There were 690 subjects, aged from birth to 18 years, 325 (47%) were males. NT-proBNP levels were very high in the first days of life with drastic decline in the first weeks. The peptide levels continued to decline gradually with age, with significant decrease between age 1 month and 18 years (r = 0.43, p < 0.001). Male and female levels were only different in the age group of 10 years to 14 years (medians: male 38 pg/ml, female 56.5 pg/ml, p = 0.002). However, the upper limit of normal for males and females was not different even in this age group. The mean values, upper and lower limits of normal of NT-proBNP levels are shown in the table.

Age interval	n	mean ± SD	5%tile	95%tile
0-2d	43	2.820 ± 3.725	321	11.987
3-11d*	84	1.800 ± 2.795	263	5,918
>1m to ^1y	50	143 ± 206	37	646
>1y to ^2y**	38	123 ± 125	39	413
>2y to ^6y	81	73 ± 82	23	289
>6y to ^14y	278	46 ± 64	10	157
>14y to ^18y	116	36 ± 54	6	158

*no data between 12 days and 30 days; ** a significant decrease with age in this age interval; Values are in pg/ml.

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Conclusion: NT-proBNP levels are elevated in the first days of life and decrease drastically thereafter. There is a mild gradual decline with age throughout childhood. Girls have somewhat higher levels during puberty. Normal range for each age group is established.

P-23

Successful implantation of ventricular assist devices in children

Fuchs A. (1), Diterich J. (1), Beier G. (1), Kozlik-Feldmann R. (1), Daebritz S. (2), Schmitz C. (2), Netz H (1)
Department of Pediatric Cardiology, Medical School Grosshadern, Munich, Germany (1); Department of Cardiac Surgery, Medical School Gro sshadern, Munich, Germany (2)

Ventricular assist devices are successfully used in the treatment of end-stage cardiac failure in the pediatric population. To evaluate early and late results of the implantation of ventricular assist device systems in children as bridge to recovery or bridge to transplant.
27 patients received assist devices, with ages ranging from 2 days to 23 years. There was size differences with weights ranging between 2.7 and 75 kg. 18 patients were supported with ECMO with 6 patients weighing less than 3 kg, 4 patients with Medos, 2 patients with Berlin Heart and 3 patients with Novacor. Patients were supported with an average of 4.2 days (ECMO), 14.7 days (Medos), 14.1 days (Berlin Heart) and 8.3 days (Novacor). 16 patients were successfully weaned from the device, with 9 patients undergoing transplantation.
Diagnosis were in 18 pts congenital heart diseases and in 9 pts acquired heart disease. Indications for implantation of an ECMO were postcardiotomy heart failure in 16/18 pts and reanimation in 2/18 pts, the indication for the use of a Medos, Berlin Heart and Novacor assist device was bridging to transplant in DCM in all pts. Myocardial recovery was observed in 6/18 of the ECMO pts. Overall, 14 pts were successfully weaned from the device. 9 pts underwent transplantation: 21% of the ECMO patients (3/18), 50% of the Medos pts. (2/4), all Berlin Heart pts. (2/2) and 67% of the Novacor pts. (2/3). 13 pts (48%) were discharged and are long-term survivors.
These results demonstrate the efficacy of the implantation of ECMO or other VAD systems in children with cardiac disease as bridge to transplant.

P-24

Psychosocial adjustment and quality of life in children attending a hypertrophic cardiomyopathy clinic

Kaski J.P., O'Curry S., Winter-Beatty J., Mead-Regan S., Tome-Esteban M., Deanfield J.E., McKenna W.J., Elliott P.M.
Great Ormond Street Hospital, London, U.K.

Background: Hypertrophic acrdiomyopathy (HCM) is an inherited heart muscle disease that causes substantial morbidity and premature death in the young. Health-related quality of life (QoL) among children with HCM has not been evaluated previously. Furthermore, the psychosocial impact of regular clinical screening of unaffected, asymptomatic individuals is unknown.
Methods: 139 children (89 males [64%]) aged 5-18 years (median 13 years) evaluated in a HCM clinic between October 2006 and February 2007 participated in the study. 110 (79.1%) were unaffected but had a family history of HCM and attended the clinic for screening, 21 (15.1%) fulfilled diagnostic criteria for HCM, and 8 (5.8%) harboured HCM-causing genetic mutations but had normal echocardiograms. Patients completed the Generic Core Scales (n = 130; 93.5%) and Cardiac Module (n = 129; 92.8%) of the PedsQL questionnaire. Parents completed the parent/proxy versions of the Generic Core Scales (n = 128; 92.1%) and Cardiac Module (n = 127; 91.4%) of the PedsQL questionnaire, and the Strengths and Difficulties Questionnaire (SDQ). These are widely used and extensively validated measures of QoL and behavioural problems in children, assessing several physical and psychosocial dimensions.
Results: Children with HCM had significantly lower scores overall and in most dimensions of the General Core and Cardiac PedsQL modules compared with unaffected individuals and gene carriers. Anxiety and behavioural scores on the parent-rated SDQ were similar across all groups and not different to scores from the normal population. However, children with HCM were more likely to fulfil criteria for clinically important behavioural and emotional problems. Importantly, QoL scores in the screening group were not different to scores from a normative UK population.
Conclusion: Children with HCM have a significantly reduced quality of life and substantial impairment in social and emotional functioning. QoL and psychosocial adjustment scores among unaffected children attending for screening did not differ from a normative UK population. This study identifies a need for psychological input as part of the multidisciplinary management of children with HCM.

P-25

Coronary arteries in children with TGA late after arterial switch operation

Moll M. (1), Dryzek P. (2), Moszura T. (2), Mazurek-Kula A. (2), Moll J.J. (1), Sysa A.(2)
Department of Cardiosurgery, Polish Mother's Memorial Hospital Research Institute, Lodz, Poland (1); Department of Cardiology, Polish Mother's Memorial Hospital Research Institute, Lodz, Poland (2)

Introduction: Evaluation of coronary arteries is one of the most important factors determining late results of the arterial switch operation. Estimation of the anatomy, localization and size of the coronary arteries may be determining the patient's future. In TGA, LVmass may be influenced by the coexisting anomalies but also by the anatomy of the coronary arteries.
Methods: Since 1992 until 2007 496 children with transposition of the great arteries underwent arterial switch operation. In 136 of these patients (aged 6 to 10 years) selective coronary arteriography was performed. In each case coronary pattern was analyzed. Indexed diameter and cross-section area of LAD, Cx and RCA divided by BSA was measured. LVmass was evaluated using 2-D ECHO.
Patients were divided into 3 groups:
1. Simple TGA – 87pts
2. TGA and VSD (including Taussig-Bing) – 39pts
3. TGA with anomalies of the aortic arch (CoA, HAA) – 10pts
Further, every group was divided into subgroup with typical (T) and anomalous (A)coronary anatomy.
The indexed diameter of LAD, Cx and RCA was compared between the groups. These parameters, in correlation to LVmass, were compared also within the subgroups T and A.
Results: In group 1 there was a significant difference ($p < 0{,}001$) in the indexed diameter of Cx between patients T and A. Cx/BSA was higher in group A. In group 2 there was a significant difference ($p < 0{,}006$) in the diameter of indexed RCA between patients T and A. RCA/BSA was higher in group T. Indexed diameter of RCA was significantly higher comparing groups 1 and 2 in patients with T.
The comparison of LVmass showed significant difference between groups. LVmass was highest in group 2 and lowest in group 3.

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There was no difference between the groups 1-3 concerning ratio LVmass to the summ of cross sectional areas of LAD and Cx.
Conclusions: Size of the coronary arteries in relation to BSA in TGA may vary depending on the artery pattern.
In TGA children late after correction LVmass may be still influenced by the earlier coexistence of VSD.
LVmass does not depend on the anatomy of the coronary arteries.

P-26

Associations between single nucleotide polymorphisms in the VEGF gene and congenital heart defects in human

Smedts H.P.M. (1), Isaacs, A. (2), Uitterlinden, A.G. (3), Duyn van C.M. (2), Costa de D. (1), Helbing W.A. (4), Steegers E.A.P. (1), Steegers-Theunissen R.P.M. (1,4,5,6)
Obstetrics and Gynaecology, Division of Obstetrics and Prenatal Medicine, Erasmus MC, University Medical Centre, Rotterdam, The Netherlands (1); Genetic Epidemiology, Erasmus MC (2); Internal Medicine, Erasmus MC (3); Paediatric Cardiology, Erasmus MC - Sophia Children's hospital (4); Epidemiology, Erasmus MC (5); Clinical Genetics, Erasmus MC (6)

Introduction: The tight regulation of vascular endothelial growth factor expression (VEGF) is essential for endocardial cushion formation. Disturbances in VEGF synthesis can result in congenital heart defects (CHDs), particularly outflow tract defects (OTDs). Three functional single nucleotide polymorphisms (SNPs) in the VEGF gene, -2578 C/A and -1154 G/A in the promoter region, and -634 G/C in the 5'UTR region are associated with increased VEGF expression for the -2578C and -1154G allele respectively. The objective was to investigate whether 3 functional SNPs in the VEGF gene are associated with congenital heart defects in human.
Methods: A case-control study was conducted in an ethnically homogenous population in the Western part of The Netherlands. At the study moment of 16 months after the index-pregnancy, blood samples were collected from 294 CHD case triads, including 194 OTDs comprising of VSD (n = 81), AVSD (n = 31), Fallot's tetralogy (n = 30), pulmonary stenosis (n = 46) and aortic stenosis (n = 6), and 317 control triads. Genotyping was performed by TaqMan discrimination assay. Genotype and haplotype frequencies were compared between case- and control triads by χ^2 test. The family based association test (FBAT-o) was used to establish linkage and association for the VEGF alleles and haplotypes.
Results: Allele frequencies of the three VEGF polymorphisms were comparable in case and control triads. The genotypes were in Hardy-Weinberg equilibrium. VEGF alleles -2578C and -1154G were transmitted more frequently to CHD cases, $P = 0.02$ and $P = 0.04$, and in particular to OTDs, $P = 0.003$ and $P = 0.002$. The VEGF -2578A/-1154A/-634G haplotype was significantly less transmitted to OTDs ($P = 0.002$). There was no evidence for a parent of origin effect.
Conclusion: The VEGF -2578C and -1154G alleles were overtransmitted to CHD cases, in particular those with OTDs. This data should be confirmed in replication studies.

P-27

An atherogenic maternal lipid profile is a risk factor for congenital heart defect offspring

Smedts H.P.M. (1), Uitert van E.M.(1), Valkenburg O. (2), Laven, J.S.E. (2), Eijkemans M.J.C. (3), Lindemans J. (4), Steegers E.A.P. (1), Steegers-Theunissen R.P.M. (1,5,6,7)
Obstetrics and Gynaecology, Division of Obstetrics and Prenatal Medicine, Erasmus MC, University Medical Centre, Rotterdam, The Netherlands(1); Obstetrics and Gynaecology, Division of Reproductive Medine, Erasmus MC (2); Public Health, Erasmus MC (3); Clinical Chemistry, Erasmus MC (4); Epidemiology, Erasmus MC (5), Clinical Genetics (6), Paediatric Cardiology, Erasmus MC-Sophia Children's Hospital (7)

Objectives: Hyperglycaemia and hyperhomocysteinaemia in early pregnancy contribute to development of congenital heart defects (CHD). In adult life both metabolic derangements also increase the risk of atherosclerosis. We investigated whether derangements in the maternal lipid profile, determinants of adult cardiovascular disease, are also associated with CHD offspring.
Methods: At 16 months post partum, a case–control study was conducted among 261 mothers of a child with CHD including 179 outflow tract defects (OTD) and 82 non-outflow tract defects (non-OTD), and 325 controls. Pregnant or lactating mothers or those who changed their diet were excluded. Primary outcome parameters were total cholesterol (C), LDL-cholesterol (LDL-C), HDL-cholesterol (HDL-C), triglycerides (TG), Apolipoprotein B (ApoB) and Apolipoprotein A-I (ApoA-I). Biomarkers ((geometric) means) were compared by Student's T-test. Risk estimates were expressed in Odds ratios (OR) and 95% confidence intervals (95%CI) and adjusted for age, diabetes, BMI, and periconception folic acid supplement use.
Results: Biomarker concentrations are shown in the table. CHD risk increased with rising maternal ApoB concentrations (Ptrend = 0.001). ApoB concentrations above 85 mg/dL almost two-fold increased CHD risk, OR 1.8 (95% CI: 1.3-2.6), and resulted in a population-attributable CHD risk of 21.1%.
Conclusions: An atherogenic maternal lipid profile is associated with the risk for CHD offspring. Thus, having CHD offspring may also be an early predictor for the mother to develop adult (cardio) vascular disease.

	total CHDs (*n* = 261)	OTDs (*n* = 179)	non-OTDs (*n* = 82)	controls (*n* = 325)
C, mmol/L*	4.8 (3.6 – 6.5)†	4.8 (3.6 – 6.3)	5.0 (3.7 – 6.8)‖	4.7 (3.5 – 6.4)
HDL-C, mmol/L	1.46 (0.93 – 2.15)	1.42 (0.93 – 2.05)	1.54 (0.91 – 2.19)	1.48 (0.96 – 2.09)
TG, mmol/L*	0.93 (0.48 – 1.97)	0.95 (0.47 – 2.14)	0.87 (0.48 – 1.88)	0.93 (0.50 – 1.99)
LDL-C, mmol/L*	3.2 (2.1 – 4.8)†	3.2 (2.0 – 4.7)	3.2 (2.2 – 5.2)‖	3.0 (1.9 – 4.6)
ApoA-1, mg/dL	145.2 (113.1 – 179.9)	143.8 (113.0 – 176.8)	148.2 (110.8 – 183.5)	146.5 (114.5 – 184.7)
ApoB, mg/dL	84.0 (57.9 – 115.2)‡	84.6 (57.5 – 118.6)§	82.8 (57.9 – 113.0)	80.0 (56.4 – 114.0)

*geometric mean (p5-p95); total CHDs vs. controls, †P < 0.05, ‡P < 0.01; OTDs vs. controls, §P < 0.01; non-OTDs vs. controls, ‖P < 0.05

P-28

Fibrillin-1 Gene Polymorphism and Risk of Mitral Valve Disease

Ozdemir O. (1), Olgunturk R. (2), Karaer K. (3), Ergun M.A. (3), Tunaoglu F.S. (2), Oguz D. (2), Kula S. (2)
Department of Paediatric Cardiology, Kecioren Education and Research Hospital, Ankara, Turkey (1); Department of Paediatric Cardiology, Gazi University, Medical Faculty, Ankara, Turkey (2); Department of Medical Genetic, Gazi University, Medical Faculty, Ankara, Turkey (3)

Introduction: Mitral valve prolapse (MVP) (autosomal dominant pattern for familiar cases, the role of fibrillin genetic variant in the MVP has locally been studied), rheumatic carditis (the mitral valve is the primary focus of endocardial involvement, but genetic data isn't found about different involvements of mitral valve in the rheumatic carditis), and connective tissue diseases such as Marfan

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syndrome (MFS) (autosomal dominant pattern, mutation in the fibrillin 1 gene on chromosome 15) and Ehlers-Danlos syndrome (EDS) (mutations in collagen gene), the most common anomalies of the mitral valve apparatus throughout childhood. Fibrillin (FBN) is one of the structural components of the elastin-associated microfibrils found in the mitral valve.
A case-controlled study has performed to investigate the relationship between FBN1 gene polymorphisms and risk of mitral valve disease and evaluate the genotype and phenotype correlation in the patients.
Methods: Thirty seven patients with MVP, 40 patients with rheumatic carditis diagnosed by clinical evaluation and echocardiography and 59 age- and sex-matched normal controls were studied. Polymorphisms of intron 52 and intron 56 of the FBN1 were identified by polymerase chain reaction (PCR)-based restriction analysis.
Results: There was a significant difference in the distribution of FBN1 intron 56 genotypes ($p = 0.001$) and allelic frequency ($p = 0.021$) between the cases and controls in favor of healty children. There was not genotype and phenotype correlation between mild/moderate cases and severe cases with mitral valve disease for intron 56 polymorphisms. A significant difference was not seen in genotype distribution or allelic frequency between the cases and controls for intron 52 polymorphism ($p = 0.738$).
Conclusions: Patients with mitral valve disease (MVP, connective tissue diseases or rheumatic carditis) have higher frequencies of FBN1 intron 56 GC genotypes. Healty children have higher frequencies of FBN1 intron 56 CC genotypes. We speculate that the higher frequency of FBN1 intron 56 G allel increases the risk of mitral valve disease.
Key words: Mitral valve disease, fibrillin-1, mitral valve prolapse, fibrillinopathies, acute rheumatic fever.

P-29

The Effect of Liver Transplantation on Cardiac Function, Hepatopulmonary Syndrome, and Portopulmonary Hypertension

Varan B. (1), Özçay F. (2), Kanra S. (2), Gökdemir M. (1), Cındık N. (1), Özbay F. (2), Tokel K. (1), Haberal M. (3)
Başkent University Faculty of Medicine, Ankara, Turkey, Department of Pediatric Cardiology (1), Department of Pediatric Gastroenterology (2), Department of General Surgery (3)

Introduction: Cardiac and pulmonary complications may occur during the course of chronic liver disease which are mainly cirrhotic cardiomyopathy and hepatopulmonary syndrome. There are scarce data in children about the reversibility of these complications after liver transplantation.
Aim: To investigate occurence of pulmonary arteriovenous fistula (AVF), hepatopulmonary syndrome and portopulmonary hypertension and to evaluate cardiac function after liver transplantation.
Patients and Method: Fifty-nine patients (7.9 ± 5.3 years old) who underwent liver transplantation for cirrhosis or acute liver failure are included in the study. Arterial blood gases and pulse oximetry are used to detect hypoxemia. Echocardiography and contrast study were used to evaluate cardiac function, pulmonary AVF, and portopulmonary hypertension.
Results: Contrast echocardiography was done in 39 patients before transplantation and 10 (25.64%) had pulmonary AVF, 8 had hepatopulmonary syndrome. There was no difference between pulmonary AVF(+) and (-) patients in terms of sex, transplantation age, cause of cirrhosis, presence of ascites, clubbing, and varices, albumin and bilirubin levels, prothrombin time, pCO2, mortality rate, and Pediatric End Stage Liver Disease score. Alveolar-arterial oxygen gradient was higher and pO2 was lower in AVF(+) group. Contrast echocardiography was negative in all of seven patients who were reevaluated for AVF 6 months to 3 years after liver transplantation. Alveolar-arterial oxygen gradient and pO2 levels were normal. Portopulmonary hypertension was diagnosed in three of 47 patients (6.4%) before transplantation in whom estimated pulmonary artery pressure could be calculated. After transplantation pulmonary artery systolic pressure returned to normal levels in two of three patients and the other patient had died. Left ventricular end-diastolic dimension and left ventricular mass indexed for body surface area decreased after transplantation.
Conclusion: Hepatopulmonary syndrome, portopulmonary hypertension and cardiac hypertrophy are reversible in cirrhotic patients after liver transplantation.

P-30

Risk factors for restenosis after isolated discrete subaortic stenosis removal

Kaneva A., Pilossoff V., Tzonzarova M., Lazarov S.
National Heart Hospital, Sofia, Bulgaria

Restenosis (RS) is e main complication after discrete subaortic stenosis (DSS) removal. The aim of the study was to define the risk factors for RS and reoperation (ReOp) in patients with isolated DSS (IDSS) and favorable immediate postoperative result. RS was defined as PG > 25 mm Hg during follow up.
For a 17 year period 66 IDSS patients were operated on. The study group comprises 52 patients with favorable early postoperative result (peak gradient (PG) < 25 mm Hg)- 43 membrane, 9 fibro-muscular shelf; preoperative PG 79 ± 22 mm Hg, removed by resection in 21, enucleation in 31 and additional myectomy in 31. After mean follow up 9 ± 5.3 (1-19) years PG increased from 7.8 ± 7.6 mm Hg to 47.4 ± 36.4 mm Hg ($p = 0.0001$). Patients were divided in two groups: Group 1- no RS (32 pts.) and Group 2– RS (20 pts). The type of IDSS, distance to the aortic valve; operative technique were not different between groups. Patients with RS were more frequently operated below age of 7 years ($p = 0.012$), had higher preoperative PG ($p = 0.011$), immediate postoperative PG ($p = 0.0001$) and year rate of PG increment ($p = 0.008$), smaller aortic ring ($p = 0.007$) and longer postoperative follow up ($p = 0.018$). The narrow aortic ring and year rate of PG increment > 2.1 mm Hg/year enter the multiple regression model for RS (OR 13 $p = 0.027$).
During the follow up ReOp was performed in 8 patients (15.4%). The freedom from ReOp diminished from mean 97.7% at 5; to 92.6% at 10; 78% at 15 and 68% at 17 year after operation. Cox regression analysis revealed the age of operation < 7 year as an independent risk factor for ReOp- OR 4.57, $p = 0.042$.
Conclusion: Multiple factors contribute to RS after IDSS removal- anatomic- size of the aortic annulus; hemodynamic- quality of immediate result. RS that requires ReOp develops in children with high preoperative potential (operated below the age of 7 years). It is likely that in some children IDSS removal does not interrupt the natural course of the disease.

P-31

Respiratory Infections in Down Syndrome with and without Congenital Heart Disease. The CIVIC 21 Study in Spain 2006-2007.

Medrano C., Garcia-Guereta L., Lirio J., Garcia-Perez J., Ballesteros F., Casaldaliga J., Cuenca V., Escudero F., Garcia de la Calzada L., Grueso J., Insa B., Luis M., Mendoza A., Carretero J., Rodriguez M.M., Suarez P., Quero C.
In representation to the CIVIC group from the Spanish Society of Pediatric Cardiology and Congenital Heart Diseases (SECPCC).

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Objectives: In the CIVIC study we described hospitalization rates of respiratory infections in young children with Hemodinamically Significant Congenital Heart Disease (CHD-HS) and specifically Down Syndrome as a significant risk factor. In the further CIVIC 21 study we therefore evaluated Down Syndrome infants, with and without congenital heart disease.
Methods: Seasonal (from October 2006 to April 2007), epidemiological, multicentric (53 public hospitals in Spain), observational and prospective. 1084 patients less than 24 months old were included. 279 with chromosomal diagnosis of 21 trisomy, classified in relation to congenital heart disease in: Group 1 (N=105): without CHD (No CHD); Group 2 (N=38): with non-hemodinamically significant CHD (i.e. patent foramen ovale, minimum patent ductus arteriosus or minimum muscular ventricular septal defect, discrete pulmonary valvular stenosis) (CHD-Non-HS); and Group 3 (N=135): with hemodinamically significant CHD (CHD-HS). Other 805 patients without Down Syndrome and with CHD-HS were included in the general study.
Results (see table): The global hospitalization rate is 13.1% (CI95%: 11.2-15.2%); In the Non-Down patients is 11%. In the Down patients is 19.1% with a significant difference (p=0.037) between groups. The most frequent clinical diagnosis was bronchiolytis. Respiratory syncytial virus (RSV) was the most frequent microbiological diagnosis, with significant differences (p=0.03) between group in Down patients. Prophylaxis against RSV, recommended in cases with CHD-HS, was completed in a lower rate in the Down patients specific group with CHD-HS. Hospitalization in the intensive care unit and mechanical ventilation rates in hospitalized patients were higher in the Down syndrome patients versus non Down: 28.4% vs. 22.9% and 14.7% vs. 11.4% respectively.

Down syndrome	No	Yes			
Congenital Heart disease type	CHD HS	Total Down	CHD HS	CHD Non HS	No CHD
Hospitalization rate (%)	11	19,1	23	26,3	11,4
Hospitalization by RSV infection (%)	3,2	7,5	8,6	15,8	3,0
RSV prophylaxis (%)	83,4	39,9	74,6	7	7

Conclusion: In children with Down Syndrome the presence of Congenital Heart Disease Non Hemodinamically Significant is associated with the higher hospitalization rate and the higher specific Respiratory Syncytial Virus infection rate. Probably Respiratory Syncytial Virus prophylaxis will be considered in this group.

P-32
Safety profile of Clopidogrel in children with heart disease
Edwards S. (1), Naqvi N. (1), Jiggins M. (2), Franklin R.C.G. (3), Slavik Z. (4)
Royal Brompton Hospital, Royal Brompton & Harefield NHS Trust, London, United Kingdom (1)

Introduction: Following evidence for Aspirin resistance in adult cardiac patients and the risks, and poor compliance in many paediatric patients with Warfarin based anticoagulation, we explored the safety of Clopidogrel use in children with heart disease.
Methods: Retrospective review of patients' pharmacy and hospital records was complemented by prospective questioning of patients and their relatives in 45 children (median age 64months, range 3-164 months) with congenital or acquired heart disease receiving Clopidogrel with (96%) or without Aspirin treatment in tertiary paediatric cardiac referral centre between January 2004 and December 2007.
Results: None of the patients received Clopidogrel after artificial valve insertion. All but 5 patients (11%) had complex congenital heart malformations. Clopidogrel was used with Aspirin as an alternative to Warfarin following total cavopulmonary connection in 23 patients (51%) all of whom had a degree of needle phobia and in whom it was often difficult to obtain blood samples. Other patients included those with dilated cardiomyopathy, following shunts and conduit procedures, and after placement of stents and septal device occluders. The mean daily dose of Clopidogrel was 1.5 mg/kg (range 1.0–2.4 mg/kg). Increased bruising was reported in 10 patients (23%) treated by Clopidogrel and Aspirin and in no patients treated with Clopidogrel only over follow-up period between 1 and 48 months (median 18 months). No life-threatening bleeding complications were encountered and none of the patients developed clinically and/or echocardiographically apparent thrombo-embolic complications.
Conclusion: Based on our data, the use of Clopidogrel in paediatric patients with heart disease was not associated with any clinically significant complications. We also believe that Clopidogrel added to Aspirin is a safe alternative to Warfarin treatment in selected patients after total cavopulmonary connection in the short term follow-up, particularly in those who are needle phobic. Increased bruising is to be expected in about one quarter of the patients where Clopidogrel is used with Aspirin. This has minor clinical implications but may be of importance in the child protection area with significant social impact.

P-33
Long-term outcome of pediatric heart transplant recipients from donation after cardiac death
Tissot C. (1), Pietra B.A. (1), Boucek M.M. (2), Campbell D.N. (1), Ripe D.W. (1), Mashburn C. (1), Mitchell M.B. (1), Ivy D.D. (1), Miyamoto S.D. (1)
The Children's Hospital, Denver, Colorado, US (1); Joe DiMaggio Children's Hospital, Hollywood, Florida, US (2)

Introduction: Donation after cardiac death (DCD) has been introduced into clinical practice to address waning donor supply and associated increased risk of death while on the transplant waiting list. The most critical concern is damage to the donor heart derived from prolonged warm ischemia time which could affect graft function. Few data are available concerning the long-term outcome of pediatric heart transplant recipients from DCD.
Methods: We retrospectively reviewed the outcome of the pediatric heart transplant recipients from DCD in our institution. The protocol was approved by the institutional review board and informed consent was obtained from all patients. Anti-thymocyte globulin, intravenous immunoglobulins and methylprednisolone were used for induction followed by cyclosporine and mycophenolate mophetyl (MMF) at hospital discharge.
Results: Three patients aged 5, 3 and 3 months, all blood group O underwent orthotopic heart transplantation from DCD for complex congenital heart disease. The mean time on the waiting list was 37 days for DCD recipients as compared to 90 days for blood group O infants (n=89) transplanted from donors meeting brain death criteria. The mean weight at transplant was 3.6±0.4 kg. The mean ischemic time was 162±52 minutes (56±27 minutes warm ischemia). The mean duration of inotropic support was 9±7 days and the mean length of hospital stay 20±14 days. One patient was on extracorporeal membrane oxygenation (ECMO) support prior and after transplantation, underwent an atrioseptostomy when on ECMO closed percutaneously 6 months later. The cardiac function was normal in all at one month post transplant (mean shortening fraction 39±6%). There were 2 episodes of acute graft

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rejection in the first year post-transplant. A median of 1.5 episodes of infection per patient was documented. Current immunosuppression is cyclosporine and azathioprine for two patients, cyclosporine and MMF for one. No patient has transplant coronary artery disease. All patients are alive with a survival range of 8 months to 3.5 years.
Conclusion: Heart transplantation from DCD has a favorable outcome in infants. It is particularly suitable for blood group O patients as it decreases their wait time and risk of death on the waiting list, without an increase in post-transplant complications.

P-34
Cardiac features of Kawasaki Disease in France: a single-center esperience
Di Filippo S. (1), Lega J.C. (2), Cimaz R. (3), Cochat P. (3), Floret D. (3), Nicolino M. (4), Bellon G. (4), Bozio A. (1)
Hôpital Cardiologique, pediatric cardiology, Lyon, France (1); Hôpital Jules Courmont, internal and vascular medicine,Lyon, France (2); Hôpital Edouard Herriot, Pediatrics, Lyon, France (3); Hôpital Debrousse, Pediatrics, Lyon, France (4)

Little is known about Kawasaki Disease (KD) features in France. The aims of this retrospective single-center study were to assess the characteristics of a French KD population over a long period of time, and to describe cardiac lesions.
Methods: We retrospectively reviewed the medical records of 417 patients referred to echocardiography for KD suspicion between August 1983 and April 2007.
Results: 210 patients met criteria for diagnosis of KD, at the age of 2.7±2.5 years (median 2). Fever was present in all patients. Time to diagnosis was 7±4.6 days (median 6days), time to hospitalization 5.7±4.3 days (median 5days). Time to first echocardiography was 11.4±7.8 days (median 9days), shorter in more recent period. Median time to IVIG administration was 8 days (1 to 39). At initial evaluation, 63.8% were free from cardiac lesions, 23.8% (52 cases) had coronary lesions (aneurisms : 25, dilatation : 27) and 12.4% had « hyperechogen » coronary arteries. Among 52 cases with abnormal coronary arteries, 40 were < 5mm in diameter, 9 were 5-8 mm, and 3 were > 8mm (giant aneurisms): one third localized on one coronary vessel, one third on 2 and one third one all 3 coronary arteries. Echographic pericarditis was found in 31 patients, mitral insufficiency in 20 and aortic insufficiency in 2. All patients recovered, except 1 who died from cardiogenic shock due to ruptured chordae. Coronary lesions resolved in 17 of 52 cases (32.6%) and persisted in 35 (67.4%, i.e. 16.7% of all patients): 14 with aneurisms and 19 with dilatations. No patient developped significant long-term coronary artery stenosis. The incidence of aneurisms was lower over the past decade (7.2%).
Conclusion: In our experience, the occurrence of coronary lesions in KD have lessened over time and long-term cardiac outcome is favourable despite persistent coronary lesions.

P-35
Sub-optimal Dosing Patterns of ACE-inhibitors in Paediatric Cardiology Patients
Roche S.L., Balasingam M., Timberlake K., Kantor P.F.
The Hospital For Sick Children, Toronto, Canada

Introduction: Angiotensin-converting enzyme inhibitors (ACEi) are frequently prescribed to paediatric cardiac patients. Adult data suggest that underutilization of ACEi is frequent. We sought to assess the adequacy of captopril prescription in our paediatric population and examine staff attitudes towards ACEi.
Methods: Fifty consecutive children (< 18 years) commencing captopril while cardiology inpatients between 2005/09/01 and 2007/03/19 were identified from our pharmacy database. Case notes of these patients were reviewed. After literature review, an "optimal" daily captopril dose was defined as 3mg/kg given in 3 divided doses and daily doses of > 2.7mg/kg as "on target". Patient doses at discharge and 8 weeks later were compared to these definitions.
Forty clinical members of the pediatric cardiology division were surveyed regarding attitudes to and use of ACEi.
Results: Median patient age was 2.7 (IQ 1-5.9) months. 66% of patients were prescribed captopril following cardiac surgery, 44% while on inotropes. The median hospital stay after first dose was 19 (IQ 10-38) days. On average the initial dose was increased 3.6 (SD 2.5) times prior to discharge. Asymptomatic hypotension was noted during up-titration in 11/50 patients. Renal impairment was not seen.
At discharge 34/50 remained on ACEi, 25 on captopril. 9/25 were within target range doses by hospital discharge, 15/25 were discharged on < 1/2 the optimal dose. At eight weeks after discharge only 23% of those remaining on captopril were on target doses.
Although 27/30 survey respondents felt captopril should be given three times a day opinion varied regarding an optimal dose. Only 13/22 who answered this question with a value suggested an answer between 2.7 and 3.3mg/kg/day. Opinion on dose up-titration varied immensely.
Conclusion: Captopril dosing of pediatric cardiology patients is sub-optimal at the time of hospital discharge and remains so at eight week follow-up. Although pediatric formularies recommend initial and safe maximum doses no guidance is given as to the best method of up-titration or target dosages. As evidenced here, the physician is left uncertain of an optimal dose and how best to achieve it. This, and not the occurrence of side-effects, was likely a limiting factor in utilization of ACEi within our patient population.

P-36
Neoaortic Aneurysm: Following Arch Reconstruction for Hypoplastic Left Heart Syndrome
Linter K.J. (1), Davies B. (2), Stumper O. (1), Reinhardt Z. (1), Stickley J. (1), Wright J.G. (1), Miller P.(1), Dhillon R. (1), Chikermane A. (1), Jones T.J. (2), Barron D.J. (2), Brawn W.J. (2), Giovanni J.V.D. (1)
Departments of Cardiology(1) and Cardiac Surgery (2), Diana, Princess of Wales Children's Hospital, Birmingham Children's Hospital NHS Trust, Birmingham, UK

Background: Staged surgical management for Hypoplastic Left Heart Syndrome (HLHS) is well established. Aortic reconstruction is achieved using a variety of techniques and materials. The normal size and growth profile of the reconstructed ascending aorta in this group of patients is not defined. We report a series of patients with a large ascending aorta. In this context rupture or dissection is unlikely but the risk is unknown. Compression of the left pulmonary artery requiring intervention is well documented.
Methods: Ten year retrospective review of 119 patients undergoing palliative surgery for HLHS. All patients underwent modified Norwood operation and subsequently completed the Fontan pathway. Measurements of aortic size were obtained at cardiac catheterization prior to second and third stages of surgery. A ratio was calculated for each patient of ascending:descending aorta dimension, measured from standard angiographic projections, and plotted against age and body surface area at time of catheter.
Results: Median follow-up for this cohort of patients is 7.0 years (range, 3.4 to 13.7 yrs) after stage 1. Angiography was available in standard projections for 76 patients. We calculated the mean ratio of ascending:descending aorta dimension for the group to be 2.5 (range 1.4 to 4.6). In four asymptomatic patients the ratio, of ascending aorta size compared to the native descending aorta, was above the normal

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distribution for the group (>3.4). Balloon angioplasty, for significant arch gradient, pre-dated neoaortic dilatation in 2/4 patients. During follow-up, one patient with asymptomatic progressive enlargement of the ascending aorta and arch with competent neoaortic valve underwent valve-sparing aortic replacement when maximal dimension of ascending aorta reached 6 cm, with the ratio measuring 4.6. Three patients remain under surveillance.
Conclusion: The implications of an enlarged or dilating reconstructed ascending aorta are unknown. Defining normal size and growth profile of the reconstructed arch in this group is important to identify patients in whom the ascending aorta dimensions are disproportionately large and potentially may be at risk.

P-37
Radial artery percutaneous coronary angiography in teenagers
Kirk C.R., Zaman A.
Freeman Hospital, Newcastle upon Tyne, UK

Introduction: The percutaneous radial arterial approach has gained favour in the adult population for coronary angiography with the advantages of a reduced incidence of vascular complications and quicker ambulation. We used this approach in a 12 year old patient post transplant undergoing coronary angiography in which the femoral access had been unsuccessful. Following this success we offered a radial approach to teenagers undergoing coronary angiography post cardiac transplantation and report our experience here.
Methods: 12 teenagers (13 procedures). 8 girls, 4 boys. A modified Allen's test utilising a pulse oximetry checked adequate ulnar flow. 2% lignocaine was used for skin anaesthesia. After access was obtained a cocktail of 2.5mg Verapamil, 3000 iu Heparin and 200mcg isosorbide dinitrate was administered into arterial sheath to reduce spasm. A 5F sheath was used with routine Judkin or Amplatz coronary catheters. Following the procedure haemostasis was achieved with a TR band left in place for 3 hours. All procedures were done as day cases.
Results: The mean (range) age was 16.1 yrs (12.1–18.8), mean weight 57.6 kg (39-81). 70% were under local anaesthetic. Radial access was successful in 85% of procedures. In one failure to access the radial artery occurred because EMLA cream had been applied and the artery was impalpable. 3 patients had significant spasm leading to abandoning the approach in one patient and failure to complete the study adequately in 2 patients. No vascular complications occurred. All but one of the patients would choose a radial approach in future.
Conclusion: Our experience demonstrates that the radial approach is technically possible in teenagers and in general is preferred by them. The radial approach is more challenging for the operator and this may have contributed to technical failures in accessing the radial artery and failing to adequately complete the study. Spasm is a significant issue and difficult to reverse once occurred. The use of premedication may reduce the incidence in this population. This is to our knowledge the first report of this technique in this age.

P-38
Clinical outcome of children with uncompacted left ventricle: a large experience from a single centre over 20 years.
Gagliardi M.G., Pilati M., Toscano A., Saffirio C., Del Pasqua A., Adorisio R., Sanders S.P.
Bambino Gesù Hospital, Rome, Italy

Background: Uncompacted left ventricle (UCLV) is an extremely rare congenital disorder. Due to the lack of reabsorption of endomyocardial trabeculations with consequent failure of the LV. Few and heterogeneous data are currently available.
The aim of this study was to identify the clinical characteristics and outcome in children.
Methods: Our database was searched for all children with UCLV between January 1985 and January 2006. Medical records were reviewed to document clinical characteristics and dysmorphic features. All the echocardiograms were re-analyzed.
Results: Among 234 children, 27 (10%, 11 F) were identified as suffering from UCLV. The median age at presentation was 1 year (range 0,1 to 14 years), and the median duration of follow-up was 6 years. The most common clinical presentation was low cardiac output and congestive heart failure (n = 10, 37%). Less frequent, the presence of murmur (n = 7, 25%), presence of other cardiac abnormalities (n = 7, 25%) or dysmorphic facies (n = 3, 11%).
During the follow up, 20 children (75%) remained in NYHA I-II, while 7 (25%) were in NYHA III-IV. Arrhythmias were recorded in 3 (n = 1 supraventricular and n = 2 ventricular tachycardia); in one case an internal cardioverter defibrillator was implanted. At the echo findings, the involvement of the LV was diffuse in 14 (53%), limited to the apex in 8 (29%) and located in the free wall in 5 (18%) and in 1 case exclusively to the right ventricle. Mean ejection fraction was 50 ± 21%: in 8 children (29%) severe LV dilatation and dysfunction (EF < 30%) were recorded. Restrictive cardiomyopathy was the final diagnosis in 19 (70%).
Ten children needed therapy during the follow up with digitalis, diuretics, ACE-inhibitors and in 3 pts also beta-blockers. In 5 warfarin was used. Two patients with reduced EF showed a progressive improvement of LVEF from 20% to 45% over 10 months. Three pts (10%) underwent a cardiac transplantation and 2 died (7%). The overall probability of transplant-free survival is 82% at a mean follow-up duration of 5 years.
Conclusion: This retrospective study confirm the better survival in pediatric age when compared with adult series. Restrictive pathophysiological pattern seem to prevail also in our children with UCLV.

P-39
Can treatment slow aortic dilatation in Marfan Syndrome? A Systematic Review of 103 Cases.
Chetaille Ph., St-Pierre J., Beaudoin EL., Lapointe A., Delisle G., Houde C., Côté JM.
Centre Mère-Enfant du Centre Hospitalier Universitaire de Québec, Québec, Canada

Objective: Marfan syndrome is an autosomal genetic disorder of connective tissue mainly associated with dilatation of the ascending aorta involving the sinus of Valsalva and predisposing to aortic dissection. Some pharmacological agents (ACE inhitors (ACEI) and AT1 antagonists (ATA)), beside their hemodynamic effects, might have a beneficial effect directly on the tissue matrix, preventing aortic medial degeneration. The objective of this study was to review the effect of pharmacological treatments among a cohort of Marfan syndrome patients in order to describe the effect on aortic dilatation.
Methods: We retrospectively reviewed 103 cases of Marfan syndrome patients (53 males). Diagnosis was made on the basis of a combination of a proven genetic mutation in FBN1 gene, positive family history and/or clinical phenotypic features of the syndrome. All echocardiographic data on follow-up were reviewed, and clinical events collected. Patients were stratified on the basis of their treatment (beta-blockers (BB), ACEI and ATA) versus no pharmacological therapy. Analyses were performed with SPSS package.
Results: Mean age at diagnosis is 8.2 years. Mean follow-up is 7.3 years. 47% were receiving a pharmacological agent as a monotherapy

(29.1% BB, 6.8% ACEI, 8.7% ATA). 72% had dilatation of the aorta. Patients on medication are older, have a longer follow-up, have more vascular events and have higher aortic root diameters and z-scores. Death occurred in 3 (3%) patients during follow-up (1 post-operative, 1 aortic dissection and 1 of unknown cause). 14 pts (13.9%) underwent vascular surgery during follow-up (13/14 were on medication). On follow-up, patients under medication had a significantly slower rate of dilatation of the aorta ($R2 = 0.1811$) than those without medication ($R2 = 0.6127$) independently from age. Because of small numbers in each group, different medications efficacy on aortic dilatation and clinical events could not be compared.
Conclusion: Marfan syndrome is a relatively frequent syndrome associated with severe cardiovascular and/or vascular complications. An early treatment with beta-blockers, ACEI or ATA might reduce the rate of progression of dilatation of the aorta. Whether angiotensin receptor blockade might prevent aorta from dissecting remains to be elucidated in large clinical prospective studies.

P-40

Congenitally Corrected Transposition in Situs Inversus: Report of an Original Series

Favia Guarnieri G., Adurno G., Tagliente M.R., Balducci G., Iorio F.M., Tomasco B., Troise D., Vairo U.
Pediatric Cardiology and Cardiac Surgery, San Carlo Hospital, Potenza - Italy

Introduction: Congenitally corrected transposition of the great arteries (CCTGA) in situs inversus, with [I,D,D] sequential ana¬tomy, is the mirror image of the more common type of CCTGA in si¬tus solitus [S,L,L]. In addition to peculiar anatomical features, including the D loop like course of the atrio ventricular conducting bundle from a posterior node, this abnormality is usually complicated by variable degrees of associated defects such as pulmonary outflow tract obstruction and VSD.
Methods: We report an original series of 19 patients with [I,D,D] CCTGA (11 males), whose age at diagnosis ranged between 1 day and 10 years (median 9 days). In one case diagnosis was made post mortem. In the other 18 cases, diagnosis was obtained by 2 D echocardiography and confirmed by angiocardiography in 16.
Results: Nine patients had dextrocardia (47%), 8 had levocardia (42%) and 2 had mesocardia (10%). Four patients, with dextrocardia, had no associated cardiac anomalies. A malalignment VSD was present in 14 cases (74%). In two patients with left ventri¬cular hypoplasia, there was an associated muscular VSD. Pulmonary outflow tract obstruction was seen in 18 patients (95%): valvar and subvalvar in 10 and atretic in the remaining 8. The patency of a ductus arteriosus in 10 patients (53%) represented the only source of pulmonary blood flow in the cases associated with pulmonary atresia. Third degree atrio ventricular block was present in only one patient (5%), since birth. Of the 3 patients without associated intracardiac anomalies, 2 patients underwent cardiac catheterization for an uncommon combination with a vascular ring, compressing both trachea and esophagus in one. Surgical procedures included: systemic-pulmonary shunt in 5, anatomical repair in 3, physiological repair in 4, division of vascular ring in 1, systemic-pulmonary shunt plus bidirectional cavo-pulmonary anastomosis in 2. There were 3 early death (one after shunt and two after physiological repair), and 3 late death.
Conclusion: In presence of situs inversus, the potential combination of CCTGA may be expected. Our series confirms that patients with [I,D,D] CCTGA have a relatively uniform recurrence of combined anatomical lesions, with prevalence of severe pulmonary outflow tract obstruction and VSD.

P-41

Left atrioventricular valve stenosis after Atrioventricular Septal Defect repair: Intraoperative Transesophageal Echocardiography predictors and outcome

Cabral M.B.C., Binotto M.A., Takigawa L.M., Mathias Jr W., Guerra V.C.
Heart Institute (InCor), University of São Paulo Medical School, Brazil

Introduction: Left-sided atrioventricular valve (LAVV) is the main cause for reoperation in patients after repair of atrioventricular septal defect (AVSD). Although LAVV regurgitation has been the major determinant of late morbidity, LAVV stenosis has not been fully addressed previously. We reviewed our experience with AVSD repair in order to investigate long term results and risk factors for residual LAVV stenosis.
Methods: Between 2001 and 2007, 221 patients underwent surgery for AVSD repair. Routine intraoperative TEE was done in all patients above 3.5 Kg. Among them, 18 (Group I) presented with residual moderate to severe LAVV stenosis (mean diastolic pressure gradient >8 mmHg). Median age at operation was 11 months; median weight was 6.4 Kg; 44% were partial AVSD. Group II (26 patients) with trivial/mild residual LAVV regurgitation after surgery. Groups I and II were similar regarding age, weight and BSA. Off-line measurements of total annulus, LAVV annulus (z scores) and interpapillary muscle distance were obtained from the pre and postoperative TEE images.
Results: Six patients underwent reoperation, including two requiring valve replacement. There were 3 deaths (16.6%), all in Group I. Mean follow-up was 26 months. Preoperative total common valve annulus and LAVV annulus, measured by intraoperative TEE, were not different between groups. However, the preoperative distance between the papillary muscles were significantly smaller in Group I ($p = .01$) Postoperatively, LAVV annulus were reduced comparing with preoperative values, in Group I ($p < .001$), but not in Group II. Partial AVSD was not a risk factor for residual stenosis. Regarding surgical approach (modified single, double patch, cleft closure, annulus reduction) there were no differences between groups. The LAVV mean pressure gradient remaining stable (by transthoracic echo) over the time in 13 patients (mean follow-up of 16 months). In two, the LAVV mean pressure gradient decreased. All patients remain in NYHA class I or II.
Conclusion: The distance between the two left papillary muscles (which support the trileaflet LAVV and also predict the mural leaflet length) was significantly smaller in those patients with residual LAVV stenosis. The LAVV annulus decreased after repair in patients whose LAVV became stenotic.

P-42

Angiotensin Converting Enyzme Gene Polymorphism Frequency in Normotensive Children with Positive Family History of Essential Hypertension

Kilic Z. (1), Camci L. (2), Dinleyici E.C. (2), Muslumanoglu H. (3), Ucar B (1)
Eskisehir Osmangazi University Faculty of Medicine, Department of Pediatric Cardiology (1), Pediatrics (2) and Genetics (3), Eskisehir, Turkey

Introduction: The classic candidate gene approach continues to be the most prevalent tool in the search for the genetic basis of essential hypertension (EHT) but there is some limited and sometimes even contradictory information. The aim of this study was to evaluate the possible relationship between blood pressure (BP) and angiotensin converting enyzme (ACE) gene polymorphism in normotensive children with positive family history of EHT.
Methods: A randomly selected 376 schoolchildren (147 boys, 229 girls) aged between 7–17 years, residing in urban parts of Eskisehir,

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were enrolled. Patient group was subdivided to "first degree relative group" and "second degree relative group" according to the presence of positive history for HT in parents or grandparents, respectively.
Results: Diastolic BP (DBP) and mean BP (MBP) levels were higher in children with positive family history for EHT than the controls ($p < 0.0001$ for both), systolic BP (SBP) levels were also higher in both groups than the controls without statistical difference ($p > 0.05$). Allelic frequencies of DD genotype of ACE gene were higher in children with positive history in the first and second degree relatives for EHT than the controls ($p < 0.05$), but ID and II genotypes frequencies were similar between the study group and the controls ($p > 0.05$). Children with positive family history of EHT and DD genotype of ACE have significantly higher SBP, DBP, MBP levels ($p < 0.05$ for all) than the children with ID or II genotypes. Children with positive history for EHT in the first degree relatives and DD genotype of ACE have significantly higher SBP, DBP, MBP levels ($p < 0.05$ for all) than the controls. Children with positive family history for EHT in the second degree relatives with DD genotype of ACE have significantly higher DBP than the children with II genotype of ACE ($p < 0.01$).
Conclusion: We consider that ACE gene DD genotype is common and BP levels were higher in children with positive family history of EHT. Because of ACE gene can play crucial role in EHT pathogenesis, we suggest the usage of this parameter for the prevention of complications associated with HT by establishing early preventive measures in children with genetic predisposition.

P-43
Pulmonary hypertension complicating isolated atrial septal defect in infancy
Pfammatter J.P. (1), DiBernardo S. (2), Götschmann S. (1), Pavlovic M. (1), Sekarski N. (2)
Pediatric Cardiology, University Children's Hospitals Berne (1) and Lausanne (2), Switzerland

Atrial septal defect (ASD) typically is asymptomatic in infancy and early childhood and elective defect closure usually is performed at an age between 4–6 years. Severe pulmonary hypertension (PHT) complicating an ASD is seen in adulthood and has only occasionally been reported in small children.
A retrospective study was undertaken to evaluate the incidence of severe PHT complicating an isolated ASD and requiring early surgical correction in the first year of life. 355 pediatric patients underwent treatment for an isolated ASD either surgically or by catheter intervention during a 10 year period (1996–2006) at two tertiary referral centers. 297 patients had secundum ASD and 58 primum ASD with mild to moderate mitral regurgitation. 8 infants were found with isolated ASD (six with secundum and two with primum ASD) associated with significant PHT, accounting for 2.2% of all ASD patients in our centers. These 8 infants had invasively measured pulmonary artery pressures between 50 and 100% of systemic pressure. Median size of the ASD at the time of surgery was 14mm (7–20). They were operated in the first year of life and had complicated postoperative courses requiring specific treatment for PHT for up to 16 weeks (median 12) postoperatively. Compared to ASD patients without PHT these infants had prolonged postoperative ICU stay of 5–9 days (median 8) and prolonged perioperative overall hospital stay of 8–32 days (median 15). Ultimate outcome in all 8 infants was good with persistent normalization of pulmonary pressures during mid-term follow-up of between 8 to 60 months (median 28). All other ASD patients had normal pulmonary pressures and mean age at defect closure was higher being 6.2 years for secundum ASD and 3.2 years for primum ASD.
In conclusion, ASD is rarely associated with significant PHT in infancy but then requires early surgery to normalize the prognosis of the patients.

P-44
The unnatural history of Critical Aortic Stenosis
Maschietto N. (1), Grosso G. (1), Reffo E. (1), Vida V.L. (2), Padalino M.A. (2), Stellin G. (2), Milanesi O. (1)
Department of Pediatrics, Cardiology Unit, University of Padua-Medical School, Italy (1); Institute of Cardiovascular Surgery, Pediatric Cardiac Surgery Unit, University of Padua-Medical School, Italy (2)

Background. Critical aortic stenosis is still a challenging lesion both for surgeons and cardiologists. The optimal treatment for these newborns is controversial. The purpose of our study is to present our experience and to identify any early predictors associated with a better outcome.
Methods: Between January 1990 and January 2007 in our Institution 36 patients under 30 days of age have undergone balloon aortic valvuloplasty (BAV) for critical aortic stenosis (cAS).
Results: 23 patients (pts) had cAS alone and 13 had associated lesions (mitral stenosis, coarctation of the aorta (CoAo), mildly hypoplastic LV (HLV) and endomiocardial fibroelastosis). 17 pts were on PGE1 at the time of the first cardiac catheterization. All patients underwent BAV through a transcarotid approach. Mean age at valvuloplasty was 9 days (2–30). No procedural mortality.
Four pts (11%) failed to achieve a biventricular circulation. Twenty five interventions/reinterventions on the aortic valve were performed in 18 pts: 10 pts underwent a second valvuloplasty, and 15 surgical interventions were performed in 13 pts. Eight of the 15 surgical interventions were due to the development of a significant sub-aortic stenosis.
One patient (2.7%) died suddenly few months later the first BAV, two pts (5.5%) after a Norwood procedure and 4 pts (11%) after a surgical intervention.
At 1, 5 and 10 years of follow-up, freedom from significant AS were respectively 73%, 48% and 36%, from significant AR were 81%, 76% and 52% and from surgical intervention were 63%, 53% and 41%. The overall survival at 1, 5 and 10 years were 86%, 79% and 79%.
We have identified several early predictors for a worse outcome. CoAo (HR 13.7), HLV (HR 13.1) and a smaller relative reduction in the AS gradient at the initial procedure (HR 1.04) were associated with a significant restenosis, a more than mild AR after the first BAV with significant AR (HR 11.4), PGE1 with surgical intervention (HR 5.2) and the presence of any associated lesions at birth with death (HR 31.5).
Conclusion. Critical aortic stenosis is a life long disease with a very severe prognosis in terms mortality and morbidity despite any treatment.

P-45
Length, weight and head circumference in newborns with hypoplastic left heart syndrome
Tulzer G., Peterlechner T., Arzt W., Mair R.
Children's Heart Centre Linz, Linz, Austria

Altered hemodynamics in fetuses with hypoplastic left heart syndrome (HLHS) may lead to impaired somatic growth, especially head circumference, which might impact long term neurological outcome.
The purpose of this study was to assess length, weight and head circumference and its relation to ascending aortic diameter in newborns with HLHS.

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Patient and Methods: Patient charts and echocardiograms of 105 consecutive newborns (64% male, 36% female) with HLHS were retrospectively reviewed for birth weight and length; the diameter of the ascending aorta right above the sinutubular junction was measured from the first available postnatal echocardiogram. Aortic atresia with retrograde flow in the ascending aorta was present in 66% of patients, aortic stenosis in 33%.
Results: Median length was 49.24 cm (41–55 cm), median weight was 3200 g (1320–4700 g), median head circumference was 34 cm (26.5–38 cm).

Percentile	<3	<10	>90	>97
Weight	4.85	10.68	2.19	0.97
Length	7.37	23.16	3.16	2.11
Head circumference	4.21	18.95	3.16	0

When comparing the head circumference to the diameter of the ascending aorta there was a significant correlation ($p < 0.05$), however if the percentile of the head circumference was used instead for correlation there was no more significance.
Conclusion: newborns with HLHS have shorter than average length, lower than average birthweigths and head circumferences although the vast majority was within the 3rd and 97th percentile. This confirms the findings from earlier studies, however the prevalence of microcephaly was lower than previously reported. The diameter of the ascending aorta might not be a reliable predictor for cerebral growth. Whether impaired somatic growth is in fetuses with HLHS is a primary problem or the result of altered hemodynamics and whether it impacts long term neurological development still has to be determined.

P-46
Attitude toward illness in children with mitral valve prolapse

Janiec I., Werner B., Florianczyk T.
Department of Pediatric Cardiology and General Pediatrics. The Medical University of Warsaw, Poland

Aim: The goal of the study was to assess attitude toward illness in children with mitral valve prolapse (MVP) and comparison with reported symptoms and results of cardiovascular system examinations.
Patients and Methods: The prospective study involved 65 children 46 girls, 19 boys aged 8-18 years (average 13.8 years) divided into 4 groups according to age and into 2 groups according to CATIS: achieving 1-3 score and 4-5 score. Polish version of Child Attitude Toward Illness Scale (CATIS) and own questionnaire of 18 clinical symptoms were used. CATIS is a short self-reported instrument designed to provide a complex assessment of how favorably or unfavorably children feel about having chronic physical condition. The scale score range from 1 to 5, higher score reflects more positive attitude. The results of Holter ECG and treadmill test (t.t.) were analyzed, statistical analysis based on Fisher exact probability test.
Results: Child Attitude Toward Illness Scale Results:

	CATIS score – mean values			
	8-10 years	11-13 years	14-16 years	17-18 years
all	3.48	3.57	3.52	3.46
girls	3.23	3.54	3.50	3.42
boys	4.00	3.70	3.55	3.58

Holter ECG and treadmill test results versus CATIS score:

		Number of children (% of all 65 children)						
		Holter ECG arrhythmia			Treadmill test result		Effort tolerance in t.t.	
CATIS score	No of children	SVEBs	VEBs	ventricular couples	positive	negative	normal	diminished
1-3	15	1 (1.5%)	0	2 (3.1%)	5 (7.7%)	10 (1.5%)	8 (12.3%)	7 (10.8%)
4-5	50	9 (13.8%)	4 (6.2%)	2 (3.1%)	9 (13.8%)	41 (63%)	38 (58.5%)	12 (18.5%)
p		>0.05	>0.05	>0.05	>0.05	>0.05	>0.05	>0.05

Patients with CATIS score 1-3 reported more clinical symptoms than patients with higher CATIS score – average number of clinical symptoms was 8 and 5,4 respectively.
Conclusion: 1. Most of children with MVP have positive feelings about their illness. Boys achieve higher CATIS score than girls. 2. Child Attitude Toward Illness Scale results are not related to age, occurrence of arrhythmia and effort tolerance in treadmill test. 3. Patients complaining of higher number of clinical symptoms have worse attitude toward illness, what reflected in CATIS score.

P-47
Switching immunosuppression from CsA to Tac after pediatric heart and heart lung transplantation

Fuchs A., Diterich J., Stroetgen J., Beier G., Urschel S., Dalla-Pozza R., Kozlik-Feldmann R., Netz H.
Department of Pediatric Cardiology, University Hospital Großhadern, Munich, Germany

In a prospective study we investigated the effects of conversion from Cyclosporine A (CsA) to tacrolimus (TAC) in pediatric recipients of cardiac allografts. Side effects of CsA led to a switch in the immunosuppressive regimen.
22 heart (HTx) and 3 heart lung (HLTx) transplanted patients (pts) with stable graft function were assigned to the conversion to TAC. Indications for the switch to TAC were severe hypertrichosis in 7 pts, gingival hyperplasia in 5 pts, renal insufficiency in 5 pts, hypertonia in 3 pts, hyperlipidemia in 3 pts and atopic eczema in 2 pts. Demographic data: n = 25, 15 boys and 9 girls, weight 47.5 ± 20.9 kg, age 10 ± 7.5 years (yrs), follow-up time 23.4 ± 4.5 months. Diagnosis leading to transplantation were dilative cardiomyopathy: n = 8, restrictive cardiomyopathy: n = 3, Kawasaki Disease: n = 1, idiopathic pulmonary hypertension: n = 1, congenital heart disease: n = 12.
Renal insufficiency was evaluated in values of serum creatinine and creatinine clearance, atopic eczema was assessed by calculating the Scorad score, hypertrichosis by calculating the Ferriman-Galway-Index. TAC was introduced directly with doses adjusted to a range of 8-10 ng/ml. Concomittant immunosuppressive therapy, consisting of mycophenolate mofetil and low dose steroids in the heart-lung-transplanted patient was maintained after the switch. Hypertrichosis (Ferriman Galway index) and gingival hyperplasia had strongly resolved in all pts 3 months after the switch. Renal function improved in terms of a decreased serum creatinine 3 months after the switch. Hyperlipidemia and hypertonia improved 3-6 months after the switch. Atopic eczema disappeared in 1 pt and remained unchanged in the other one.
Conversion from CsA to TAC in pediatric heart transplanted pts is effective. CsA related side effects improved within a few months, resulting in a better quality of life and in an improved cardiovascular risk profile.

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P-48

"Non causally related" pulmonary artery hypertension in congenital heart diseases

Basquin A., Levy M., Agnoletti G., Bajolle F., Boudjemline Y., Bonnet D.
M3C-Pediatric Cardiology, Necker-Enfants Malades, Paris, France

Background: The revised clinical classification of Pulmonary Artery Hypertension (PAH) (Venice, 2003) identifies PAH associated with congenital systemic-to-pulmonary shunts and with left heart diseases. However, PAH can be observed in congenital heart disease (CHD) while elevated pulmonary vascular resistances can not be causally related to the physiology of the CHD.
Objective: To report a series of PAH associated CHD without causal relationship.
Methods and Patients: We identified from our database all children with PAH and CHD. All patients with left-to-right shunts (ASD –VSD) were excluded, as well as patients with PAH and left heart disease, particularly pulmonary vein stenosis (including post-mortem diagnosis).
Results: Eleven patients were considered to have PAH without causal relationship between PAH and CHD: five transpositions of the great arteries, two scimitar syndromes, one successfully repaired coarctation of the aorta, one VSD with severe pulmonary stenosis, one previously closed patent ductus arteriosus, and one absent left pulmonary artery. During follow-up, four patients died, three underwent a Pott's shunt (two alive) and all received targeted therapy for PAH.
Conclusion: The distribution of CHDs associated with PAH in our series has been consistently described, suggesting a specific subgroup that could be named "non causally related PAH in CHD". This very well known condition could either be a fortuitous association between CHD and PAH or a specific subtype of associated PAH that should appear in classifications.

P-49

Echocardiographic versus CMR evaluation of right ventricular size after TOF repair.

Munkhammar P. (1), Carlsson M. (2), Liuba P. (1), Sarna S. (3), Pesonen E. (1)
Children's Hospital, University Hospital, Lund, Sweden; Department of Clinical Physiology, University Hospital, Lund, Sweden; Department of Public Health, University of Helsinki, Helsinki, Finland

Background. Transannular patching of the right ventricular outflow tract during surgical repair of TOF is a common cause of significant pulmonary regurgitatation and subsequent RV volume overload. On follow-up 50% of patients develop restrictive right ventricular physiology, which reduce the growth of RV and its diastolic filling capacity. Restrictivity has been shown to be beneficial in reducing the risk for late sudden death and malignant ventricular tachycardia. Echocardiography is used for sizing RV. By tracing the internal RV surface of a two-dimensional (2-D) echocardiographic pictures RV area can be calculated. This technique excludes essential parts of the RV from measurements such as the outflow tract and the trabeculated apical portion. We compared the volume estimated from the two dimensional echocardiogaphy pictures with volume measurement from cardiac magnetic resonance (CMR) images to test the accuracy of the echocardiographic method.
Methods and Results: Thirty-two patients were examined after TOF repair with transthoracic echocardiography (TTE) and CMR. The RV area was measured with TTE from an apical four-chamber view in end diastole and systole and averaged across 3 consecutive beats. The measurements were compared with CMR-derived RV volume measurement across the same time-interval. In univariate linear regression analysis, the RV`s area assessed by TTE significantly correlated with volume measurements by CMR ($p < 0.0001$, $r^2 = 0.53$ in systole and 0.38 in diastole).
Conclusion. Transthoracic echocardiography is not an adequate method for evaluating the RV volume in individual patient. The technique is not reliable for risk evaluation of restrictivity-related complications later in life.

P-50

Vectorcardiographic recordings of QT interval predicts LQTS diagnosis in children

Diamant U.B. (1), Winbo A. (2) Rydberg A. (2) Stattin E.L. (3) Jensen S.M. (1)
Heart Centre(1), Paediatric Cardiology(2), Medical and Clinical Genetics (3) Umeå University Hospital, Umeå, Sweden

Introduction: Manual measurements of QT intervals are time consuming and burdened with inter- and intra- individual variation. QTc measurements in the paediatric population are even less reliable. We hypothesized that VCG recordings could determine the QT interval with high precision and predict whether an individual has LQTS or not.
Method: Electrocardiography (ECG) and vectorcardiography (VCG) were performed in a population consisting of 23 children, 14 LQT1, 3 LQT2 and 6 normals. Mean age was 8.0 year (1 to 15), 15 female and 8 male. The 12-lead ECG automatic measurement and interpreting of QTc where performed with a Mac®5000, GE Medical system, the VCG automatic measurement were made with Mida®1000, Ortivus. Four experienced observers performed the manual measurements of QTc in lead II, V5 and V6, this was repeated after one week. Bazett's formula was used for rate correction. The patients were classified according to Goldenberg 2006, children 1–15 years: normal < 440 ms, borderline 440–460 ms, prolonged > 460 ms, borderline values were counted as having LQTS. The measurements from the observer with the smallest intraobserver relative error were used for comparison.
Result: The sensitivity, specificity and predictive values, compared to genotyping as gold standard are shown in the Table.
Conclusion: The VCG measurement of QTc seems to be a better predictor of LQTS in children than manual or automatic measurement and interpreting on scalar ECG. This should be confirmed in larger series including other LQTS types in a paediatric population.

Table

	Sensitivity	Specificity	Positive predictive value	Negative predictive value
Observer 1	0.882	0.500	0.883	0.600
Mida®1000	1.000	0.833	0.944	1.000
Mac®5000 automatic interpreting	0.471	0.833	0.889	0.357
Mac®5000 automatic measured	0.611	0.800	0.917	0.364

P-51

Use of betablockade in patients with catecholaminergic polymorphic ventricular tachycardia

De Klerk J.C.A. (1), Clur S.A. (2), Delhaas T. (3), Blank A.C. (4), Blom N.A. (5), De Kezel C.C.A. (6), Rammeloo L. (7), Van de Heuvel F. (8), Reimer A. (9), Wilde A.A.M. (2), Ten Harkel A.D.J. (1)

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Erasmus MC-Sophia, Rotterdam, The Netherlands (1); Academic Medical Center, Amsterdam, The Netherlands (2); University Medical Center Maastricht, Maastricht, The Netherlands (3); University Medical Center Utrecht, Utrecht, The Netherlands (4); Leiden University Medical Center, Leiden, The Netherlands (5); Elisabeth Hospital, Tilburg, The Netherlands (6); VU Medical Center, Amsterdam, The Netherlands (7); University Medical Center Groningen, Groningen, The Netherlands (8); University Medical Center Nijmegen, Nijmegen, The Netherlands (9)

Introduction: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a congenital rhythm disorder. These patients can suffer from syncope or cardiac arrest. They should be treated with beta-blockade to limit these risks. However, side effects may reduce compliance In asymptomatic patients. We therefore studied the use of betablockers and its side effects in a group of young CPVT patients.
Methods: The medical files of all CPVT patients under the care of a pediatric cardiologist in seven academic hospitals and one non-academic hospital in the Netherlands were retrospectively analyzed. Information about medication and potential side effects were collected.
Results: The study population consisted of 34 CPVT patients. Mean age was 12 ± 4 (4–18) years. Age at diagnosis was 8 ± 4 (1–17) years. Molecular genetic analysis showed a RYR2 gene mutation in 26 patients (77%), and a CASQ2 gene mutation in 1 (3%) Seven patients had a history of syncope (N = 4), epileptic seizures (N = 2) or cardiac arrest (N = 1), while the other 27 patients were diagnosed after family screening. Treatment has been instituted in 23 patients (68%), all were treated with betablockers in combination with ICD (N = 2) or pacemaker (N = 1). Of the 11 patients that did not receive therapy, 4 parents did not want to start prophylactic therapy for religious reasons. Two patients changed from betablocker, while two have received an additional ICD. Therapy was changed because of persistent symptoms of syncope in 3/4 patients. Eight patients (35%) experienced side effects of their treatment with beta-blockade. These side effects include tiredness (n = 5) and cold limbs (n = 3).
Conclusion: The study population consisted of 34 CPVT patients. Most patients are being treated with betablockade, although a minority refused preventive therapy for various reasons. One third of the patients experienced side effects. These side-effects should be discussed with patients and/or their parents.

P-52

Use of betablockade in patients with long QT syndrome

De Klerk J.C.A. (1), Clur S.A. (2), Delhaas T. (3), Blank A.C. (4), Blom N.A. (5), De Kezel C.C.A. (6), Rammeloo L. (7), Van de Heuvel F. (8), Reimer A. (9), Wilde A.A.M. (2), Ten Harkel A.D.J. (1)
Erasmus MC-Sophia, Rotterdam, The Netherlands (1); Academic Medical Center, Amsterdam, The Netherlands (2); University Medical Center Maastricht, Maastricht, The Netherlands (3); University Medical Center Utrecht, Utrecht, The Netherlands (4); Leiden University Medical Center, Leiden, The Netherlands (5); Elisabeth Hospital, Tilburg, The Netherlands (6); VU Medical Center, Amsterdam, The Netherlands (7); University Medical Center Groningen, Groningen, The Netherlands (8); University Medical Center Nijmegen, Nijmegen, The Netherlands (9)

Introduction: Patients with long QT syndrome (LQTS) may suffer from ventricular arrhythmias, leading to syncope and sudden death. Although treatment with betablockers reduces these risks, side effects may limit patient compliance. We therefore studied a series of LQTS patients with respect to use of betablockers and its side effects.
Methods: The medical files of all patients with LQTS under the care of pediatric cardiologists in seven academic hospitals and one non-academic hospital in the Netherlands were retrospectively analyzed. Information about medication and potential side effects were collected.
Results: We included 153 LQTS patients with a mean age of 12 ± 5 (0.5–26) years. Age at diagnosis was 7 ± 5 (0–17) years. In 117 patients a genetic diagnosis was available (76%). LQT1 was present in 46 patients (30.1%), LQT2 in 58 patients (37.9%), LQT3 in 11 patients (7.2%), and Jervell Lange-Nielsen in 2 patients. QTc was 472 ± 34 msec (416–570) for LQT1 patients, 465 ± 40 msec (390–570) for LQT2 patients, 463 ± 36 msec (419–520) for LQT3 patients and 569 ± 27 msec (550–588) for the Jervell Lange-Nielsen patients. Treatment with betablockers was provided to 123 patients, in some patients in combination with ICD (N = 6) or pacemaker (N = 1). An ICD (N = 2) or a pacemaker (N = 5) was implanted in 7 additional patients. These seven patients all had LQT3. In 13 asymptomatic patients treatment was postponed to an older age. In 10 patients, side effects were the reason to change from betablocker; 28 additional patients changed to a once-daily regimen of the new drug. Forty five patients (34%) experienced side effects of their treatment with beta-blockade. The mostly reported side effects are tiredness (n = 12; 21%), cold limbs (n = 7; 12,1%), dizziness (n = 7; 12,1%) and headache (n = 7; 12,1%).
Conclusion: We studied a large LQTS population with 76% positive mutation analysis. Although most patients were treated according to present guidelines one third experienced side-effects of betablockade. These side-effects have to be discussed with patients and/or their parents.

P-53

Junctional ectopic tachycardia after surgery for congenital heart disease: incidence, risk factors and outcome.

Mildh L. (1), Hiippala A. (2), Rautiainen P. (1), Sairanen H. (3), Pettilä V. (4), Happonen J.-M. (2)
Department of Anaesthesia and Intensive Care (1), Paediatric Cardiology (2), Paediatric Cardiac Surgery (3), Hospital for Children and Adolescents; Division of Anaesthesia and Intensive Care (4), Helsinki University Hospital, Helsinki, Finland

Introduction: Junctional ectopic tachycardia (JET) is associated with surgery for congenital heart disease and can cause serious hemodynamic problems. An incidence of 6–11% has been previously reported. The purpose of this study was to analyze the incidence, risk factors and outcome of JET in a consecutive single centre patient cohort.
Methods: All 1026 patients under 18 years, who underwent open heart surgery for congenital heart disease in our hospital during 2000–2004 were included in this retrospective analysis. The patients with JET were identified by analyzing the ECG's of the patients who received amiodarone during postoperative intensive care. Three controls, adjusted for age and type of surgery, were selected for each patient diagnosed with JET.
Results: JET was diagnosed in 51 patients (5.0%). The median age was 0.3 yrs (interquartile range 0.03–0.91) for the JET group, and 0.54 yrs (interquartile range 0.19–2.86) for the whole patient cohort. VSD closure was part of the surgical procedure in 33/51 (64.7%) of the JET patients (VSD ± ASD closure, TOF correction, Rastelli, ASO for TGA+VSD, PA+VSD correction, AVSD correction). Other types of surgery among the JET group were ASO for simple TGA (6/51, 11.8%), TCPC (5/51, 9.8%), and other (7/51, 13.7%).
JET patients had longer cardiopulmonary by-pass time (CPB) (138 vs. 119 min, $p = 0.002$), higher body temperature at the onset of tachycardia (38.0 vs. 37.4°C, $p = 0.013$) and higher level of postoperative TnT (3.7 vs. 2.1 μg/l, $p < 0.001$) compared to controls. They

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also needed longer ventilatory support (3 vs. 2 days, p = 0.004) and ICU stay (7 vs. 5 days, p < 0.001). The mortality was 10% in the JET group and 4% in the controls (p = 0.066). There was no difference in the plasma levels of potassium, magnesium or calcium, or use of inotropic drugs. JET was not an independent risk factor for death in the whole cohort in logistic regression analysis.
Conclusions: The incidence of JET was 5.0% in this unselected paediatric open-heart surgery patient group. Compared to controls, JET patients had longer CPB time and higher level of TnT, possibly reflecting bigger surgical trauma. JET was not an independent risk factor for death.

P-54
Automatic atrial threshold measurement is reliable in pediatric pacemaker patients
Hiippala A. (1), Serwer G. (2), Clausson E. (3), Davenport L. (4), Brand T. (4), Happonen J.-M. (1)
Pediatric Cardiology, Helsinki University Hospital, Helsinki, Finland (1); University of Michigan Congenital Heart Center, University of Michigan Health System, Ann Arbor, MI, USA (2); Medtronic AB, Kista, Sweden (3); Medtronic, Inc., Mounds View, MN, USA (4)

Automatic threshold measurement and output adjustment are used as default settings in modern pacemakers. While the safety and reliability of these algorithms have been studied in adults, there are no comparable studies on automatic atrial threshold measurements in children. The purpose of this study was to assess the performance of Atrial Capture Management (ACM) present in Medtronic pacemakers in pediatric patients.
Forty pacemaker patients, 23 boys and 17 girls, were enrolled. The median age was 9.9 yrs (range 0.8–17.5 yrs). The atrial leads had been implanted for mean 2.8 yrs (range 0.1–12.1 yrs). An epicardial atrial lead was used in 45% of the patients. The pacing indication was high grade AV block in 82% of patients and sinus node disease in 18%. Operated congenital heart disease was present in 20 patients (50%); single ventricle physiology was present in six patients. The objective was to compare manual atrial threshold and ACM measurements. Clinical equivalence of the measurements was defined as ACM threshold being within −0.25 to +0.5 V of the manual threshold to allow for some circadian variation as the ACM measurements were often performed at night.
ACM measurements were within the expected variation in 38/40 (95%) of the patients. One patient had too high intrinsic heart rate for ACM to be able to measure at all. Data were not available in another patient. Successful ACM measurements within three days prior to follow up visit were present in 37/40 pts (93%). The results are presented in the table:

	Manual threshold V @ 0.4 ms	ACM threshold V @ 0.4 ms	Mean threshold difference V	95% CI V
All patients	0.69 ± 0.32	0.68 ± 0.35	0.013	-0.027 to 0.053
Epicardial leads	0.67 ± 0.27	0.66 ± 0.32	0.014	-0.066 to 0.094
Endocardial leads	0.70 ± 0.36	0.69 ± 0.38	0.012	-0.032 to 0.056

ACM measures atrial thresholds reliably in pediatric patients with both endocardial and epicardial leads allowing its use in both. Constant high intrinsic atrial rate may prevent automatic threshold measurement in young children.

P-55
Pediatric ablation in an adult electrophysiology laboratory: A safe and effective setup
Chetaille Ph. (1), Cote JM. (1), Philippon F. (2), Champagne J. (2), Gilbert M. (2), Molin F. (2), Blier L. (2), Houde C. (1), Delisle G. (1), O'Hara G. (2)
Centre Mère-Enfant du Centre Hospitalier Universitaire de Québec, Québec, Canada (1); Québec city Heart Institute, Hôpital Laval, Québec, Canada (2)

Introduction: Catheter ablation in children can be complex due to the patient size and cardiac anatomy. Pediatric cardiology centers may not offer adequate or up to date equipment and trained personnel required for such procedures due to the small number of patients involved.
Objective: The purpose of this study was to evaluate our 14-year experience of pediatric ablations in the adult setup of the Quebec Heart Institute.
Method: We retrospectively reviewed the database of the EP lab for all the pediatric electrophysiology procedures done to evaluate indications, success/failure rates, complications and long term evolution.
Results: From September 1993 to December 2007, 420 invasive procedures were performed in pts aged 9 months-17 yrs, mean 13 yrs. Age was ≤10 yrs in 19%. Most procedures were performed by a pediatric electrophysiologist assisted in most cases by one of six adult electrophysiologists.
Indications for EPS were SVT in 92%. The EPS findings were: WPW/concealed pathway 178(42%), AVNRT 119(28%), PJRT 19(4%), EAT 16(4%), Flutter 11(3%), VT 7(2%), Mahaim 3, JET 2, EPS without ablation 65(15%). Ablation was attempted in 355 with overall success of 311/344 (90%), failure rate was 37/344 (10%), and no ablation attempted in 11 (3%). 155/178 (87%) WPW/CP were successfully ablated, there were 15 failures. Ablation was not attempted in 8 (7/8 antero-septal close to the AV node). AVNRT was diagnosed in 119, 118 (99%) were successfully ablated with no AV block. There were 2 late recurrences which were successfully ablated in a second procedure. 19 procedures were performed in 13 patients with PJRT. At follow-up, they are all cured. There were 16 EAT procedures in 15 patients. At follow-up 13/15 were free of arrhythmias and free of anti-arrhythmic drugs. There were 11 flutter ablations in 8 pts, including 3 Fontan. In the whole series, there were no serious complications including death, heart block, tamponade, or stroke.
Conclusion: The combination of pediatric and adult electrophysiologists in a high volume adult EP lab that offers up to date equipment and trained technical personnel proved to be a very safe and successful set up for arrhythmia management of our pediatric patients.

P-56
Use of automatic ventricular threshold measurement and adjustment (AutoCapture™) in infants
Hiippala A., Happonen J.-M.
Pediatric Cardiology, Helsinki University Hospital for Children and Adolescents, Helsinki, Finland

Introduction: AutoCapture (AC) in St. Jude pacemakers offers beat-to-beat capture verification enabling pacing safety marginal of only 0.3 V. The evoked response signal (ERS) amplitude needs to be sufficient for this generator saving function. In a previous study ERS decline over time has been reported to happen in 20% of children with transvenous leads, while another study shows no difference during 4-year follow-up with epicardial leads. The purpose of this study was to evaluate the performance of AC in infants with epicardial leads.
Methods: Between Jan 1998 and Dec 2004, 16 newborns with normal cardiac anatomy were implanted with a Microny® pacemaker through subxiphoidal sternotomy. Medtronic bipolar epicardial lead 4968 was used in 88% and 10366 in 12%. All patients had congenital complete AV block due to maternal autoimmune disease (Sjögren 69%, LED 25%, and MCTD 6%) and needed 100% pacing. Follow-up time was median 4.0 yrs (range 0.4–8.6 yrs); end-point for follow-up was generator or lead exchange, or death.. The ER signals at discharge, at 12 ± 2 months and long term AC performance were recorded retrospectively.

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Results: In this cohort of 16 newborns median birth weight was 2570 g (range 1750–3250 g), gestational age 37 weeks and heart rate 50 bpm. The median threshold was 0.8 V (range 0.3–2.3 V), R-wave 17.4 mV and lead impedance 1180 ohm at pacemaker implantation. AC could be activated in all newborns with mean ERS of 10.5 ± 4.4mV at discharge.
During the first year of follow-up, ERS declined with mean 49% of the original amplitude and measured mean 4.6 mV at 12 ± 2 months (N = 13). It fell below 2.5 mV in 3 pts (19%) eliminating the possibility to use AC. The ERS and polarization safety margins became too low in additional 3 pts (19%) resulting in AC deactivation. Long term AC use was possible in 9 patients (56%). One patient died at the age of 11 months despite good LV-function and pacing parameters.
Conclusion: In infants, the ERS should be evaluated during every pacemaker follow-up because significant decline may occur. Long term low output pacing from birth was possible in 56 % of patients.

P-57

Transseptal Procedures [TSP] in Pediatric EP and Cardiology: Long Term Follow Up and Risk Analysis in more than 1100 Patient-Years

Will J.C., Botsch M., Krings G., Berger F.
Pediatric Cardiology, Charite University Hospital Berlin, Germany

Introduction: After antegrade dilatations of congenital AS and CoA has not been shown to be the primarily used method there is an increasing use of TSP due to electrophysiologic ablation procedures (left sided accessory pathways, IARTs, atrial fibrillation). Purpose of the study: Incidence of TSP related complications in pediatric cardiology/EP, identification of potential risk factors.
Methods: Retrospective analysis of all TSP since 1994.
Results: 5020 invasive catheterisation procedures in pediatric and congenital patients [pts]. 169 TSP were performed (3.4%) in 144 pts, 91 were male. Mean pts age was 11.5 yrs(range 0.1–63 yrs), mean weight 44.5 kg (range 3–113), mean length 142 cm (range 48–189). In 97 pts (57%) CHD was present. 35% of the TSP were performed for ablation procedures, 22% for ballon/stent interventions, 17% during diagnostic caths and 12% for PDA occlusions. 7% of the TSP were through baffles/conduits or ASD patches. During a cumulative follow up of 1101.2 patient-years (mean 7.24±5.04 years) 3 minor complications were observed (1.8%): 2 residual shunts (1 after puncture through a surgical baffle, 1 spontaneous closure after 1 month, 1 pericardial effusion without need for further intervention).
Conclusion: TSP in pediatric catheterization and EP diagnosis and therapy is safe and unrelated to patient age, diagnosis and weight.

P-58

Sensitivity and Specificity of QT-RR Relationship on Holter in LQT1, LQT2 versus Gene-negative Patients Being Assessed for Long QT Syndrome (LQTS)

Hamilton R.M., Fenwick L.M., Gross G.J.
The Hospital for Sick Children, Toronto, Canada

Introduction: Genetic LQTS diagnosis allows for specific therapy, risk stratification, and family expansion but is costly and not routinely funded. LQTS patients (pts) have abnormal QT/RR relationships on Holter recordings. We assessed the sensitivity (SENS) and specificity (SPEC) of QT/RR for gene-proven LQT1 and LQT2 versus gene-negative pts being evaluated for LQTS.
Methods: Pts (0 to 50 yr, median = 12 yr) assessed for LQTS between 2005 and 2007 underwent genetic testing (PGX Health) and Holter analysis using GE Medical automated QT/RR analysis. Pts were classified as LQT1 or LQT2 if a class 1 or 2 mutation was present, and gene-negative if no class 1 or 2 mutation of LQT1, 2 or 3 was identified. QRS to T-wave peak (QTp) and T-wave offset (QTo) were measured during waking (W), sleep (S), and total (T) hours of recording in leads V5 and V2, and QT/RR slope measured. LQT1 and LQT2 were compared to gene-negative by unpaired T-test, and SENS and SPEC were determined.
Results: The table shows results for 18 LQT1 Holters (15 pts) and 8 LQT2 Holters (6 pts) versus 10 gene -ve Holters (9 pts). QT/RR measurements were possible in V5 in 36/36 and in V2 in 29/36 Holters. LQT1 pts were best identified by QTp during S in V5 (SENS 83% and SPEC 80% at cutpoint slope = .14), and LQT2 by QTp during W in V5 (SENS 87.5% and SPEC 70% at cutpoint slope = .16) Conclusions: Both LQT1 and LQT2 pts can be identified by QT/RR relationships on Holter.
QT/RR Results by Gene and State (*p < .05, **p < .01)

		QTo			QTp		
Lead	State	LQT1	LQT2	Gene -ve	LQT1	LQT2	Gene -ve
V5	W	.226 ± .069**	.276 ± .095**	.157 ± .047	.205 ± .058**	.271 ± .093**	.141 ± .051
	S	.200 ± .065**	.177 ± .094	.132 ± .040	.177 ± .050**	.154 ± .116	.120 ± .035
	T	.234 ± .043*	.287 ± .098*	.191 ± .053	.213 ± .037*	.276 ± .104*	.178 ± .052
V2	W	.201 ± .087	.300 ± .111*	.171 ± .041	.159 ± .046	.287 ± .086**	.152 ± .058
	S	.177 ± .111	.227 ± .088*	.140 ± .039	.136 ± .075	.201 ± .135	.104 ± .022
	T	.210 ± .070	.330 ± .077**	.197 ± .070	.173 ± .049	.320 ± .063**	.185 ± .074

P-59

Intraoperative ablation of ventricular tachycardia associated with right ventricular remodelling during valve implantation for pulmonary regurgitation and aneurysmal RVOT dilatation: a multidisciplinary approach to GUCH patients.

Tavera M.C. (1), Foresti S. (1), Lupo P.P. (1), Ali H. (1), Pittalis M. (1), De Ambroggi G. (1), Chessa M. (2), Giamberti A. (3), Negura D. (2), Cappato R. (1)
Electrophysiology and Cardiac pacing Department (1), Pediatric Cardiology, GUCH Unit (2), Cardiac Surgery, GUCH Unit, IRCCS Policlinico San Donato, San Donato Milanese, Italy

Introduction: arrhythmia is the main cause of morbidity and mortality in grown up congenital heart disease (GUCH) patients. Pulmonary regurgitation with right ventricular (RV) dilatation is a possible complication late after correction of congenital RVOT obstruction. In patients with this condition, sudden death may occur due to ventricular tachyarrhythmias (VT).
Methods: Adult patients (>16 years) with pulmonary regurgitation and RV dilatation requiring cardiac surgery comprehensive of RV remodelling were studied. Complete clinical and instrumental evaluation included baseline Electrophysiological Study (EPS) with an aggressive protocol for induction of VT. When VT was induced an electroanatomical map of the RV was performed. Sites of conduction slowing were identified as potential isthmi of reentrant circuits. During surgery, ablation was performed using an irrigated unipolar radiofrequency device to obtain a transmural lesion at the critical isthmi. In all patients, EPS was repeated 6 months after surgery.
Results: 26 patients were studied from October 2005 to December 2007. 4 of them presented clinical VT. In 6/26 patients VT was reproducibly inducible (3/6 experiencing clinical VT). In these 6 patients high density electroanatomical map was performed. During surgery, ablation was carried out at sites identified by prior mapping. 1 patient, with clinical pulseless VT, not inducible at baseline EPS, underwent ICD implantation after surgery. Six months follow up data are available for 16/26 patients. None of them presented clinical VT. VT at 6 months FU has been induced in 1/12 patients with negative baseline EPS and 1/4 patients with positive baseline EPS undergone surgical ablation. In the first patient inducibility was

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suppressed by drug therapy, in the second, VT induced resembles the same circuit of pre-operative VT, but with longer cycle lenght and haemodinamic stability, so trans-catheter ablation has been planned. One-year follow up has been completed for 14/26 patients, none of them experienced clinical VT.
Conclusion: intraoperative ablation guided by electroanatomical mapping appears to be effective in patients with congenital heart disease presenting clinical VTs. Multidisciplinary approach is feasible and seems to provide a good therapeutical option in GUCH patients at hight risk for VT or sudden death.

P-60
Safety and efficacy of paediatric outpatient radiofrequency catheter ablations
Tomaske M. (1), Candinas R. (1), Weiss M. (2), Bauersfeld U. (1)
Division of Paediatric Cardiology (1), Department of Anaesthesia (2), University Children's Hospital Zurich, Switzerland

Introduction: Radiofrequency catheter ablations (RFA) are frequently performed as treatment for supraventricular tachycardia in children older than 4 years of age. Aim of this study was to evaluate safety and efficacy of paediatric outpatient RFA.
Patients and Methods: Between 06/2002 and 03/2007, 201 RFA were prospectively analyzed. Exclusion criteria for outpatient procedures were complex RFA in congenital heart disease, arterial access or distance to home more than 1 hour. All RFA were performed under general anaesthesia. In case of transseptal puncture, patients received a single-shot dose heparin. All patients underwent postprocedural echocardiography and electrocardiogram and were discharged within 6 hours after conclusion of RFA. To identify potential complications after discharge, parental follow-up phone calls the day after outpatient RFA procedure were performed.
Results: A total of 65/201 (32%) patients aged 13.6 ± 3.8 years qualified for outpatient RFA. Accessory pathway ablations (n = 33) and atrioventricular node modifications (n = 28) were the most common RFA. A transseptal approach was performed in 24 RFA. Median procedure time was 1.5 hours (range: 1.1–4.3), with a median fluoroscopy time of 10 minutes (range: 5–86). RFA was successful in 63/65 (97%) patients. Postprocedural echocardiography with special attention for intracardiac thrombi, pleural effusion and inflow patterns from systemic veins or the coronary sinus were normal in all patients. Anaesthetic adverse events, predominantly post-interventional nausea and vomiting, were observed in 9 (10%) patients. Hospital discharge within 6 hours after conclusion of RFA was practicable in all but one patient due to ongoing nausea. Follow-up phone calls did not reveal further complications. Recurrence of tachycardia after RFA was observed in 4 of 65 (6%) patients.
Conclusion: Outpatient RFA are feasible and safe in selected paediatric patients. No RFA related complication was observed. Anaesthetic adverse events were nausea and vomiting due to general anaesthesia. Success rate and recurrence rate of tachycardia was favourable after outpatient RFA.

P-61
Arrhythmia's in Patients with Ebstein Anomaly and RF Ablation
Szumowski L. (1), Orczykowski M. (1), Szufladowicz E. (1), Derejko P. (1), Urbanek P. (1), Kepski R. (1), Bodalski R. (1). Włodarska K. (1), Knecht M. (2), Bieganowska K. (2), Kawalec W. (2), Walczak F. (1)
Institute of Cardiology, Warsaw-Anin, Poland (1); The Childrens Memorial Health Institute, Poland (2)

Introduction: In patients (pts) with Ebstein anomaly exists a complex electro-anatomical substrate for recurrent arrhythmia – atrioventricular reentry tachycardia (AVRT), atrial fibrillation (AF) and life-threatening ventricular tachyarrhythmia.
The aim of this study is assessment of the presence and type of arrhythmia in pts with Ebstein anomaly who underwent RF ablation.
Methods: 930 with WPW syndrome, mean age 34.8 years, underwent non-pharmacologcal treatment (years 1988-2007).
18 of them (2%), 12 female (F), (mean age 26 yrs) and 6 male (M) (mean age 35 yrs) had Ebstein anomaly.
The results of 12 lead ECG Holter, ECG monitoring, electrophysiological tests and an echocardiography study were analyzed.
Results: 18 pts had have 26 accessory pathways (AP), in 4 pts (27%) it was "wide" AP.
7 pts (40%) had have 2 AP, 1 (5%)-3 APs. In 17 pts (95%) right-sided AP, in 1 pts (5%) left-sided AP was observed. 14 pts (80%) had have overt WPW syndrome, 2 pts (10%) intermittent, 2 (10%) concealed.
15 pts (84%) had ortodromic AVRT, 3 (16%) antydromic. 2 pts had AF, 2 AT, 2 AVNRT, 1 AFL. 2 pts (13%) had have episodes of AVRT, AF and AT.
2 (13%) had AVRT and AVNRT. 1 pts with primary VF had have 3 APs and had intermittent long QT.
Conclusions: 1. In analyzed pts with Ebstein anomaly multiple APs occurred frequently or APs were wide.
2. In more than 50% of pts with Ebstein anomaly existed more than one type of arrhythmia.

P-62
Dilative Cardiomyopathy due to an eccentric electrical activation of the ventricles by an accessory pathway
Emmel M., Sreeram N. Brockmeier K.
Cologne Heart Center, Pediatric Cardiology, University of Cologne, Germany

Introduction: An accessory pathway may cause an eccentric electrical activation of the myocardium which can be seen as a delta-wave in the surface ECG. On the other hand it is well known, that patients under long term pacemaker therapy (with pseudo-LBBB) may develop a "pacemaker cardiomyopathy") due to an eccentric electrical activation of the ventricles.
Patients: We report on eight Patients (age 12-156 months) with dilative cardiomyopathy. In all patients the ECG showed a pre-excitation fitting to an accessory para-Hissian pathway, but none of these patients had had a clinical or documented tachycardia.
Results: In two of the patients the cardiomyopathy resolved spontaneously when the pre-excitation had disappeared in the surface ECG. In six patients we ablated the accessory pathway. During the EP-study there was no tachycardia inducible in any patient with usual stimulation technique. Five patients had a right-sided para-Hissian pathway and in one patients we ablated a fasciculo-ventricular pathway. Myocardial specimen taken in three patients during the EP-study revealed no pathology. Left-ventricular dimensions and function turned back to normal within three weeks after ablation
Conclusion: The excentric electrical activation of the ventricles secondary to an accessory pathway (electrical remodelling) may cause a mechanical remodelling with development of a dilative cardiomyopathy. When the pre-excitation persists the ablation of the pathway is a causal therapy allowing reverse remodelling of the ventricular myocardium.

P-63
Radiofrequency catheter ablation of ectopic supraventricular tachycardia in children
Bieganowska K. (1), Szumowski Ł. (2), Miszczak-Knecht M. (1), Rękawek J. (1), Szymaniak E. (1), Brzezinska-Paszke M. (1), Kaszuba A. (1), Kawalec W. (1), Walczak F. (2)

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Department of Cardiology, The Children's Memorial Health Institute (1), Cardiac Arrhythmias Department, Institute of Cardiology (2), Warsaw, Poland

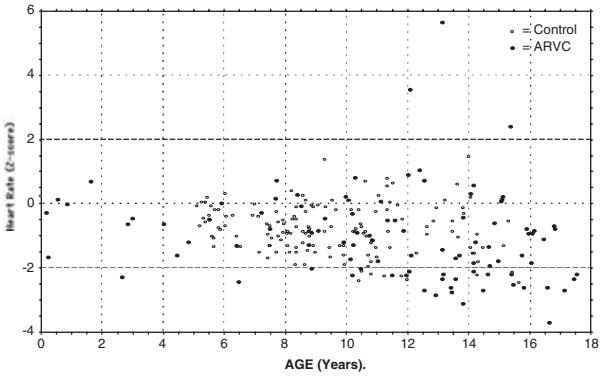
Introduction: Ectopic supraventricular tachycardia (ESVT) in children is rare and usually resistant to pharmacological therapy. Radiofrequency (RF) catheter ablation has became the procedure of choice for patients with supraventricular tachycardia.
Methods: Among the 195 children with SVT treated by radiofrequency (RF) catheter ablation we had 10 children (aged 8 to 18 years, 5 girls) with ESVT. All children have had incessant form or attacks of ESVT with heart rate from 110 to 220 beats per minute, all were on permanent pharmacotherapy (beta-blockers, sotalol, propafenone). Six children had normal heart, in 3 mitral valve prolapse was diagnosed, one had tachycardia-induced cardiomyopathy. The RF catheter ablation followed by electrophysiology testing was performed under general (7 children) or local (3 children) anesthesia.
Results: In 9 patients right atrial ectopic tachycardia (located in: ostium of coronary sinus – 3, crista terminalis – 2, lateral wall – 1, anterial wall – 1, septum – 1, ostium of inferior vena cava –1) and in one junctional ectopic tachycardia was diagnosed. In all children RF ablation of automatic focus was successfully performed. During the procedure in 3 patients atrial flbrillation was induced (electrical cardioversion was performed in 2 of them). Follow-up period ranged from 1.3 to 2.75 years (mean 2 year). In one girl after the RF ablation first and second atrio-ventricular block occurred, in one adolescent the tachycardia attacks returned. All patients remained in sinus rhythm without the need for medication. The heart of the girl with tachycardia-induced cardiomyopathy had normalized.
Conclusion: In children with ESVT radiofrequency catheter ablation is effective procedure and it is possible to avoid serious complications.

P-64
Regular Holter ECG monitoring for follow up of pacemaker patients – is it necessary?
Nagel B.H.P. (1), Gamillscheg A. (1), Anelli-Monti M. (2), Beitzke A. (1)
Paediatric Cardiology (1), Cardiac Surgery (2), Medical University, Graz, Austria

Background: Whether Holter ECG's (HECG) can contribute to optimal pacemaker (PM) settings, is not known.
Patients and Methods: 95 PM follow ups were performed in 52 patients (16.3 (1.9–29) years) in 2006. 38/52 asymptomatic patients had a HECG. Four patients with a DDD PM and normal testing showed abnormal features in their HECG's. Patient 1 (14.8 years, male): congenital AV block, own heart rate < 30/min. HECG demonstrates 6 pauses (2-2.5 s). These persist despite reducing the sensitivity in the ventricle. Surgical revision shows a loosened connection between the ventricular lead and the PM. Following replacement of the PM aggregate HECG is normal. Patient 2 (19.8 years, male): complete AV block post VSD closure. HECG shows 10 pauses (2-2.2 s) with muscle artefacts. Repeat testing demonstrates typical ventricular oversensing caused by muscular provocation. With reduction of the ventricular sensitivity from 2 to 2.8 mV no more oversensing is seen. Patient 3 (19 years, female): complete AV block post AVSD repair. Testing shows 97% atrial and ventricular pacing. HECG reveals continuous fusion beats of sensed and paced ventricular beats due to intermittent own AV conduction. Maximum sense and pace AV intervals are prolonged, the proportion of atrial and ventricular pacing can thus be reduced to 75%. Patient 4 (19.8 years, male): TGA post Senning repair with sinus node dysfunction. HECG shows fusion beats of sensed and paced ventricular beats and frequent ventricular pacing in the T wave. Repeat testing reveals atrial far field sensing, therefore atrial sensitivity is reduced and maximum pace and sense AV intervals are prolonged. HECG shows no more fusion beats, no pacing in the T wave and preferred atrial pacing with own conduction raises from 34 to 84%.
Discussion: In 10% of asymptomatic PM patients HECG revealed ineffective pacing, inadequate pauses, unfavourable high stimulation rates and far field sensing. In all cases the PM setting could be improved, in one case surgical revision of the PM was indicated. HECGs should be performed in all PM patients on a regular basis.

P-65
Sinus Bradycardia is a Frequent Presenting Finding in Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)
Hamilton R.M., Huh J., De Souza L.
The Hospital for Sick Children, Toronto

Introduction: ARVC is associated with frequent ventricular tachycardias (VT) and occasional atrial tachyarrhythmias, but sinus bradycardia has not been described.
Methods: We sought to identify whether an increased proportion of ARVC patients have resting sinus bradycardia. Patients with LBBB morphology VT or a family history of ARVC were assessed with ECG, signal-averaged ECG, Holter, echocardiogram and MRI. Those with additional findings also underwent RV angiogram and biopsy. Patients were classified as ARVC or possible ARVC, or excluded if another diagnosis was identified. Control individuals were assessed from a school population. A Z-score of ECG sinus rate was calculated for both patients and controls based on the published normal values of Davignon and colleagues (1980), and compared for the proportion falling short of -2.0 Z by Chi-square analysis.
Results: The figure demonstrates the sinus rate of patients (filled circles) compared to 149 controls. Sinus bradycardia (heart rate Z-score < -2.0) was present in 27/97 (28%) patients compared to 9/149 (6%) controls ($p < 0.0001$). Assesing only patients meeting Task Force criteria, bradycardia was present in 6/29 (21%) compared to controls ($p < 0.01$).
Conclusions: Sinus bradycardia is a frequent finding in patients referred for assessment for ARVC, being present in 21–30%. Whether this represents a physiology of the disorder or a predisposition of patients to both sinus bradycardia and early ARVC presentation (such as athletic individuals) remains to be determined.



P-66
Safety and Efficacy of Transvenous Lead Implantation After Catheter Closure of Intracardiac Septal Defects
Snyder C.S. (1), Cannon B. (2), Dortan M. (1), Lucas V. (1), Braynt R. (3)
Ochnser Clinic Foundation, New Orleans USA (1); Texas Children's Hospital, Houston USA (2); University of Florida, Jacksonville USA (3)

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Background: Transvenous lead implantation in patients with a septal defect may result in complications such as clot formation and lead misplacement. Due to this, patients undergo epicardial implantation or anti-coagulation therapy. The purpose of this study is to evaluate the outcomes of device closure as well as and the transvenous lead placement and follow up.
Methods: An approved retrospective review of patients referred for transvenous lead implantation that had a transvenous septal occlusion device implanted. Data collected included: demographics, cardiac diagnosis, indication for septal closure, follow-up of closure device, indication for trans-venous lead implantation, and follow up of lead thresholds.
Results: We identified 8 patients who underwent transvenous septal occlusion followed by implantation of a transvenous lead(s). Cardiac diagnoses included complex anatomy in 5 (all previous surgical repairs) and 3 with an atrial septal defects or patent foramen ovale. 9 transvenous septal occluders (8 atrial, 1 ventricular) were placed with one patient receiving both an atrial and ventricular device. Median age at time of occlusion was 13 years (range 9-18.) No complications were noted with follow-up [average 46 months (range 5-90).] Transvenous leads were implanted during the same hospital stay (n=5), 3 months later (n=2) or 8 months later (n=2.). Indication for device implantation [pacer (n=5) and ICD (n=3)] was complete atrio-ventricular block (3), sinus node dysfunction (2) and aborted sudden death (3). No complications occurred during lead implantation at follow-up [average 44 months (range 5-84)]. Lead thresholds were excellent with no significant difference during follow-up (average: Atrium; implant=0.5 V at 0.4 ms, follow up=0.6V at 0.4 ms,Ventricle; implant=0.6V at 0.5 ms, follow-up 0.9V at 0.5 ms.)
Conclusion: Implantation of transvenous pacing and ICD leads can be successfully performed after implantation of transvenous septal occlusion devices. Transvenous leads can be implanted during the same hospital stay as their septal occlusion is performed. Acute and chronic lead thresholds are not affected by septal occlusion device placement in short-term follow-up. A large study with long term follow up is warranted to evaluate lead and device longevity.

P-67
The Diagnostic Yield from Implantable Loop Recorders in Children and Young Adults
Gass M. (2), Sreeram N. (1), Emmel M. (1), Brockmeier M. (1), Bennink G. (1), Apitz C. (2), Ziemer G. (2), Hofbeck M. (2)
University Hospital of Cologne, Germany (1); University Hospital of Tuebingen, Germany (2)

Background: Syncope and palpitations occur frequently in young patients. Noninvasive diagnostic testing may be inconclusive.
Objectives: To assess the diagnostic yield of implantable loop recorders in young patients.
Patients and Methods: Thirty three young patients underwent implantation of a loop recorder for long-term monitoring of cardiac rhythm, to establish symptom-rhythm correlation. They belonged to one of three subgroups: those with structurally normal heart, normal electrocardiogram at rest, and negative family history (n=16); patients with structural heart disease and previous surgical repair (n=11), and patients with proven or suspected primary electrical disease (n=6). A combination of automatic and patient-activated recordings was used to monitor cardiac rhythm during symptomatic episodes.
Results: There were no procedural complications. Diagnostic electrograms could be obtained in all patients. A high degree of symptom-rhythm correlation was established. In 8/33 patients, no recurrence of symptoms was observed either until end of battery life of the device (n=6) or until last follow-up (n=2). Specific cardiac therapy was required, based on rhythms recorded by the device in 17 patients (until last follow-up). This consisted of catheter ablation of a tachyarrhythmia (n=7), pacemaker implantation or upgrade (n=5) or ICD implantation (n=5). In the remaining patients (n=8), recurrence of symptoms was associated with a normal electrocardiogram, and in two of these patients a non-cardiac diagnosis was made.
Conclusion: In selected patients, the implantable loop recorder provides valuable diagnostic information to guide further therapy.

P-68
Transcathetrer ablation of supraventricular tachycardia in patients with complex congenital heart disease
Foresti Sara (1), Tavera M.C. (1), Lupo P.P. (1), Ali H.(1), Pittalis M. (1), De Ambroggi G. (1), Chessa M. (2), Giamberti A. (3), Negura D. (2) and Cappato R. (1)
(1) Electrophysiology and Cardiac Pacing Departement (2) Pediatric Cardiology, GUCH Unit (3) Cardiac Surgery, GUCH Unit, IRCCS Policlinico San Donato. San Donato Milanese. Italy.

Background: Arrhythmia is the main cause of morbidity and mortality in grown up congenital heart disease patients (pts). Atrial arrhythmia are often refractory to antiarrhythmic drug and may be haemodinamically poorly tolerated.
Methods: Since October 2006 to December 2007 we performed transcatheter ablation for supraventricular arrhythmias in 5 adult patients with complex congenital heart disease. Underlying disease was single ventricle (Fontan circulation) in one pt, complete AV channel in one pt and Tetralogy of Fallot in three pts. Most of the patients underwent multiple surgery (median 2 interventions, range 1-3). In all patients tachycardia was refractory to antiarrhythmic therapy (1-3 drugs). Three pts underwent previous attempt of transcatheter ablation and in two patients intraoperative ablation was performed during previous surgery. At the time of Electrophysiological study (EPS) 4 patients were in arrhythmia and 1 in SR. The presumptive EKG diagnosis of the arrhythmias was atrial tachycardia in all five pts. During EPS clinical arrhythmia was identified as macroreentrant right atrial tachycardia in three patients, typical atrial flutter in one pt and atrio-ventricular nodal reentrant tachycardia (AVNRT) with 2:1 conduction block mimicking atrial tachycardia in one pt .
In 3 patients with macroreentrant tachycardia 3D high density elettroanatomical mapping was performed to identify the critical isthmus for the arrhythmia circuit; RF delivery at this site resulted in arrhythmia termination. In the patient with atrial flutter RF was applied at the isthmus between the inferior vena cava and the tricuspid annulus resulting in tachycardia termination and bi-directional conduction block at that level. In the patient with AVNRT the slow pathway was ablated delivering RF at postero-septal region of the tricuspid annulus.
At a mean follow up of 10 months (range 1–17months) all patients are in regular SR (1/5 pt taking antiarrhythmic drug for symptomatic atrial premature beats).
Conclusion: Transcatheter ablation of supraventricular tachycardia is feasible and effective in selected patients with complex congenital heart disease. EKG diagnosis of arrhythmia can be misleading in these patient and diagnostic manoeuvres during EPS are mandatory to obtain successful transcatheter ablation.

P-69
Pulmonary valve insertion in an animal model of tetralogy of Fallot: refinement in device use
Basquin A., Pineau E, Roussin R., Mollet A., Agnoletti G., Bonnet D., Sidi D., Boudjemline Y.
Necker For Sick Children Hospital, Paris, France

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Background: Following our experience in valvulation of large native RVOT in animals through transcatheter technique. We conducted a study to refine the use of a modified pulmonary artery reducer in an animal model of tetralogy of Fallot.
Methods: Twelve sheep were included in the study and divided into two groups. All animals had a pulmonary trunk enlargement using a pericardial patch and a creation of a pulmonary insufficiency. Two months after this surgery, we intended to implant a device to percutaneously reduce the diameter of the PA followed by the insertion of a laboratory made Melody valved stent. Animals were sacrificed after a follow-up of 2 months. Animals from group 1 (normal size group) received a device that as an external diameter 0 to 5-mm greater than the measured PA diameter where animals from group 2 (oversized group) received a device with an external diameter greater than 5-mm. Anchoring properties and paraprosthetic leak were studied at baseline, one and two month follow-up.
Results: Prior to valve insertion, all animals had significant PR and RV dilatation on angiography. All reducers were inserted successfully through the right jugular vein. A valved stent could be placed in all animals but one. In one animal from group 1, the reducer embolized during advancement of the delivery system containing the valved stent. At one and two month evaluation, no significant paraprothetic leak was noticed in any animal from group 2 where 4 out of 6 animals from group 1 had significant leak. All animals were sacrificed according to the protocol. At autopsy, reducers were fixed to the pulmonary wall and completely covered by a fibrous tissue in animals from group 2. In group 1, devices were incompletely covered especially anteriolry completely explaining the leak observed in these animals.
Conclusion: Implantation of a pulmonary valve is possible in sheep through a transcatheter approach when the RVOT is aneurismal and exceeds 22-mm in diameter using a reducer. This reducer should be oversized by at least 5-mm to avoid embolization and paraprosthetic leak.

P-70
Embryonic Development of the Atrio-Ventricular Node from Triple AV-Nodal Primordia in Relation to Arrhythmia Etiology

Kolditz D.P. (1,3), Pijnappels D.A. (1), Blom N.A. (2), Hahurij N.D. (2,3), Gittenberger-de Groot A.C. (3), Schalij M.J. (1).
Institute A (1): Department of Cardiology, Leiden University Medical Center, Leiden, The Netherlands; Institute B (2): Department of Pediatric Cardiology, Leiden University Medical Center, Leiden, The Netherlands; Intitute C (3): Department of Anatomy and Embryology, Leiden University Medical Center, Leiden, The Netherlands

Introduction: Atrioventricular Nodal Reentrant Tachycardia (AVNRT) accounts for approximately 13–16% of supraventricular tachycardias in children and adolescents. While dual atrioventricular nodal physiology is present in approximately 62% of these cases, the developmental origin of these anatomical substrates still remains a subject of debate. We hypothesized that the AVN has a triple origin in the left sinus horn (LSH) myocardium, the AV-junctional myocardium and the specialized ring tissues.
Methods: Isolated embryonic hearts of the Japanese quail (HH 22-35, n = 20) were stained with Periodic Acid Schiff (PAS), anti-MLC2a, anti-Nkx2.5 and anti-connexin43. The AVN was analyzed in serial sections and in 3D AMIRA computer reconstructions.
Results: In the looped heart, the myocardium was identified by MLC2a, Cx43 and Nkx2.5 positive staining. At HH 22, a MLC2a and Cx43 positive and Nkx2.5 negative region could be distinguished surrounding the left common cardinal vein (LCV) in the early pacemaking region. Around HH 24, an identical Nkx2.5 negative structure was found surrounding the right common cardinal vein (sinoatrial node region). Due to the outgrowth of the right atrium and rightward migration of the LCV to eventually transform into the coronary sinus (CS) (HH 29-31), the myocardial tissue surrounding the LCV is ultimately positioned close to the CS (adult AVN region). At the same time, formation of the isolating annulus fibrosis is initiated by fusion of the epicardial AV-sulcus and endocardial AV-cushions with incorporation of the Cx43-negative AV-junctional myocardium in the lower atrium. With ventricular septation, a small part of the posterior AV-junctional myocardium, carrying along the converged specialized sinoatrial and atrioventricular ring, becomes continuous with the interventricular septum and is incorporated within the upper part of the central fibrous body where the adult AVN resides.
Conclusions: The tissues of the LSH myocardium, AV-junctional myocardium and embryonic specialized ring tissues all seem to contribute to the AVN anlage. Based on the spatiotemporal relation of these primordia, we furthermore postulate that the LSH myocardium gives rise to the medioanterior AVN (fast pathway region), the AV-junctional myocardium to the medioposterior AVN (slow pathway region) and the remnants of the specialized ring tissues to the compact part of the AVN.

P-71
Time-related changes in biventricular systolic and diastolic function after chronic pulmonary artery banding in rats

Faber M.J. (1), Dalinghaus M. (1), Lankhuizen I.M. (1), Steendijk P. (2), Duncker D.J. (3), Lamers J.M.J. (4), Helbing W.A. (1)
Erasmus MC-Sophia Children's Hospital, Dept. of Pediatric Cardiology, Rotterdam, The Netherlands (1); Leiden University Medical Center, Dept. of Cardiology, Leiden, The Netherlands (2); Erasmus-MC, Dept. of Experimental Cardiology, Rotterdam, The Netherlands (3); Erasmus-MC, Dept. of Biochemistry, Rotterdam, The Netherlands (4)

Introduction: Residual RV pressure overload in (corrected) congenital heart disease will eventually lead to RV failure, but determining the appropriate timing of (re)intervention is difficult. Therefore, we sought to determine hemodynamic signs of imminent RV failure in a model of pulmonary artery banding (PAB).
Methods: Rats received sham (n = 28) or PAB (n = 27) surgery and were housed for 6, 12 and 20 weeks respectively. After the housing period, biventricular hemodynamics were measured at baseline and during dobutamine infusions using pressure-volume loops. Systolic function was characterized by ESPVR, PRSW, and dPdt/max-Ved relations. Diastolic function was characterized by Tau, dPdt/min (early), and chamber stiffness constant (late). In addition, molecular determinants of β-adrenergic signaling and Ca2+-cycling and the degree of fibrosis were measured.
Results: RV pressure was increased to 60% of systemic level in PAB, RV mass was 2-fold increased ($p < 0.01$). There were no signs of heart failure. In PAB rats RV systolic function was 3-fold increased in all three time groups ($p < 0.01$) and dobutamine response was unimpaired. RV diastolic function parameters indicated both impaired early ($p < 0.01$) and late ($p < 0.01$) diastolic function, with similar response to dobutamine in PAB and sham. LV systolic and diastolic function were unaffected, except for a somewhat impaired early diastolic function after 20 weeks. Even though RV and LV function parameters tended to be remarkably stable throughout time, alterations at molecular level in PAB rats were observed. The β-ARs were decreased at 20 weeks (−32%, $p < 0.05$), while GRK2 was decreased both at 6 and 20 weeks (−27% and −48%, $p < 0.05$). Furthermore, at 6 and 20 weeks both SERCA (−30% and −40%,

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$p < 0.05$) and PLB (−14%, and −32%, $p < 0.05$) were decreased, while NCX progressively increased (+41% and +90%, $p < 0.05$). There were no signs of increased RV fibrosis in PAB.
Conclusion: Prolonged PAB resulted in compensated RV hypertrophy, with maintained RV systolic function and impaired diastolic function. LV function was hardly affected. The decrease in β-ARs and SERCA, commonly found in heart failure, were compensated for by decreases in GRK2 and PLB, respectively, and an increase in NCX. These molecular alterations are compatible with our hemodynamic findings of compensated hypertrophy.

P-72
Stem cells vascular differentiation in a right ventricular congestive heart failure model
Castellani Chiara (1), Angelini A. (1), Dalla Libera L. (2), Gobbo V. (2), Piccoli M. (3), Pozzobon M. (3), Valente M. (1), Thiene G. (1), Vescovo G. (4)
Cardiovascular Pathology (1); CNR Padua, department of biomedical science (2); department of pediatric Oncohaematology of Padua (3); division of Internal Medicine of Vicenza Hospital (4)

Background: Monocrotaline treated rats represent an animal model of right ventricular heart failure induced by pulmonary vascular injury and hypertension. Accumulating evidence suggests that stem cells may participate in the regeneration and remodelling of remote organs. Aim of our study was to investigate the role of human amniotic stem cells and rat stromal adipocite-derived vascular GFP positive cells in animal model of right heart failure.
Methods: 16 male Sprague –Dowley rats weighing 80 to 100 g were studied. They developed a secondary CHF at three weeks monocrotaline injection and received four million of human amniotic stem cells (hAFC) labelled with CMFDA (green fluorescence molecule) or rat vascular stromal adipocite-derived GFP (rGFP) cells via vein tail. At different times after SC injection hearts, lung and scheletal muscles were harvest. Stem cells detection and differentiation was studied by double immunofluorescence technique (c-Kit, oct4, SMA, alpha human mitochondria), with confocal microscopy.
Results: Green fluorescent-protein cells were detected at 24 hours and 72 hours after injection. Stem cells showed an undifferentiated phenotype with positivity for some of stem cells markers as c-Kit, oct-4 and SSEA-4 (1.9%). At one week after injection stem cells were detected in the vessel pulmonary structure as infiltrated vascular stem cells (0.7% hAFC and 0.44% rGFP). Double green-fluorescent/SMA cells were detected and mainly confined to the media layer of arterioles (0.12%). In the myocardium were mainly localized in the interstitium and the percentage was 0.24% for hAFC and 0.75% for rGFP positive cells. Rats with hAFC injection showed a 0.02 percentage of SMA differentiation while rats received rats GFP positive stem cells showed 0.37% of SMA differentiation.
Conclusion: These results suggest that amniotic and vascular stromal adipocite-derived stem cells are both engrafted in lung and heart vascular structure being detected in all the three layer of arterioles. At one week after injection, stem cells can differentiate in vascular smooth muscle and endothelial cells but not in cardiomyocytes.

P-73
Impact of fibrin and collagen matrices used in tissue-engineered heart valves on bacterial-endothelial interaction in a model of endocarditis
Heying R. (1,2), Wolf C. (1), Schmidt K.G. (2), Schroten H. (1)
Pediatric Infectious Diseases, Department of General Pediatrics, University Hospital Duesseldorf, Germany (1); Department of Pediatric Cardiology, University Hospital Duesseldorf, Germany (2)

Patients requiring heart valve prosthesis are at high risk of endocarditis. Heart valves based on the principle of tissue-engineering are under construction to minimize risk factors and improve biocompatibility.
In the present study we compare the influence of a fibrin and collagen gel matrix on bacterial adhesion and invasion. In addition endothelial pro-inflammatory activity was tested.
Staphylococcus aureus, Streptococcus sanguis and Staphylococcus epidermidis are among the most important bacterial pathogens responsible for endocarditis. ECs were cultivated on the two gels and polystyrene tissue culture plates as a control matrix.
ECs incubated with S. aureus showed a similar high infection rate cultured on fibrin ($3.3 \pm 0.9\%$ of the inoculation dose) and collagen gel ($3.2 \pm 0.8\%$ of the inoculation dose). These values were significant higher in relation to ECs cultured on tissue culture plates ($1.4 \pm 0.4\%$ and $1.1 \pm 0.3\%$; $p < 0.01$).
ECs incubated with S. sanguis and S. epidermidis led to a slightly lower infection rate on fibrin gel vs. collagen gel ($0.5 \pm 0.2\%$ vs. $1.2 \pm 0.3\%$ and $0.5 \pm 0.1\%$ vs. $1.3 \pm 0.4\%$ of the inoculation dose). Compared to ECs cultured on tissue culture plates the infection levels did not differ to the ones reached on the gel matrices. Values for S. sanguis and S. epidermidis were not significant comparing the three matrices.
Endothelial pro-inflammatory response investigated by expression of ICAM-1 showed a slightly higher S. aureus induces activation of ECs cultures on fibrin gel vs. collagen gel (8.5 vs. 5.2 fold increase) in a representative experiment. ICAM-1 levels of ECs cultures on a tissue culture plate were similar to the ones on fibrin gel (7.9 fold increase). Values after infection with S. sanguis and S. epidermidis supported these pattern.
We conclude that the matrix serving as a basic structure for tissue-engineered heart valves has an impact on the risk of endovascular infections. Especially investigations on endothelial activation suggest an influence of the matrix on endothelial pro-inflammatory response. Current further investigations might state the importance of different matrices as inducers of pathways to evoke inflammation, tissue damage and fibrin deposition at the infected endovascular sites.

P-74
Characterization of paediatric pulmonary hypertension: not as straightforward as the Venice classification
van Loon R.L.E. (1), Roofthooft M.T.R. (1), van Osch-Gevers M. (2), Delhaas T. (3), Strengers J.L.M. (4), Kapusta L. (5), Blom N.A. (6), Berger R.M.F. (1)
Department of Paediatrics, Divisions of Paediatric Cardiology, University Medical Center Groningen, University of Groningen, Groningen(1); Erasmus Medical Center Rotterdam, Rotterdam(2); University Hospital Maastricht, Maastricht(3); University Medical Center Utrecht, Utrecht(4); University Medical Center Nijmegen, Nijmegen(5); Leiden University Medical Center, Leiden(6); The Netherlands

Introduction: The Venice classification distinguishes 5 subtypes of pulmonary hypertension (PH), including pulmonary arterial hypertension (PAH). In paediatric patients, age-specific combinations of PH subtypes occur and various systemic-to-pulmonary shunt-defects (CHD) may be present. We aimed to describe characteristics of paediatric PH.
Methods: Between 1993 and 2007, 63 children were seen at the Dutch referral centre for paediatric PH. At presentation, PH subtype diagnosis was assessed, as well as associated conditions, transcutaneous oxygen saturation (TcSO2), World Health Organization functional class (WHO class) and hemodynamics.

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Results: Median age at presentation was 5.8 years (range 0.1–17.4). Symptoms included: dyspnoea at rest n = 16 (25%), exercise induced dyspnoea n = 48 (76%), chest pain n = 2 (3%), syncope n = 8 (13%). Mean TcSO2 was 91 ± 8%, WHO class I/II/III/IV (n = 2/16/29/16).
Strict adherence to the Venice classification led to the following diagnoses: idiopathic PAH (iPAH) 21 (33%), CHD associated PAH (PAH-CHD) 40 (63%), respiratory disorder associated PH (respPH) 1 (2%) and thromboembolic PH (CTEPH) 1 (2%).
Of the 40 PAH-CHD patients, 14 had "classic" Eisenmenger syndrome, 5 had PAH after previous shunt closure, 4 had accelerated PAH-CHD (already in the first months of life), and in 17 the CHD was regarded not explanatory for the PAH. In 7 of the latter patients, diagnostic work-up revealed additional respiratory problems as contributing factor to the PH; they were assigned to "respPH". The remaining 10 patients were reclassified as iPAH, after exclusion of other possible underlying conditions.
Thus, after adjusting for patients presenting with more than one associated condition, diagnostic classification revealed: iPAH 31 (49%), PAH-CHD 23 (37%), respPH 8 (12%), and CTEPH 1 (2%). Furthermore, accompanying chromosomal abnormalities or syndromes were present in 27 (43%) patients: Down 13 (21%), Noonan 2 (3%), velocardiofacial 2 (3%), Jacobsen 1 (1.6%), 1P36 deletion 1 (1.6%), NOMID syndrome 1 (1.6%), and non-specified developmental abnormalities or dysmorphic features 7 (11%).
Conclusions: Presentation of paediatric P(A)H is complex. Combinations of conditions associated with PH are often present. Determination of the contribution of each of these conditions to the course of PH may be difficult in the individual patient. In our series, the CHD was judged not primarily responsible for the PAH in 17 of 40 patients with PH and CHD. Furthermore, associated syndromal abnormalities were frequently present (43%).

P-75
Hematologic Abnormalities In Children With Dilated Cardiomyopathy
Kammache I., Elie C., Marini D., Legendre A., Bonnet D., Agnoletti G.
Necker Enfants Malades, Paris, France

Background: The aim of this work was to evaluate the prevalence and the prognostic value of haematological abnormalities in children with dilated cardiomyopathy (DCM).
Methods: From January 1985 to December 2005, 110 consecutive children had a first diagnosis of DCM. All patients had clinical, echographic, radiological and biological examination.
Results: Median age at diagnosis was 0.6 years (0–17).
Severe mitral regurgitation was found in 7 patients, cardiothoracic index > 0.7 in 7 and fractional shortening < 20% in 75. 39 patients needed inotropic support.
Anaemia (Hb < 10 g/dL) was present in 22 children, neutropaenia (< 1500) in 10 patients and lymphopaenia (< 1500) in 3 patients. Renal insufficiency (uraemia > 8 mmol/L) was found in 20 patients and was not related to the presence and degree of anaemia.
Patients were followed for a median period of 4.8 years (CI 95% 2.9 - 6.5). There were 26 deaths, occurred at a mean interval 68 days (1-2072) from diagnosis; 16 patients underwent cardiac transplantation. Age at diagnosis was not a predictor of poor outcome. Anaemia was present in 54% of patient who died or underwent cardiac transplantation and was highly related to a poor outcome (p = 0.005) and to the need of inotropic support (p = 0.02). The need of inotropic support was also predictive of poor outcome (p < 00001).
Conclusions: Haematological abnormalities are relatively frequent in children with DCM. In particular, the presence of anaemia is not related to renal insufficiency and is a sensitive prognostic factor or death and/or transplantation.

P-76
Bosentan for the treatment of Pulmonary Arterial Hypertension in Children
Gonçalves M.R., Freitas I., Paramés F., Martins J.D.F., Nogueira G., Trigo C., Pinto F.F., Kaku S.
Hospital de Santa Marta, Pediatric Cardiology Department, Lisbon, Portugal

Introduction: Bosentan, a dual endothelin receptor antagonist, has been introduced as an important oral Pulmonary Arterial Hypertension (PAH) therapy targeting over expression of endothelin-1. Short-term improvements have been demonstrated in adults; however data on paediatric use is limited.
Aims: To evaluate bosentan safety and effectiveness in children with PAH.
Methods: 4 years retrospective descriptive study of children with PAH treated with bosentan on top of conventional therapy, between January 2003 and December 2007. Data was collected from clinical charts, concerning baseline and follow-up. Age, gender, clinical diagnosis, New York Heart Association (NYHA) functional class, arterial oxygen saturation, calculated right ventricle pressure (echocardiography) and haemodynamic data, Pro-BNP levels, 6-min walking distance and adverse effects were collected. All children were admitted for a few days to initiate the therapy.
Results: Fifteen children were included. Ages ranged from 5 months to 21 years, 10 were female. Thirteen patients had congenital cardiac disease. All cases presented with cardiac failure, 9 with NYHA class IV and 6 NYHA class III. The average baseline arterial oxygen saturation was 82%. The media baseline right ventricle systolic pressure was 69 mmHg. Two children died during this period with no relation to bosentan (one during cardiac catheterization, one with respiratory infection). At the last follow-up, all the remaining patients (13) had improved (= 1 NYHA class), had higher arterial oxygen saturation and the media right ventricle systolic pressure was 60 mmHg. Decreasing levels were found in five out of the eight patients with Pro-BNP levels performed at the follow up. Three minor adverse effects were registered, none required treatment.
Conclusions: In our series, bosentan demonstrated to be both safe and effective. All patients had demonstrated clinical improvement during the follow up and no serious adverse effects were documented. Our experience suggests that Bosentan is an important treatment option for these children, but larger numbers studies are needed.

P-77
Beta-Blocker Therapy in Children with Dilated Cardiomyopathy: A Single Centre Experience
Gagliardi M.G., Pilati M., Saffirio C., Adorisio R., Sanders S.P.
Bambino Gesù Hospital, Rome, Italy

Background eta-adrenergic blockade showed the beneficial effects on motality and mobility in adult population with heart failure. Conflictive data exist in children.
Aim: This retrospective study is aimed to establish the efficacy and safety of beta-blocker therapy in our population of pediatric patients suffering from idiopatic dilated cardiomyopathy (DCM).
Methods: We reviewed the medical records and re-analyzed the echocardiograms of 15 patients, referred to our hospital from 2000 to 2006, with DCM (biopsy proven) and an ejection fraction

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(EF) < 40%, who were treated with beta-blocker therapy as adjunct to standard therapy including ACE-inhibitors. The mean age of the pts was 4.6 ± 3.9 years, mean weight 21.9 ± 14. Mean EF at diagnosis was 29,7 ± 11,1 %; 6 (40%) pts were in I NYHA functional class, 5 (33%) pts in II NYHA class, 3 (20%) pts in IV NYHA class, 1 pts (7%) in III NYHA class. Values are presented as mean ± SD. Statistical analysis was performed with a two-sample t test.

Results: In our patients beta-blocker therapy was started after a median of 1.5 years after the diagnosis. All the patients started the therapy at a dosage of 0,1 mg/Kg/die and the dose was up-titrated upon a mean of 0.7 ± 0.2 mg/Kg/die. Over a follow-up of 1.6 ± 1.2 years the EF increased significantly from 29.7 ± 11.1% to 42,7 ± 8% (p 0,0008). NYHA functional class improves in 13 pts (87%), one patient worsened to a IV NYHA functional class and one patient was transplanted and then died (7%). No pts had adverse effects related to the therapy and no pts discontinuated therapy.

Conclusions: In our population of children affected by DCM the addition of beta-blocker to standard therapy improves EF and NYHA functional class, without major adverse effects. Nevertheless there is the need of a prospective randomised study with a longer follow up to clarify the effects of this class of drugs on mortality.

P-78

Aggressive Treatment of Pulmonary Hypertension before Heart Transplantation in Children

Gagliardi M.G., Adorisio R., Pilati M., Di Donato R., Parisi F., Sanders S.P.
Bambino Gesù Hospital, Rome, Italy

Background: Increased pulmonary vascular resistance (PVR) is an independent predictor of mortality after orthotopic heart transplantation (OHT) in both adults and children and is considered a contraindication.

Aim: To evaluate the hemodynamic effects of a period of aggressive treatment with pulmonary vasodilators ± a ventricular assistance device (VAD) in children with restrictive cardiomyopathy and PAH.

Method: Three children not eligible for OHT because of PAH, were treated with sildenafil (0.5–1 mg /kg 4 times/day). In one case, epoprostenol and BVAD (Berlin Heart) were added after 2 mos. Two of the three cases underwent cardiac catheterisation after 4 mos. Mean PAP, CPWP, TPG and PVR were measured at baseline, under hyperoxia and with 25 ppm NO.

Results

#	Weight (Kg)	Age (yrs)	# Cath	Mean PAP (mmHg) (basal/O2/NO)	CPWP (mmHg) (basal/O2/NO)	TPG (mmHg) (basal/O2/NO)	PVR (U.W./m2) (basal/O2/NO)
1	28	12	1	22/24/22	12/17/17	10/7/5	4/2.8/1.4
2	21	6	1	32/38/38	17/24/24	15/14/14	8.3/7.7/8.2
			2a	36/36/25	16/16/15	20/20/10	8/8/4
3	13	4	1	35/35/35	16/16/25	19/19/10	8.26/8.26/6.5
			2b	35/34/30	19/19/20	16/15/10	3.5/3.5/2.2

2a = after 4 mos of sildenafil; 2b = after4 mos of sildenafil and 46 days of bi-vad and epoprostenol

Patient 1 did not have a second catheterization and underwent a successful HT.

In patient 2, treated with sildenafil alone, TPG and PVR did not improve either at baseline or under hyperoxia. However, the responsiveness to NO increased significantly after exposure to sildenafil. This child is awaiting transplantation. Patient 3, after 60 days of sildenafil, received a BVAD for intractable heart failure. Epoprostenol was added for an additional 46 days. A cardiac catheterization showed a reduction in the TPG and the PVR with no response to hyperoxia but a significant response to NO. She underwent a successful HT 25 days later.

Conclusions: These preliminary data indicate that aggressive treatment of PAH with vasodilators and mechanical assistance can reduce PVR and increase sensitivity to NO allowing successful orthotopic heart transplantation in some patients previously considered ineligible because of PAH.

P-79

Aortic Isthmic and placental flow patterns in fetuses with hypoplastic left heart syndrome: Implications for brain development

Fouron J.-C. (1), Siles A. (1), Roxanne Gendron R. (2), Lamoureux J. (1), Bureau N. (1,2), Andelfinger G. (2)
(1) Fetal Cardiology Unit, (2) Cardiovascular Genetics, Pediatric Cardiology Service, Department of Pediatrics, Sainte Justine Hospital, University of Montréal, Québec, Canada

Decreased blood flow to the fetal brain may contribute to neurologic sequelae in survivors of hypoplastic left heart syndrome (HLHS). In HLHS fetuses, placental characteristics and flow patterns at the aortic isthmus, which supplies the entire cerebral circulation, have not yet been investigated. We sought to study the role of circulatory competition between placenta and brain in fetuses with HLHS.

Echocardiographic isthmic Doppler flow patterns, umbilical and middle cerebral artery pulsatility indices (UAPI and MCAPI, respectively) were investigated in 39 HLHS fetuses. Autopsy records of 12 of these fetuses and 8 additional HLHS fetuses were compared with age-matched normals for organ weights.

In diastole, 32/39 (82%) fetuses had forward isthmic flow (F-IDF), indicating placental steal at the expense of the cerebral circulation. At autopsy, the linear correlation of placental and brain weight in normal fetuses ($n = 21$, $r = 0.614$, $p = 0.002$) was lost in HLHS ($n = 20$, $r = -0.061$, $p = 0.414$), which could be attributed to disproportionately small placentas in HLHS fetuses. This suggests a placental contribution to developmental perturbations in HLHS. Paradoxically, higher placental versus cerebral resistance was associated with reduced reverse isthmic systolic flow integrals (MCAPI/UAPI versus R-ISFI, $r = 0.455$, $p = 0.025$).

These findings identify isthmic flow patterns as cerebral risk markers in HLHS fetuses. Further investigations into consequences and possible developmental causes of placental steal are warranted.

P-80

Mechanisms in fetal bradyarrhythmia: seventeen years experience with established and new Doppler echocardiographic techniques

Sonesson S.-E., Eliasson H.
Pediatric Cardiology, Karolinska Institutet, Stockholm, Sweden

Introduction: AV-block (AVB) and blocked supraventricular bigeminy (BB) cause fetal bradycardia. Whereas BB resolves spontaneously without treatment, congenital AVB might respond to steroid treatment if incomplete at time of diagnosis. As steroid treatment has potentially serious side effects, a correct diagnosis is of major importance. To more precisely establish the electrophysiological mechanisms in fetal bradycardia we have not

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only used conventional m-mode and Doppler echocardiographic techniques but also included flow velocity recordings from the pulmonary artery and the ductus venosus.
Objectives: To evaluate the magnitude of the clinical problem in differentiating fetal AVB from BB and the potential of current and new Doppler echocardiographic techniques in making a correct diagnosis.
Methods and Results: Retrospective analysis of eighty-five consecutive patients from 1990 to 2007, referred to a tertiary centre of fetal cardiology, for evaluation of bradycardia +/- suspicion of heart malformation. Patients were divided into two groups. A. (HR < 100) comprised 24 fetuses with AVB (4 incomplete, 20 complete) of which 7 had a cardiac malformation and one LQTS. Twenty-six had BB (5 sustained, 21 intermittent) of which one had a cardiac malformation and 3 later developed supraventricular tachycardia. B. (HR > 100) included 16 fetuses with intermittently blocked premature atrial contractions. Of another 19 fetuses with regular heart rate, 3 were postnatally diagnosed with LQTS. Intermittent BB was usually diagnosed in late gestation, whereas almost all cases of sustained BB and AVB were diagnosed during the second trimester. A HR < 65 was never seen in BB (range 65–88), and a HR < 60 was only seen in complete AVB (range 25–70). A relative time interval constructed from the ductus venosus records; the interval between the conducted and following blocked atrial contractions divided by the time interval between two conducted atrial beats, was 0.24–0.47 in BB and 0.49–0.50 in AVB II. Pulmonary artery recordings were particular helpful in differentiating AVB III from AVB II and BB.
Conclusion: BB is a common cause of fetal bradycardia. During mid-gestation it is frequently sustained and with a higher degree of resemblance with AVB. Using new Doppler techniques a correct diagnosis can still be made.

P-81
Pre and post-natal outcome of fetal tachycardias: retrospective analysis of 41 consecutive cases
Paoli F., Poinsot J., Vaillant M.C., Chantepie A.
Hôpital Clocheville, CHRU, Université François Rabelais, Tours, France

Objective: To review the pre and postnatal management and outcome of fetal tachycardia
Methods: From 1984 to 2006, 41 consecutive cases of fetal tachycardia from a single tertiary centre for paediatric cardiology were retrospectively evaluated.
Results: The median gestational age at presentation was 31,5 weeks (range 21 to 40). The diagnosis was made during routine fetal monitoring in 39 cases, and further to a decrease of the fetal active movements in 2 cases. Thirty three fetuses had a supraventricular tachycardia (SVT), 6 had atrial flutter and 2 had multifocal atrial tachycardia. Thirty four fetuses were non-hydropic and 7 were hydropic (all of them with SVT). Seven fetuses with non-sustained SVT did not receive any treatment. Transplacental therapy as first line therapy was given in 34 patients, digoxin only (16), digoxin associated to amiodarone or sotalol (8), amiodarone (6), sotalol (2), flécainide (1), verapamil (1). Prenatal conversion of the tachycardia was achieved in 25 of the 27 treated non-hydropic fetuses (89%) and in 4 of the 7 treated hydropic fetuses (57%). Partial success was observed in 2 non hydropic patients. Drug failure occurred in 3 hydropic patients of whom one died. The overall mortality is 3% in the treated fetuses. Two adverse effects occurred in relation to maternal digoxin toxicity: second degree AV block and vomiting ; a transient hypothyroidy was observed in a patient treated with amiodarone. The conversion rate obtained with digoxin was 65% (13/20)) in non-hydropic fetuses and 25% (1/4) in hydropic patients. Among the 40 alive newborns, 25 received an anti-arrhythmic treatment during 6 months. Six neonates presented a supraventricular arrhythmia . There were 2 patients with recurrent or chronic tachycardia.
Conclusion: In the absence of hydrops, digoxin is effective in most of fetal tachycardias. The risk of maternal toxicity leads to propose a close monitoring of maternal EKG and digoxin serum level. The treatment of tachycardia in hydrops fetus is still a challenge and requires a multicenter trial to determine the optimal strategy. The post-natal prognosis is excellent.

P-82
High Frame Rate Vector Velocity Imaging: a Novel Approach to Assessment of Fetal Cardiac Function in Congenital Heart Disease using a Speckle Tracking Algorithm
Matsui H. (1, 2), Germanakis I. (1, 2), Roughton M. (2), Gardiner H. (1, 2)
Faculty of Medicine, Imperial College at Queen Charlotte's & Chelsea Hospital, UK (1); Royal Brompton Hospital, London, UK (2)

Background: Vector velocity imaging (VVI) tracks natural myocardial echogenic foci in the 2D echocardiographic image (speckle tracking) enabling measurement of myocardial strain and torsion. Currently commercially available equipment enables capture of low frame rate images of 25 Hz reducing ability to speckle track in the fetus. We have developed a novel method of high frame rate capture at 100Hz and compare cardiac function in normal and abnormal fetal ventricles.
Methods: We prospectively stored original DICOM data using a dummy electrical signal from 4 chamber loops in 84 normal singleton pregnancies, median 22 (14–36) weeks' gestation. Off-line VVI ventricular analysis was performed using Syngo-VVI software (Siemens) by creating a tracking movie automatically from the 4 chamber view. The longitudinal myocardial strain of LV and RV free walls were measured and we compared these data with 27 fetuses showing cardiac malformations: Coarctation of aorta (7), Tetralogy of Fallot (4), atrioventricular septal defect (4), hypoplastic left heart syndrome (HLHS; 3), Ebstein anomaly (2), transposition of great arteries (3), mild pulmonary stenosis (2), critical pulmonary stenosis (1) and cardiac tumor (1).
Results and Discussion: Normal Fetuses: median frame rate was 87 Hz (36–191) and median fetal heart rate 145 beats per minute (36 frames per beat). Automatic tracking of VVI was impossible in 52 normal (25 RV and 27 LV) and 16 abnormal ventricles. Although systolic and diastolic velocities showed a significant gestational increase with (RV > LV), longitudinal strain did not alter between LV and RV free wall. (0.21 in LV vs. 0.20 in RV: $p = 0.80$). CHD: compared to normal values, longitudinal strain in HLHS and cAS was not significantly different in RV median 0.23 (0.16–0.28), but reduced in LV 0.09 (0.03–0.14). In critical PS longitudinal strain was normal at 0.21 in RV and at 0.23 in LV. In Ebstein anomaly, reduced longitudinal strain of the RV (0.16 (0.13–0.21)) suggested impaired RV function while the other malformations showed normal longitudinal strain (0.22 ± 0.05, $p = 0.85$) suggesting the ventricular stress-strain relationship was preserved.
Conclusion: High frame rate speckle tracking is useful to assess fetal cardiac function and may refine assessment of timing of intrauterine interventions or need for delivery.

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P-83

Progression of pulmonary-aortic asymmetry in foetus: an aid to predict neonatal coarctation

Bonnet D., Goulet M., Fermont L., Le Bidois J., Maltret A., Bajolle F., Stos B.
M3C-Pediatric Cardiology, Necker-Enfants Malades, Paris, France

Background: Prenatal diagnosis of coarctation of the aorta (CoA) suffers from high false positive and negative rates.
Objective: To measure the progression of main pulmonary artery/ascending aorta ratio (MPA/Ao) between 2nd and 3rd trimesters and to correlate foetal evolution with post-natal outcome.
Methods: 120 foetuses with suspicion of CoA during 2nd trimester were followed sequentially during pregnancy. MPA/Ao was measured at each echoscan. Progression of asymmetry was considered present if MPA/Ao increase was >0.3. Outcome of pregnancy was noted. Postnatal evaluation included: diagnosis of CoA, presence of bicuspid valve at first echocardiography and any left heart anomaly during follow-up (congenital mitral stenosis or insufficiency, subaortic stenosis) in patients who did not have CoA.
Results: Termination of pregnancy was performed in 19 cases (15 chromosomal or extracardiac anomalies; 4 severely diminutive left ventricle). Fifteen foetuses were lost for follow-up. 86 neonates were born alive: 49 developed a CoA and 37 did not. MPA/Ao was significantly higher in foetuses with post-natal CoA both at 2nd and 3rd trimester ($p < 0.001$). MPA/Ao increase >0.3 had a 88.5% specificity to predict CoA. When bicuspid aortic valve was present, 91% of foetuses with MPA/Ao increase had a CoA. Finally, considering all left heart anomalies during follow-up, 93% of foetuses with MPA/Ao increase had at least one left heart lesion requiring follow-up.
Conclusion: Progression of left-right asymmetry aids to predict postnatal CoA or left heart lesions with a high specificity. In the absence of MPA/Ao increase, false positive rate remains high.

P-84

Comprehensive considerations for prenatal counseling in presence of fetal cardiac rhabdomyoma

Chockalingam P. (1), Degueldre S. (1), Mivelaz Y. (1), Di Bernardo S. (1), Pfammatter J.P. (2), Barrea C. (3), Sekarski N. (1), Jeannet P.Y. (1), Fouron J.C. (4), Vial Y. (1), Meijboom E.J. (1)
University Hospital of Lausanne, Switzerland (1); University Hospital of Bern, Switzerland (2); University Hospital of Saint-Luc, Bruxelles, Belgium (3); Centre Hospitalier Universitaire de Sainte-Justine, Montreal, Canada(4)

Background: This study analyses the long-term cardiac and neurological outcome of patients with cardiac rhabdomyoma (CR) in order to allow comprehensive prenatal counseling.
Methods and Results: A retrospective study including all cases with echocardiographic diagnosis of CR encountered from August 1982- September 2007.
Of 72 CR patients identified, 19 were diagnosed in-utero at a gestational age between 20-35 weeks and 53 postnatally between 10 days-11 years. 50 had multiple CR and 22 had single CR. They were situated predominantly in the LV (64%), RV (43%), IVS (50%) and to a lesser extent in the atria (9%) and pericardium (1%). Follow-up echocardiography over 3 months-18 years in 56 showed complete postnatal regression of CR in 15, partial regression in 24 and no changes in 16, in 1 case the nodules were enlarged after 6 years. Cardiac complications were encountered in 12 patients, 8 arrhythmias, of which 4 with paroxysmal SVT and 2 with WPW syndrome, 2 with sub-aortic obstruction, 1 with pulmonary obstruction requiring surgical intervention and 1 with cardiogenic choc due to outflow obstruction causing death. Long-term follow-up revealed tuberous sclerosis of Bourneville (TSB) in 55, complicated by epilepsy in 44 and developmental delay in 36.
Conclusion: CR generally regresses after birth and after the perinatal period cardiac-related problems are rare, but TSB and the associated neurodevelopmental complications dominate the clinical picture and should form an important aspect of the prenatal counseling of parents. An association between multiple CR and TSB compared to single CR and TSB could not be established.

P-85

How to improve effectiveness of fetal supraventricular tachycardia treatment - causes of therapeutic failure

Dangel J.H. (1), Hamela - Olkowska A. (1), Bieganowska K. (2)
Perinatal Cardiology Clinic2nd Department of Obstetrics and GynecologyMedical University of Warsaw, Poland (1); Cardiology Department Children's Memorial Health Institute, Warsaw, Poland (2)

Introduction: Supraventricular tachycardia (SVT) is the most common fetal tachyarrhythmia. Transplacental treatment has been used since years but there is not universal approved treatment.
The aim of this study was to evaluate 50 fetuses with SVT and atrial flutter (AF). Type of arrhythmia, fetal cardiovascular profile score (CVS), treatment, neonatal follow-up, reasons for success or failure and evolution of antiarrhythmic treatment were analyzed.
Material and Results: 36 fetuses with SVT (21-short VA time, 15–long VA time), and 14 with AF were analyzed. CVS was used for evaluation of fetal condition and monitoring. Until 2002 12 fetuses with SVT and 3 with AF were treated. Digoxin was used in all, digoxin and sotalol in 3, adenosine intraumbilically in 2. Treatment was successful in 50%. There was one perinatal death and one neurological damage and death. Due to poor results of treatment in 2002 we decided to change therapeutic policy. Knowing published results of amiodarone therapy we decided to use this drug in the most resistant cases. It was drug of the first choice in fetuses with SVT - long VA time and in all kind of SVT with NIHF. If SVT - short VA time without hydrops or AF persists for more than 7 days on digoxin – amiodaron was added. Time for conversion to the sinus rhythm was 16 days in long VA-time and 5 days in short VA-time. No conversion was in in 3 cases: tachy-brady, 2 long VA time, 1 in obese mother. There were not major complications due to amiodaron in neonates, transient thyroid dysfunction was in 4. Phlebitis was in 3 mothers. After that we decided to use amiodarone only orally. Since then there were not complications among mothers.
Conclusion: Effective method of transplacental antiarrhythmic treatment was possible after some years of personal experience. Intravenous loading doses are necessary only for digoxin, amiodaron should be administered orally in decreasing doses. Adenosine might be helpful in very rare conditions, when therapeutic level is impossible to obtain. Amiodarone is an affective and save drug for tachyarrythmia treatment in fetuses. Long VA time SVT and mother's obesity were main reasons for treatment failure.

P-86

Clinical Features, Management and Survival of Children with Fetal (F) and Postnatal (PN) Diagnosis of Common Truncus Arteriosus (TA): A Single Institution's Experience Since 1990

Dillenburg R., Guerra V., Van Arsdell G., Jaeggi E.
The Hospital for Sick Children555 University AveToronto, ON, Canada

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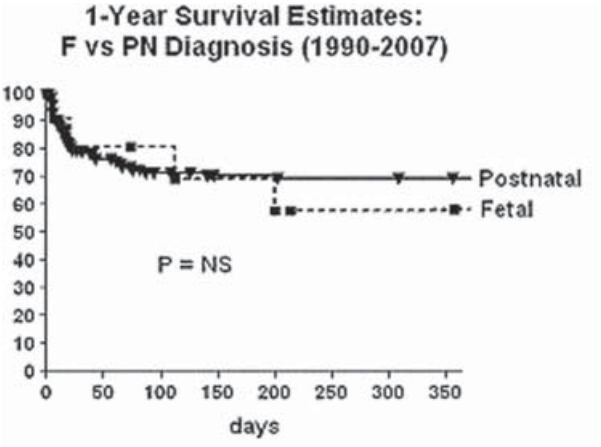
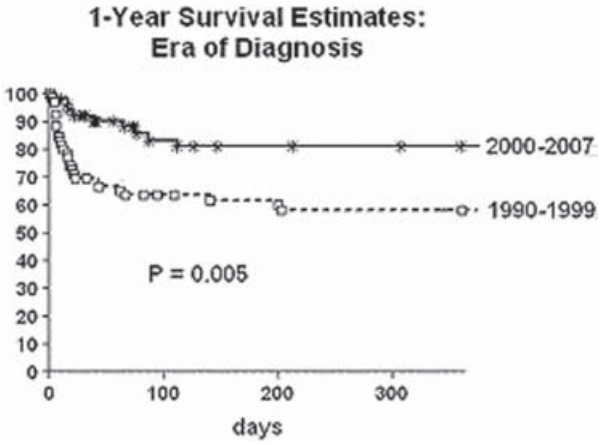
Background: TA is often associated with significant co-morbidity and mortality; however differences in survival between children with fetal versus postnatal diagnosis of TA are unknown.
Objectives: To compare clinical features, management and survival of F versus PN diagnosed TA.
Methods: We reviewed medical charts and echocardiograms of all TA cases referred between 1990 and 2007. Patient characteristics and outcomes were compared for F vs. PN diagnosis and for era of diagnosis (1990–1999 vs. 2000–2007).
Results: Of 153 patients, 25 (16) had a fetal TA diagnosis since 1990, predominantly (n = 18) during the recent 7 years. Clinical characteristics were comparable between F and PN diagnosis: most had type-1 TA (F: 68 %; PN: 57%) with functional truncal valve anomalies (60%; 70%). More frequent additional pathology other than septal defects included pulmonary artery hypoplasia (20%; 31%) and aortic arch interruption (20%; 15%). Microdeletion 22q11 was diagnosed in 45% of the tested fetal cases and in 35% of the PN cases. Pregnancy termination occurred in 32% of F cases, while 16% died in utero. One-year survival estimates of children with F and PN TA diagnosis since 1990 were 59% and 70% respectively (Figure 1). Survival to 1 year of age was significantly improved since 2000 (Figure 2) although age at surgical repair did not significantly differ among the two study eras.
Conclusion: Fetal detection rate of TA has significantly improved in recent years to 29%. While prenatal diagnosis significantly impacted pregnancy management and facilitates postnatal care, it did not to improve survival.





P-87
Redefining High Risk Referral Criteria in Fetal Cardiology
McBrien A., Sands A., Craig B., Casey F.
The Royal Belfast Hospital for Sick Children, Belfast, Northern Ireland

Introduction: Northern Ireland has a population of 1.7 million. There is a single, regional fetal cardiology service. The indications for referral to our clinic are those recommended by the fetal working group of the AEPC. Clinics are mainly consultant led. More than 90% of fetal echocardiograms performed are normal. We believe that this system is inefficient and have gathered data to facilitate streamlining the service.
Methods: Records from the fetal cardiology clinic and the regional database were used to identify all patients seen in 2005 to 2007. Referral reasons and known risk factors at first attendance were documented. The presence or absence of major congenital heart disease (CHD) was determined by the result of the fetal echocardiogram. Patients referred for assessment for fetal cardiac arrhythmia were excluded. Logistic regression was performed to identify the significant risk factors for major structural CHD.
Results: 1074 women were seen during the three years studied. 964 women met the inclusion criteria. 80 fetuses (8.3%) had major CHD. 3 women (with normal fetal echocardiograms) were excluded from the analysis as the referral reason was unknown. 3 statistically significant risk factors for major CHD were identified: abnormal/inadequate obstetric scan, fetal extra-cardiac malformation and known abnormal fetal karyotype. If only patients with any of these 3 risk factors were scanned, 621 fewer (65%) fetal echocardiograms would have been performed and 75 out of 340 (22%) would be positive for major CHD. Using this policy, 75 of the 80 (94%) major abnormalities would have been diagnosed. The risk for other referral reasons is 5/621 (0.8%).
Conclusions: By altering referral criteria, the efficiency of fetal cardiology clinics could be greatly improved. Many current referral criteria do not provide sufficient numbers of patients with CHD to justify their use. We suggest that only women with any of the 3 highest risk factors should be seen at the consultant led clinic. Those at lower risk should be scanned by ultrasonographers with expertise in fetal echocardiography.

P-88
Prenatal brain pathology in congenital heart disease
Maschke-Mlczoch E. (1), Brugger P. (2), Hanslik A. (1), Salzer-Muhar U. (1), Prayer D. (2)
Clinical Department for Pediatric Cardiology, University Clinic for General Pediatrics and Neonatology, Medical University of Vienna, Austria (1); Clinical Deparment for Neuroradiology, University Clinic for Radiology, Medical University of Vienna, Austria (2)

Purpose: To identify the type and incidence of fetal brain pathology in fetuses with cardiac malformations
Material and Methods: 57 pregnant women underwent 1 or 2 fetal MR-examinations between 20 and 38 gestational weeks. MR was done on a 1.5T superconducting system. The type of cardiac malformation was defined by fetal ultrasound. Genetic workup was available in 15 cases.
Results: In 33 cases (59%) we found brain pathology. 18 fetuses presented with malformations, consisting of commissural anomalies (3), holoprosencephaly (1), pathologies of the posterior fossa (4), tuberous sclerosis (3), malformations of cortical development (3), and combined malformations (4) In 12 fetuses symmetrical /asymmetrical widening of the internal/ external cerebrospinal

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fluid spaces was seen, consisting of ventricular enlargement, and/or widened subarachnoid spaces, 2 showed germinolytic cysts, and one displayed hemorrhagic lesions.
Cardiac malformations consisted of: Tetralogy of Fallot (9), transposition of great arteries (3) ventricular septal defect +/- associated cardiac malformations (9), rhabdomyoma (3), Coarctation (3), Atrioventricular Canal Defect (3), cardiomegaly (uni-or bilaterally) (3) other pathologies, such as aneurysm, cardiac teratoma, missing inferior vena cava, and double outlet right ventricle were seen one each, in the remaining cases the cardiac malformation could not yet be classified .
Genetically, in 3 cases a Microdeletion 22q11 was found, in two a trisomy 13. The remaining 11 tested cases had normal chromosomes.
Conclusion: The association between congenital heart disease and cerebral impairment is well known (1). Fetal MRI of the brain in congenital heart disease is a new method to investigate early onset signs of cerebral changes in fetal brain development. The prenatal origin of brain pathology in such cases was suspected. This is now proved by the results of this study. In addition, the wide range of anomalies is demonstrated, consisting of malformations with, at least partly, genetical background, and acquired disease as a consequence of altered cerebrovascular hemodynamics (2)

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(2) Kaltman et al. Impact of congenital heart disease on cerebrovascular blood flow dynamics in the fetus. Ultrasound Obst. Gyn 2005; 25: 32-36

P-89

Assessment of Diastolic Ventricular Function in Fetuses of Diabetic Mothers Using Tissue Doppler

Zielinsky P., Hatem M.A.B., Hatem D.M., Nicoloso L.H., Manica J.L., Piccoli Jr A.L., Zanettini J., Oliveira V., Scarpa F., Petracco R.
Institute of Cardiology of Rio Grande do Sul / FUC. Porto Alegre, Brasil

Objective: To identify the presence of ventricular diastolic dysfunction by tissue Doppler in fetuses of diabetic mothers, with or without septal hypertrophy, in comparison to fetuses of nondiabetic mothers.
Methods: A contemporary transversal study in which the diastolic function was studied by tissue Doppler and pulsed wave Doppler of the atrioventricular diastolic flow in fetuses with a gestational age between 25 weeks to term whose mothers had previous or gestational diabetes and who were referred to the Fetal Cardiology Unit of the Institute of Cardiology in Porto Alegre, Brazil. Pearson's correlation tests and variance analysis with Student-Neumann-Keuls post hoc test. An alfa of 0.05 was considered significant for statistical analysis.
Results: Mean myocardial velocities of the E´and A´ waves at the posterior mitral annulus, in fetuses of diabetic mothers with myocardial hypertrophy were, respectively, 7.00 plus or minus 1.6 cm/s and 10.24 plus or minus 3.3 cm/s; in the fetuses of diabetic mothers group without myocardial hypertrophy were they were, respectively, 7.19 plus or minus 2.4 cm/s and 10.77 plus or minus 3.77 cm/s; and in the control group they were 4.81 plus or minus 0.85 cm/s and 8.01 plus or minus 2.2 cm/s. The difference between the velocities in fetuses of diabetic mothers and in fetal normal mothers was statistically significant (p less than 0.05). Statistically significant differences were also observed in E' and A' diastolic waves at the anterior mitral annulus, as well as at the tricuspid annulus by tissue Doppler in the same sample. Mean E/E' ratio of mitral and tricuspid waves in the control fetuses (normal mothers) was significantly higher than in fetuses of diabetic mothers.
Conclusion: Pulsed tissue Doppler in fetuses of diabetic mothers, independently of the presence of myocardial hypertrophy, shows evidences of impaired diastolic function, when compared with fetuses of non diabetic mothers.

P-90

Prenatal Diagnosis Of Congenital Heart Defects: A 5-year UCL Experience

Barrea C., Nimbona G., Sluysmans Th., Bernard P., Moniotte S.
Cliniques Universitaires Saint Luc, Brussels, Belgium

Introduction: Fetal echocardiogram is a standard of care in our institution in fetuses declared at risk for congenital heart defect (CHD) by obstetrical ultrasound. Our objectives were 1) to analyse the spectrum of observed CHD by transabdominal echocardiogram in this high risk population; 2) to evaluate associated extracardiac and chromosomal anomalies and outcome and 3) to compare pre and postnatal diagnosis.
Materials: From 01/2003 to 08/2007, 295 pregnancies were referred for echocardiogram for suspicion of CHD or arrhythmias (n = 37). The following variables were collected prospectively (n = 258 fetuses with CHD): gestational age at diagnosis, fetal and neonatal cardiac diagnosis, associated extracardiac anomalies, karyotype, pregnancy outcome and autopsy when appropriate.
Results: The first echocardiogram was obtained at a mean gestational age of 26 weeks (range 15-39 weeks). The most common CHDs were single ventricles (64/258, 24.8%) of whom one third were hypoplastic left heart syndrome. The prevalence of atrio-ventricular septal defects (AVSD), ventricular septal defects, tetralogy of Fallot and variants was quite similar (~10%). Suspected coarctation of the aorta and isolated transposition of the great arteries were present in ~5%. Extracardiac anomalies were present in 26.4 % (68/258) and chromosomal anomalies in 20.1% (36/179). The chromosomal abnormality rate was the higher in AVSD (75%). As for pregnancy outcome, there were 71 (27.5%) terminations of pregnancy (TOP). The most common causes of TOP were single ventricles (43/71, 60.6%) and chromosomal anomalies (18/71, 25.4%). There were 9 intra-uterine fetal deaths (IUFD). Pre and postnatal comparison was possible in 175 cases (67.8%). Total agreement between pre and postnatal diagnosis was found in 77.1 % (135/175). In 2 cases the neonatal diagnosis was more severe and TOP could have been offered prenatally. In 3.4% (6/175) the neonatal diagnosis induced a change in the prognosis. There were 32 cases (18.3%) of mild errors (without prognostic change).
Conclusion: The spectrum of CHDs encountered prenatally is different from postnatally. The high association rate of heart defects with extracardiac and chromosomal anomalies requires a multidisciplinary team for appropriate management and karyotyping seems of major importance. Pre and postnatal correlation is comparable to results of the literature.

P-91

Myocardial Tissue Doppler in the Evaluation of Diastolic Function in Fetuses With Intrauterine Growth Restriction.

Naujorks A., Zielinsky P., Beltrame P.A., Castagna R., Petracco R., Busato A., Nicoloso L.H., Piccoli Jr A.L., Manica J.L.
Institute of Cardiology of Rio Grande do Sul/FUC. Porto Alegre, Brazil

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Objective: In the complex cardiac changes in fetuses with intrauterine growth restriction (IUGR), diastolic dysfunction occurs early in the process. Myocardial tissue Doppler (MTD) is a technique that allows estimation of myocardial velocities in systole and diastole. This study aimed to assess the role of MTD in the evaluation of diastolic function in fetuses with IUGR.
Methods: The sample was made up of 14 fetuses with IUGR (group 1), 13 with appropriate weigh for gestational age (AGA) of hypertensive mothers (group 2) and 29 AGA fetuses of healthy mothers (group 3). Patients with other diseases were excluded. MTD was performed with the volume- sample placed in the basal segment of the left ventricle side wall (LV), interventricular septum (IVS) and right ventricular free wall (RV). E'/A'ratio was obtained in each location.
Results: There was a difference between groups for the E'/A' in the SIV ($p < 0.001$) and LV ($p = 0.009$), being higher in group 1 (IVS: 0.92 ± 0.28; LV: 0.85 ± 0.19) than in group 2 (IVS: 0.62 ± 0.09; LV: 0.68 ± 0.14) and 3 (IVS: 0.71 ± 0.14; LV: $0,69 \pm 0.15$). In the RV position, there was no statistically significant difference in the E'/A' ($p = 0.2$), even though mean ± SD in group 1 (0.86 ± 0.31) was higher than in groups 2 (0.68 ± 0.1) and 3 (0.66 ± 0.09).
Conclusion: Early to late myocardial velocities ratios are higher in fetuses with IUGR than in AGA fetuses, regardless of maternal hypertension. MTD can be a sensitive method to assess of cardiac diastolic dysfunction in fetuses with IUGR.

P-92
Behavior of Left Atrium Shortening Fraction in Fetuses of Diabetic Mothers with and Without Myocardial Hypertrophy
Zielinsky P., Luchese S., Manica J.L., Piccoli Jr A.L., Nicoloso L.H., Leite M.F, Hagemann L., Busato A., Moraes M.R.
Institute of Cardiology of Rio Grande do Sul / FUC. Porto Alegre, Brasil

Introduction: Approximately 7% of pregnancies are affected by diabetes and the risk of complications has not been reduced. Myocardial hypertrophy may occur in up to a third of fetuses of diabetic women. In adults with myocardial hypertrophy, left atrial shortening fraction (LASF) may be more reliable to assess diastolic function than mitral Doppler.
Objective: To test the hypothesis that LASF decreases in fetuses of diabetic women with myocardial hypertrophy, compared to those without hypertrophy and to fetuses of mothers with no systemic pathology.
Patients and Methods: Echocardiography was used to examine fetuses of women with previous or gestational diabetes and fetuses of normal women at gestational ages from 25 weeks onwards. LASF was obtained by the ratio: telesystolic diameter – presystolic diameter/telesystolic diameter. Data were compared using ANOVA and Tukey's test, and Pearson's coefficient for association between variables.
Results: Ninety-eight fetuses were studied, being 45 from control mothers. Out of the 53 fetuses of diabetic women, 14 had myocardial hypertrophy and 39 had normal septal thickness. Gestational age was similar ($p = 0.577$). Fetuses of diabetic mothers with hypertrophy presented a mean LASF of $0.32 + 0.11$, without hypertrophy of $0.46 + 0.12$, and those of normal mothers of $0.53 + 0.09$ ($p = 0.001$). A significant inverse linear correlation was observed between LASF and septal thickness ($r = -0.510$, $p = 0.001$).
Conclusion: LASF is lower in fetuses of diabetic women with myocardial hypertrophy as compared to those without hypertrophy and to those of normal mothers. It is suggested that LASF could be useful as an alternative parameter in the assessment of fetal diastolic function.

P-93
Aberrant right subclavian artery in the fetus
Zidere V., Borenstein M., Nicolaides K.H., Allan L.D.
Harris Birthright Research Centre for Fetal Medicine
King's College Hospital, London, UK

Objective: An aberrant right subclavian artery (ARSA) is an asymptomatic variant, occurring in about 1–2% of normals but increased in trisomy 21, with a range of 10-35%. Our aim was to document the origin of the right subclavian artery in the fetus.
Methods: Between March 2006 and April 2007, fetal echocardiography was performed prospectively in 1,995 patients from 14 to 37 weeks of gestation. Colour flow mapping was used to identify the right subclavian artery and determine whether this was normal or aberrant. Fetuses with a right aortic arch were excluded from the study. Data concerning karyotype and outcome were collected.
Results: The right subclavian artery was successfully identified in 1852 (93.2%) of the 1987 cases, with 135 failures. The mean gestational age at diagnosis was 20 weeks. An aberrant right subclavian artery was diagnosed prospectively in 33 fetuses, 25 with a normal heart and 9 (27.3%) with associated intracardiac malformations.
The karyotype was known in 628/1852 cases (34%) and there was a normal live-birth in a further 779. Forty pregnancies ended in termination, intrauterine death or neonatal death and the karyotype status remains unknown. Outcome is not yet available in the remaining 405 cases, including three fetuses with ARSA. An aberrant right subclavian artery was observed in 11 of the 573 fetuses with normal karyotype and in 8 of the 779 normal live-births (1.4%). An aberrant right subclavian artery was found in 8 (32%) of the 25 cases with trisomy 21 and in 3 (10%) of the 30 fetuses with other chromosomal defects. Confirmation of ARSA was obtained in two cases at surgery for congenital heart disease.
Conclusion: Despite the limited nature of postnatal follow-up, prenatal images were felt to be sufficiently convincing to make the correct diagnosis in a high percentage of cases. As previously reported, the background rate of ARSA was found to be 1–2%, whereas a higher rate (>30%) was found in the fetus with trisomy 21. Identification of the course of the right subclavian artery can be used to adjust the estimation of risk for trisomy 21 during pregnancy.

P-94
Myocardial performance in fetuses with twin-to-twin transfusion (TTTS) syndrome – serial follow up
Herberg U. (1), Bürücüoglu T. (1), Kotzmann J. (1), Gembruch U. (2), Breuer J. (1), Kohl T. (2)
Pediatric Cardiology (1); Prenatal Medicine (2) of the University of Bonn, Germany

Introduction: In twin-to-twin transfusion syndrome (TTTS), intertwin placental anastomoses lead to severe volume overload of the recipient twin, resulting in ventricular dilatation, tricuspid regurgitation and severe ventricular dysfunction while the donor twin does not show cardiac disease. Laser coagulation of placental anastomoses stop intertwin transfusion, but the consecutive adaptation of ventricular function has not been serially followed up.
Methods: 10 twin-pregnancies with severe TTTS were examined serially before and after endoscopic LC (day 0, 1, 2; week 1, 2, 4, 8,

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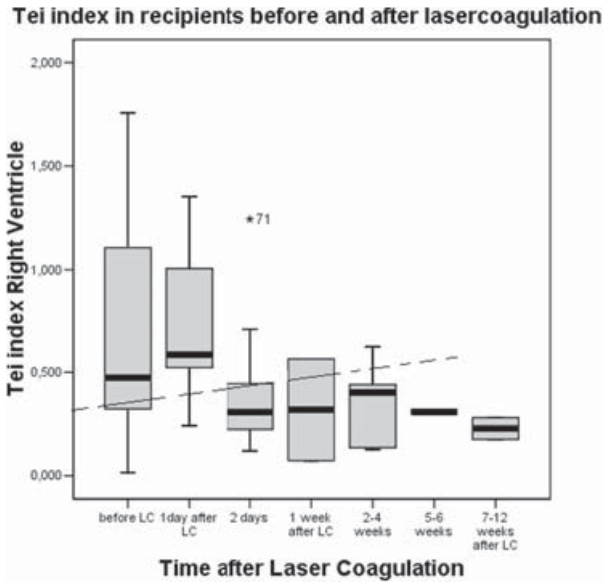
12) and compared to a control group of 4 monochorional diamniotic twins without TTTS and 47 singleton pregnancies. (Gestational age: TTTS: 22 [15-34], control group: 23 [15-35]). Ventricular function was assessed by M-mode, 2D-measures of ventricular diameter and wall thickness, Tei-index (myocardial performance index), Doppler of ductus venosus, umbilical artery and vein and cerebri media. Surviving fetus were follow up postnatally.

Results: Before LC, the recipient twin showed significant – predominantly RV – hypertrophy and dilatation. Myocardial performance index (Tei) was significantly increased for the RV and LV compared to donor twins and the control group (Tei RV: p 0.005; Tei LV: p 0.016). Tei correlated with the thickness of the interventricular septum, RV contractility and stiffness and ductus venosus PIV. After successful LC, RV and LV Tei indices decreased subsequently and reached normal values. (fig). Tei did not correlate with fetal outcome. In donors, ductus venosus PIV and Tei increased immediately after LC and decreased to normal values during follow up.

Conclusions: Even severe ventricular dysfunction in recipients with TTTS can resolve after successful LC of intrauterine placental anastomoses. The ventricular performance (Tei) index reflects ventricular stiffness, does not correlate to acute volume changes and is not a predictive factor of fetal outcome.



Tel-index in receipients before and after lasercoagulation (LC): RV Tei-index in recipients were significantly increased before LC compared to donors and the control group ($p < 0.005$; normal values for Tei = 0.38[0.13−0.62]) and declined to normal values after successful LC

P-95

Classification of foetuses with congenital heart disease (CHD) using a newly developed prenatal Medical-Aristotle-Personal and eXtracardiac score (P-MAP-X-Score) of complexity- and impairment

Freund M.W. (1), Pistorius L. (2), Heide H. ter (1), Stoutenbeek P. (2), Strengers J. (1)

Department of Pediatric Cardiology (1), Department of Obstetrics (2), Wilhelmina Children's Hospital/University Medical Center, Utrecht, The Netherlands

Introduction: Prenatal diagnostic of congenital heart defects (CHD) has further implications and demands prenatal counselling. The implications of the CHD and extracardiac defects have to be defined. A newly developed prenatal Medical-Aristotle-Personal and eXtracardiac score (P-MAP-X-score) which consists of four items, 1. medical cardiac impairment, 2. surgical complexity level, 3. physical, neurological, social impairment, and 4. extracardiac impairment, describes and predicts the overall cardiac and extracardiac situation.

Methods: Between 2004 and 2007 168 foetuses were diagnosed with a CHD. Based on the pre- and postnatal diagnosis we separately defined the cardiac and extracardiac situation using the P-MAP-X-score. The results were summarised and the variance based on pre- and postnatal diagnosis was determined.

Results: All patients could be classified using the P-MAP-X-score. In the prenatally diagnosed foetuses the medical cardiac impairment-score revealed in 25.6% a non-complex CHD, in 39.9% a complex CHD with the option of biventricular repair, in 23.8% a very complex CHD with the option of univentriculair repair or transplantation and in 10.7% a CHD without therapeutic options.

In 147/168 patients surgical interventions were predicted to be indicated postnatally. A total of 251 operations should be performed to reach the predicted goal. The mean surgical complexity level was 2.99 (out of 4).

In 41.1 % no physical impairment, in 17.5% no potential risk for neurological impairment, in all but two a social impairment later in life was predicted.

The pre-postnatal correspondence of the diagnosis was 72%. In 22.7% there were major differences which changed the leading diagnosis or its significance, and in 5.3% the diagnosis was wrong.

The pre-postnatal correspondence of the medical cardiac impairment score was 87%, of the surgical complexity level was 81.3%, and of the personal complexity score > 84.5%. Extracardiac anomalies were present in 49.4% and pre-postnatal correspondence was 86.2%.

Conclusion: A new P-MAP-X-score gives a description of the complexity and impairment of the prenatal diagnosed CHD. It transforms the diagnoses into categories. Even if a postnatal diagnosis is inconsistent, the correspondence of the predicted P-MAP-X-score is still high and will provide reliable and adequate prenatal counselling. Further prospective studies are required.

P-96

Normal values of isovolumetric contraction and relaxation time in fetus

Dangel J. (1), Hamela -Olkowska A. (2)

Perinatal cardiology Clinic 2nd Department of Obstetrics and GynecologyMedical University of Warsaw

Objective: To establish gestational age-specific values of isovolumetric contraction (ICT) and relaxation time (IRT) by pulsed Doppler in healthy singleton fetuses.

Methods: Examinations were performed using Sequoia 512 in a group of 140 healthy singleton fetuses at 18 to 40 weeks of gestation. ICT and RCT were measured by pulsed Doppler from left ventricular inflow/outflow view using convex 3,5-6 MHz probe. ICT was measured from mitral valve closure to aortic valve opening. RCT was measured from aortic valve closure to mitral valve opening.

Results: There was a slight correlation between ICT, RCT and gestational age, but is was no statistically significant (Table). No correlation was found between FHR and ICT or RCT.

Weeks	18-22	23-28	29-34	35-40	
No. of cases	36	35	35	34	p
ICT (ms)	35 ± 6	37 ± 6	38 ± 8	39 ± 9	NS
IRT (ms)	40 ± 6	40 ± 7	44 ± 6	45 ± 8	NS

NS - non-statistical

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Conclusion: The isovolumetric contraction and relaxation time is constant with gestation and fetal heart rate in normal pregnancy. Normal values may be useful for comparison with complicated pregnancy.

P-97

Frequency of and causes of ventricular disproportion in fetal echocardiograms

Clur S.A. (1,2), Ottenkamp J. (1,2), Bilardo C.M. (3)
Department of Pediatric Cardiology Emma Children's Hospital Academic Medical Centre, Amsterdam, The Netherlands (1), Center of Congenital Heart Disease Amsterdam-Leiden, The Netherlands, (2), Department of Obstetrics and Gynaecology Academic Medical Centre, Amsterdam, The Netherlands (3)

Introduction: Ventricular disproportion (VD) is not infrequently seen on fetal echocardiograms. Coarctation of the aorta and total anomalous pulmonary venous drainage (TAPVD) are commonly mentioned causes.
Objectives: To document the incidence and causes of fetal disproportion in our institution.
Methods: From April 1999 until end 2007 519 consecutive fetuses were referred to the pediatric cardiologist for fetal echocardiography with standard indications after preliminary screening by the gynaecologist.
Results: In 105 (20%)VD was identified, due to a relatively enlarged right ventricle in 83 (79%) and a relatively enlarged left ventricle in 22 (21%).VD was due to: Double Outlet Right Ventricle (+/- Transposition of the Great Arteries) (20), Hypoplastic Left Heart Syndrome (18), abnormalities of the tricuspid valve (Ebstein's, regurgitation, dysplasia, atresia) (16), unbalanced Atrio-ventricular Septal Defect (16), coarctation of the aorta (+/-Ventricular Septal Defect (VSD)) (6), abnormalities of the pulmonary valve (stenosis, atresia, absence) (5), Double Inlet Left Ventricle(3), VSD (+/- Atrial Septal Defect (ASD)) (5), Tetralogy of Fallot(3), ASD (2), Congestive Cardiac Failure (2), vena cava superior sinistra (VCSS) without VCS Dextra (1). In 8 the heart was normal postnatally. In 13 patients a VCSS was present (12.4%). There were no cases of isolated TAPVD.
Conclusions: VD is commonly encountered in fetal echocardiography and is predominantly due to left heart hypoplasia or right heart enlargement.VD may be secondary to a variety of causes. A VCSS draining to the coronary sinus is a commonly associated finding and may have a role in the pathogenesis of the VD by potentially reducing the amount of fetal blood flow over the mitral valve. When VD is present congenital heart disease should be actively excluded and the presence of a VCSS excluded.

P-98

Longterm results and prognosis after surgical treatment of valvular aortic stenosis in children - a population based study

Mattila I.P., Nieminen H.P., Sairanen H.I.
Department of thoracic and cardiovascular surgery, Hospital for Children and Adolescent, Helsinki University Central Hospital, Helsinki, Finland

Introduction: There is no population based information about the late outcome after surgery for aortic valve stenosis in children.
Methods: We evaluated all the patients whose first operation for aortic valve stenosis was performed before the age of 18 in Finland between 1953–1999.
Results: There were 233 patients with valvular aortic stenosis. Additional heart defects were coarctation (13%), subaortic stenosis (10%), aortic insufficiency (9%), mitral valve disease (4%) and fibroelastosis (4%).Total operative mortality including also reoperations was 8.6% (20/233 patients). Early mortality was highest in the neonatal group (31/37, 35%), followed by 16% in the 1–12 mo group (5/31). 1–15 years of age group consisted of 109 patients(1 died initially, 1%) and 56 patients were operated between15–18 years of age(1 death, 2%).
Late survival after succesful first operation was 86% in the neonatal group (n = 24, for 15 years follow up), 96 % (n = 26), 93% (n = 108) and 86% (n = 55) for 20 years follow up respectively. Freedom from reoperations 20 years after the first operation was 34% for those initally operated < 1 years of age, 68% for 1–15 y and 80% for the oldest group. Compared to normal population late survival was 10% lower at 20 years for the whole study group. Relative survival for those patients treated in 1990–1999 (119) was 10% better compared to those treated in 1953-1989 (114) 10 years after the initial succesful operation.
Conclusion: Aortic valve stenosis carries high mortality when presented for surgery before the age of one month. However late prognosis for the whole group is relatively good compared to normal population.

P-99

Structural Abnormalities of Pulmonary Trunk in Tetralogy of Fallot

Bedard E., McCarthy K.P., Gatzoulis M.A., Ho S.Y.
Royal Brompton Hospital, London, UK

Introduction: Little is known about pulmonary trunk (PT) histology in patients with tetralogy of Fallot (TOF), especially in the current era of reducing age at primary repair. We hypothesized that intrinsic elastic histological abnormalities of the PT are present early in life and that palliative surgery and total repair affect these changes.
Methods: PT histology of 18 cases with TOF (9 with pulmonary stenosis and 9 with pulmonary atresia) from our cardiac morphology archives were studied by light microscopy and compared with 10 normal controls. Elastic tissue configuration (ETC) of the PT was classified according to Heath et al.[1] (J Pathol Bacteriol, 1959), and collagen arrangement was examined. PT to aortic media thickness (MT) ratio was also calculated.
Results: In the TOF group (1 foetus, 4 infants, 5 children, and 8 adults), 4 patients had previous palliative surgery, and 6 had total repair (at median age 6 years; range 2.5–18 years). PT ETC in normal controls was as expected for age (< 6 months, aortic-like; 6 to 24 months, transitional; > 24 months, adult pulmonary; Table 1). Normal PT histological features were found in the youngest TOF specimens (foetus to 3 months of age). Majority of TOF cases aged ≥ 1 year had significant histological abnormalities, including patients with previous palliative surgery or total repair. Two of the 6 patients who had total repair 9 and 15 years prior to death had normal ETC. PT to aortic MT ratio was lower for TOF patients aged > 2 years (median 0.29, range 0.19-1.00) compared to normal controls (median 0.61, range 0.28-0.91, p = 0.04), especially in patients who had total repair (median 0.27, range 0.19-0.38, p = 0.004).
Conclusions: Remarkable intrinsic histological abnormalities are present from age≥1 year in the PT of patients with TOF, even after previous palliative surgery or total repair. "Normal" ETC was found in only 2 TOF cases after total repair. The repaired patients in this cohort had surgery relatively late (age ≥ 2.5 years). These data provide support for early primary repair of TOF although further characterization of PT tissue is needed and underway.

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Table 1. Elastic tissue configuration – TOF patients and normal controls

Elastic tissue configuration			Age distribution and surgical characteristics				
Heath et al.[1] classification	TOF (n)	Controls (n)		TOF (n)			Controls (n)
				Unoperated	Palliated	Repaired	
Aortic-like	8	2	Age Range	30w gest – 30y			1d
			≤2y	4	1	-	2
			>2y	1	1	1	0
Transitional	4	1	Age Range	4y – 18y			12mo
			≤2y	-	-	-	1
			>2y	1	1	2	0
Adult pulmonary	3	7	Age Range	5y – 33y			22-70y
			>2y	1	-	2	7
Hypotensive[c1]	3	0	Age Range	20y – 36y			-
			>2y	0	1	2	0

d = day; mo = months; y = years

P-100
Coagulation parameters and platelet function in whole blood samples of adults with cyanotic congenital cardiac disease

Braun S., Mebus S., Eicken A., Hager A., Hess J., Kaemmerer H.
Deutsches Herzzentrum München

Objectives: In the natural course patients with cyanotic congenital cardiac disease (CCCD) tend to develop both thromboembolic events as well as bleeding complications. The bleeding tendency in CCHD-patients may be related to reduced platelet count and function and additional defects in the coagulation system. Thrombelastometry measures the velocity of clotting as well as the firmness of the clot and has been shown to be suitable for the detection of hypercoagulability. The aim of our study was to assess the usefulness of measuring coagulation and platelet function in whole blood samples of CCCD-patients by recently developed instruments for thrombelastometry and platelet aggregometry.

Methods: We studied 34 consecutive patients presenting in the outpatient clinic of our department for congenital cardiac disease, 25 of them had pulmonary hypertension. One patient took acetylsalicylic acid, 5 had oral anticoagulation therapy with vitamin K antagonists and 7 patients received sildenafil. Thrombelastometry was performed in citrated whole blood using the ROTEM™ instrument. Whole blood impedance platelet aggregometry was measured with the Multiplate™ system. Blood cells were counted on a Sysmex XE2100 analyser.

Results: The median value of hematocrit (Hct) was 56% (range 43–78%). Negative correlations were found between hematocrit and platelet count (correlation coefficient r = −0.5898), maximum clot firmness (MCF) in thrombelastometric analysis activated with tissue thromboplastin (EXTEM) (r = −0.5961), alpha angle (r = −0.8100) and platelet aggregation after activation with ADP (r = −0.5267), arachidonic acid (r = −0.6584) and TRAP (r = −0.4624). The median MCF value in the FIBTEM test, which shows the fibrinogen component of blood coagulation after inhibition of platelet function in-vitro was 8.5 mm. This means that 50% of all patients were below the FIBTEM reference range, in the group with high hematocrit the value was even lower (Table).

Conclusions: Our findings using these two new instruments for whole blood coagulation and platelet function analysis are consistent with publications showing thrombocytopenia and suppressed platelet function in CCCD patients. Thrombometry showed that there is a tendency for reduced clot formation dynamics (alpha angle in EXTEM) and decreased fibrinogen or disturbed clot polymerization (small MCF in FIBTEM), but we found no tendency for hypercoagulability. This may be relevant for therapeutic decisions concerning anticoagulation or antiplatelet therapy in CCCD patients.

	Median [25. and 75.percentile] (Hct <56%)	Median [25. and 75.percentile] (Hct ≥56%)	Reference range
Platelet count [/nl]	192 [160–249]	146 [76–188]	200–400
ROTEM EXTEM MCF [mm]	56 [51–61]	52.5 [40–58]	50–72
ROTEM EXTEM alpha angle [°]	67 [65–72]	56 [48–64]	63–83
ROTEM FIBTEM MCF [mm]	10 [6.25–17.25]	8 [5–12.25]	9–25
Multiplate ADP-ind. Aggregation [AU*min]	501 [337–664]	306 [279 – 565]	534–1220
Multiplate A.a.- ind. Aggregation [AU*min]	753 [251–935]	513 [276–600]	745– 361

P-101
Infective endocarditis in adults with congenital heart disease

Di Filippo S., Ninet J., Sassolas F., Henaine R., Ducreux C., Ruffin C., Bozio A.
Hôpital Cardiologique, Pediatric Cardiology, Lyon, France

The aim of this retrospective study was to describe features ofinfective endocarditis (IE) in adults with congenital heart disease (CHD).

Methods: The records of all episodes of IE diagnosed from 1974 to 2006, in patients with CHD and more than 18 years of age at diagnosis, were retrospectively reviewed.

Results: Fourty-four episodes of IE occurred, 36 after 1990 (81.8%), 28 males (63.6%). Age at diagnosis was 30.3 ± 9years (median 28years). Ten were recurrent episodes (22.7%). CHD was previously repared in 15 cases (34%), palliated in 7 (16%) and non-operated in 22 (50%). Dental causes were predominant (34%), followed by cutaneous causes (25%); others were postoperative (4.5%), miscellaneous (7%) or unknown causes (29.5%). A microbial agent as identified in 95.4% of the cases: oral streptococcus and staphylococcus aureus were the leading causative agents (respectively 41% and 36%). Left heart locations were predominant (75%). Severe clinical cardiac complication occurred in 10 cases (23%), an echocardiographic complication in 18 (40%). Twenty-four patients experienced embolic events (54.5%) ; early surgical treatment was required in 25% of the cases. Three patients died due to IE (6.8%). Antibiotic prophylaxis had been neglicted despite known risk in 41% of the cases.

Conclusion: E is an ongoing life-threatening complication in adults with CHD, with significant morbidity. Faced the high rate of prophylaxis negligence and recurrent episodes, antibiotic prophylaxis should be emphasized.

P-102
Haemostatic Capacity of Eisenmengerís syndrom examined by ThrombelastographyÆ

Jensen A.S. (2), Stensballe J. (1), Johansson P.I. (1), Sondergaard L. (2)
Department of Clinical Immunology (1), Department of cardiology (2), Rigshospitalet, Copenhagen University Hospital, Denmark

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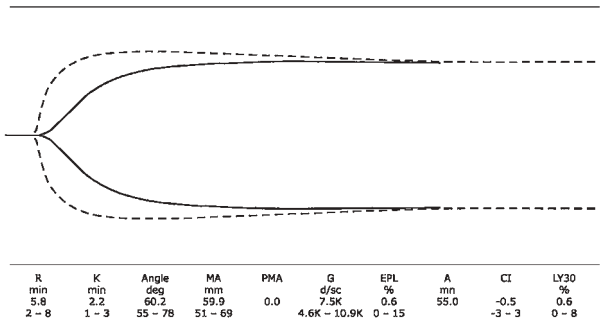
Background: Eisenmenger's syndrome is associated with an increased risk of both thrombo-embolic events and bleeding. The thrombopathogenesis is believed to be due to hyperviscosity, whereas the cause of bleeding is more uncertain. Small studies have shown different coagulation abnormalities such as thrombocytopenia, thrombasthenia and vascular impairment (1). The aim of this study was to examine hemostatic capacity using Thrombelastography (TEG®) in patients with Eisenmenger's syndrome.
Methods: Eighteen patients with Eisenmenger's syndrome where examined at out patient visits. TEG was performed using a coagulation analyzer (5000 series TEG analyzer, Haemoscope Corporation, Niles IL, USA), which measures clot formation and degradation in whole blood. Several variables are measured, the most important being R, Angle, and maximal amplitude MA. The parameter R represents the initiation of the coagulation process, Angle represents the clot build up involving fibrinogen function, and MA maximal clot strength mainly dependent on platelet function.
Results: We studied a cohort of predominantly females (n = 12/18, 67%) without active bleeding. None of the patients had a hypercoagulable TEG® profile. Only 22% (4/18) of the patients had a completely normal TEG®. Of the 78% (14/18) abnormal TEG®, 64% (9/14) had significant changes in their TEG® profile indicating a hypocoagulable state. By analysing the abnormal TEG® profiles we found that 33% (3/9) had prolonged R time, 33% (3/9) had reduced MA and 100% (9/9) had reduced Angle. Since an abnormal R and/or MA can interfere with the value of the Angle, we looked at how many had only affected Angle. That was the case for 56% (5/9) of the patients.
Conclusion: Interestingly, in this study TEG®-profiles show that patients with Eisenmenger's syndrome tends to be hypocoagulable and not hypercoagulable, which is generally accepted due to the high incidence of thrombo-embolic events. In general the TEG® revealed normal platelet function. Instead, the hypocoagulable profile seemed more related to an impaired fibrinogen function or a reduced enzymatic initiation of the coagulation. The reason for the variability in fibrogen function and enzymatic coagulation initiation is uncertain, but warrants further study.



P-103

Morbidity after pediatric cardiac surgery assessed with usage of medicines. A population-based registry study

Nieminen H.P. (1), Sairanen H.I. (1), Jokinen E.V. (2)
Department of Surgery (1) and Cardiology (2), Hospital for Children and Adolescents, Helsinki University Central Hospital, Helsinki, Finland

Introduction: The overall morbidity of the patients who had been operated on for congenital heart defect (CHD) is comprehensively unexamined. The medication of the patients can be used as an indirect indicator of the morbidity.
Methods: The data from the Finnish Research Registry of Pediatric Cardiac surgery was linked to the data of the medication registry of the Social Insurance Institution. The present study includes the medication data of all 5116 patients (mean age 33.5, range 14.7–64.8) who had been operated on for CHD during childhood, and lived in Finland in 23rd of August 2004. The use of medicines among patients was compared with age and sex matched control population (n = 10232).
Results: The overall use of medicines was frequent among patients; 62% of patients and 53% of controls had bought at least one prescribed medicine (RR 1.2, Cl 1.1–1.2). As expected, the number of patients using cardiovascular medicines (17%) and antithrombotic agents (5%) was high when compared to control subjects (RR 2.2 and 8.4, respectively). In addition, the patients needed medicinal care for epilepsy (3%), obstructive airway diseases (7%), psychiatric diseases (10%) and hormone deficiencies (2.5%) more often than control subjects (RR 2.2, 1.5, 1.3 and 1.5, respectively). Asthma and epilepsy were especially common among patients operated neonatally.
Conclusion: Patients had more chronic diseases and used more medicines than controls. The patients operated as neonates and patients with mental retardation had most often heavy burden of medicines.

P-104

Exercise capacity in young people long term after surgical correction of d-transposition of great arteries by Senning atrial switch

Pietrucha A.Z. (1), Pietrucha B.J. (2), Gackowski A. (1), El-Massri N. (1), Mroczek – Czernecka D. (1), Rudzinski A. (2), Piwowarska W. (1)
Medical School of Jagiellonian Univers Coronary Disease Department Cracow, Poland (1), Children Cardiology Department University Children Hospital CM UJ Cracow, Poland (2)

The aim of study: evaluation of physical efficiency and function of circulatory system in young people, long term after surgical correction of d-transposition of great arteries by Senning atrial switch.
We observed 16 pts (14 men, 2 women) at age 21–25 (mean 22.1) after Senning procedures of transposition of great arteries. Senning procedures were performed within 3–7 months of life.
All pts underwent 12-lead standard ECG, 24-hour ECG Holter monitoring, echocardiography examination, cardiac MRI and cardiopulmonary exercise treadmill test (ETT). Left and right ventricle function, ETT duration and workload, and max oxygen uptake (VO2max) and anaerobic threshold (AT) were evaluated. VO2max, ETT work were also expressed as % of predicted values. Serum level of NT-proBNP was also assessed in all pts.
Results: Episodic or sustain bradycardia was observed in all pts, 5 pts had an escape nodal rhythm, 2 pts had registered asymptomatic tachycardia-bradycardia syndrome. There were no RR pauses longer than 2,5 s.
Left ventricle function was not reduced in all pts (LV diastolic diameter 46 ± 4.2; LV EF 68 ± 14.7%). Right ventricle dilatation RV 38.2 ± mm with moderately reduced ejection fraction (RV EF 47 ± 13,9%) was observed in pts after TGA correction. Cardiac MRI discovered significant RV dilatation with hypertorophy of right ventricle walls with magnified trabeculation.
Exercise was well tolerated by all pts. – they achieved about 100 % of predicted workload (207.6 ± 65.3 W; 12 ± 2.5 METS). Value of VO2max was decreased in all pts. [24.3 ± 9.1 ml/kg/min] - about 42.8% of predicted values. The level of AT was decreased in comparison to predicted values (0.97 ± 0.12 l/min; 26.8 ± 9.6 %VO2max). Serum level of NTproBNP was moderately increased (328 ± 127,8 pg/ml). These results indicate mild to moderate reduction of exercise capacity.

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Conclusions:
• Young people after surgical treatment of d-transposition of great arteries by Senning atrial switch revealed good exercise toleration.
• In spite of good exercise toleration, mild to moderate reduction of exercise capacity and function of circulatory system was observed in these patients.

P-105
Benefit of beta-blockers on systemic ventricle function and exercise capacity in patients with systemic right ventricle or single ventricle
Godart F. (1), Defrance C. (1), Lions C. (2), Hossein-Foucher C. (3), Francart C. (1), Delhaye C. (1), Rey C. (1)
Paediatric Cardiology, Lille, France (1); Radiology - Lille, France (2); Nuclear Department, Lille, France (3)

Objective: To evaluate efficiency of beta-blockers on systemic ventricle function, exercise performance and functional status in patients with systemic right ventricle (SRV) or single ventricle (SV).
Methods and Results: 16 patients (12 SRV and 4 SV) with systemic ventricle dysfunction underwent clinical evaluation (NYHA class), exercise testing with metabolic gas exchange, BNP plasma level determination, and evaluation of systemic ventricle function by Magnetic Resonance Imaging (MRI), Radionuclide Ventriculography (ERV) and echocardiography (TTE), both before and after a 6-months-minimum follow-up period with beta-blockers.
Beta-blocking drugs were well tolerated and no major adverse event occured.
At baseline all patients had systemic ventricle dysfunction and diminished exercise tolerance. Under beta-blocker therapy there was a significant improvement in NYHA class ($p = 0.034$). Peak oxygen uptake was significantly increased in patients with RSV ($p = 0.012$). Systemic ventricle ejection fraction (EF) was significantly improved: from 28% to 32% by MRI ($p = 0.007$), from 34% to 41% by TTE ($p = 0.01$), from 39% to 46% by ERV ($p = 0.23$), and from 34% to 40% by "mean EF" ($p = 0.006$). BNP level remained unchanged until the end of the study ($p = 0.31$). Patients with exercise chronotropic insufficiency (peak exercise heart rate < 65% of theoretical value) had significantly lower peak oxygen uptake ($p = 0.018$) and shorter duration of exercise ($p < 0.05$) than people without chronotropic insufficiency.
Conclusion: Beta-blockers significantly improve functional status (NYHA class), exercise performance and systemic ventricle function in patients with SRV or SV.

P-106
Long-term cardiopulmonary exercise capacity after Fontan operation
Ovrutskiy S., Hauberg I., Ewert P., Miera O., Alexi-Meskishvili V., Peters B, Hetzer R., Berger F.
German Heart Institute Berlin, Germany

Objective: After successful Fontan operation in strictly selected patients physical capacity mostly improves. It was postulated that early separation of the blood circuit enhances the long-term success of Fontan hemodynamics. To test this hypothesis we analyzed the short-term vs. long-term postoperative cardiopulmonary exercise capacity in children and adults.
Methods: Spiroergometry was performed at least twice in 47 patients with the total follow-up of 3.1 (0.7–5.2 years) for early and 7.2 (3.9–14.5) years for late testing. The median age at the last testing was 14.5 (range 7–45) years. 29 patients had been operated on in childhood (median 4.5 range 1.5–14 years) and 18 as adults (16–37 years). The exercise capacity (W/kgmax) and oxygen consumption capacity (VO2max) and heart rate at peak (HRmax) were compared between children and adults.
Results: The VO2max and W/kgmax in children compared to adults was slightly better early postoperatively (median 26.6 vs. 22.9 ml/kg/min, bzw. 2.2 vs. 1.8 w/kg, n.s.) and both VO2max (median 27.5 vs. 17.2 ml/kg/min, $p < 0{,}001$) and W/kgmax were significantly better late after surgery (median 2.2 vs. 1.4 w/kg, $p < 0.001$) The HRmax was at median 135 b/min as well early as late postoperatively, but was in trend lower in adults on the late testing compare to children (119 b/min vs. 147 b/min, $p = 0.06$). There was a high correlation between HRmax and VO2max late postoperatively ($r = 0.69$, $p < 0.001$). In the patient group as a whole, there was a significant decrease of VO2max between the early and later testing (median 26.5 vs. 20.6 ml/kg/min, $p < 0.001$).
Conclusions: Exercise capacity and oxygen consumption decrease continuously in all age groups in Fontan patients, while surgical palliation in early childhood results in better cardiopulmonary capacity during long-term follow-up. The chronotrope incompetence may play a negative roll in this capacity decrease. Regular surveillance of the physical capacity by spiroergometry is indispensable for the supervision of patients with Fontan hemodynamics.

P-107
Rate responsive pacing does not improve right ventricular hemodynamics nor exercise capacity in adults with a systemic right ventricle.
Uebing A. (1,2), Diller G.P. (1), Li W. (1), Maskell M. (1), Gatzoulis M.A. (1)
Adult Congenital Heart Centre and Centre for Pulmonary Hypertension, Royal Brompton Hospital and the National Heart and Lung Institute at Imperial College, London, United Kingdom (1); Department of Paediatric Cardiology, University Hospital of Schleswig-Holstein, Campus Kiel, Kiel, Germany (2)

Background: Inappropriate heart rate response to exercise (chronotropic incompetence) and exercise intolerance are common in patients with a systemic right ventricle (SRV). We aimed to assess the relationship between heart rate increase, oxygen consumption (VO2) and the timing of the right ventricular cardiac cycle in this cohort.
Methods: We prospectively studied 9 patients with SRV and pre-existing pacemakers using treadmill exercise testing and Doppler echocardiography. Exercise tests were performed with baseline pacemaker settings and with optimized heart rate response in a random order. Echocardiography was performed at increasing heart rates. In addition, 8 age and gender matched control subjects underwent exercise testing using a similar exercise protocol.
Results: Patients with SRV had significantly lower peak VO2 compared to controls (12.6 ± 6.8 vs. 31.4 ± 6.6 m/kg/min, $P = 0.0006$) at baseline and active pacemaker reprogramming failed to increase VO2 in this cohort (12.6 ± 6.8 vs. 12.4 ± 4.9 ml/kg/min, P = NS at baseline and with active reprogramming, respectively). A marked increase in total isovolumic time, and a significant reduction in total filling time and the aortic velocity time integral ($P < 0.001$ for all) was found at higher heart rates compared to baseline conditions.
Conclusion: This study suggests that despite chronotropic incompetence at baseline rate responsive pacing does not improve exercise capacity in patients with SRV. It further indicates that

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high heart rates may be detrimental in these patients by reducing diastolic filling and stroke volume. These findings may have clinical implications when considering implantation of a permanent pacemaker in this cohort.

P-108

Ebstein`s anomaly: modified reconstruction of tricuspid valve without ventricle plication and further developments

Nagdyman N. (1), Hetzer R. (2), Ewert P. (1), Komoda T. (2), Berger F. (1)
Department of Congenital Heart Defects and Pediatric Cardiology (1); Department of Cardiothoracic and Vascular Surgery (2); Deutsches Herzzentrum Berlin, Germany

Introduction: Ebstein's anomaly (EA) is a rare congenital heart defect in which the hinges of the septal and/or posterior leaflets are displaced downward to the right ventricle. The anterior leaflet is usually not displaced but is enlarged and sail-like and valve closure is likewise displaced downwards.
Methods: Since 1988 we have operated 42 patients [30 f, 12 m; median age 26 (2–55)] years with EA using a modified repair technique of the tricuspid valve. This technique reconstructures the valve mechanism at the level of the true annulus by using the most mobile leaflet for valve closure without plication of the atrialized chamber. In 12 pts a mattress suture is placed between the body of the anterior leaflet and the ventricular septum on the opposite site and a double orifice valve was reconstructed.
We evaluated our long-term results with regard to functional capacity (NYHA functional class), tricus pid valve function, mortality and re-operation rate. We quantified the right ventricular function by measuring flow velocity integral of the pulmonary artery (VTI_{PA}).
Results: All patients survived the operation. Early mortality was 7.1% and the late mortality was 2.4%. All deceased pts. were older 50 years and at least NYHA III or IV. Since 2004 no patient died. The mean follow-up period was 64 months. So far one re-operation has been necessary.
NYHA class improved significant from 3.1 to 1.8 ($p < 0.001$). Echocardiographic studies demonstrated a significant improvement of tricuspid valve insufficiency from 3.2 to 1.9 ($p < 0.001$). No tricuspid valve stenosis was observed. Significant improvement of VTI_{PA} was observed with stable heart frequency ($p = 0.01$).
Conclusions: We conclude that reconstruction of the tricuspid valve without ventricle plication achieves good functional results. The modifications and the further developments seem to underline these results. The operation in older patients with impaired functional capacity seem to contain a higher mortality risk.

P-109

Therapeutic management for hypertension and improvement of exercise tolerance in patients after surgical repair of coarctation of the aorta

Giordano U. (1), Giannico S. (2), Cifra B. (3), Meta R. (3), Turchetta A. (1), Calzolari A. (1)
Cardiorespiratory and Sports Medicine Unit, Department of Pediatrics - Bambino Gesù Children's Hospital; Rome, Italy (1); Department of Cardiology, Bambino Gesù Children's Hospital; Rome, Italy (2); Tor Vergata University - Rome, Italy (3)

Introduction: We sought to analyze a group of patients (P) operated upon for coarctation of the aorta in early childhood to examine the presence of hypertension during exercise testing (ET) and 24-h ABPM, the efficacy of a new therapeutic approach, if needed, based on Candesartan cilexetil (C) or Atenolol (A), respectively in those patients with a hypertensive response during exercise with a reduced increment of heart rate (HR) and in those with a hypertensive response with sinus tachycardia.
Methods: 46 P (17 female, 29 male, mean age of 14.9 ± 3.3 yrs) were studied to examine blood pressure (BP) behavior and to observe the results of therapy to achieve a normotensive status. Selection criteria: absence of associated cardiac abnormalities (with the exeption of aortic bicuspid valve), no significant arm-leg gradient at rest (< 15 mmHg). P were evaluated by BP at rest, during ET on cycle ergometer (James protocol) and 24-h ambulatory blood pressure monitoring (ABPM).
Results: 21/46 P (45%) were normotensive; 25/46 P (55%) need antihypertensive therapy (AT) to control exercise systolic (S) BP and/or ABPM. 13/25 were treated with C and 12/25 with A. Optimal BP control was achieved in 12/13 (93%) treated with C and in 6/12 (50%) treated with A. No patient on C had side-effects while three treated with A needed to modify therapy (two for excessive bradycardia and one for dizziness while at rest). P on C had a significant better exercise tolerance at similar SBP values (in terms of time of exercise and Watt) and maintained nighttime SBP fall when compared to P on A

GROUP	TIME EXERCISE		HR MAX (beats/min)	SBP MAX (mmHg)	SBP DAY (mmHg)	SBP NIGHT (mmHg)
	minutes	Watt				
C group	9.4 ± 1.4	153 ± 15	169 ± 14	196 ± 20	123 ± 10	111 ± 7
A group	7.6 ± 2	129 ± 22	135 ± 18	178 ± 27	126 ± 4	117 ± 5
t-test	0.01	0.004	< 0.001	n.s.	n.s.	0.02

Conclusion: AT for patients after coarctation repair should be based on the results of SBP and HR during exercise and ABPM.

P-110

Prospective follow-up study of children with Univentricular heart: Neurodevelopmental outcome at age 12 months

Sarajuuri A. (1), Lönnqvist T. (1), Mildh L. (2), Rajantie I. (1), Eronen M. (3), Mattila I. (4), Jokinen E. (3)
Division of Neurology (1), Anesthesiology and Intensive Care Medicine (2), Paediatric Cardiology (3) and Surgery (4), Department of Paediatric and Adolescent Medicine, Helsinki University Central Hospital, Helsinki, Finland

Objectives: Despite recent advances in the treatment of children with univentricular heart, their neurodevelopmental outcome remains a major concern deserving study.
Methods: This prospective follow-up study evaluated the neurodevelopmental outcome of 23 patients with hypoplastic left heart, 14 with other forms of univentricular heart, and 46 healthy controls at a median age of 12.2 months. The Griffiths developmental scale and Atlanta Infant Movement Scale served for developmental evaluation.
Results: The mean Griffiths Developmental Quotient of children with hypoplastic left heart was significantly lower (91.6) than in control children (DQ 106.8, $p < 0.001$). Patients with univentricular heart scored significantly lower than controls only in the gross motor domain ($p = 0.001$), not in overall development (DQ 100.6, p = non-significant). Atlanta Infant Motor Scale scores were significantly lower in children with hypoplastic left heart (37.5, $p < 0.001$) and with univentricular heart (43.5, $p = 0.011$) than in controls (53.3). In linear regression, a diagnosis of hypoplastic left heart ($p = 0.016$), a clinical seizure history ($p = 0.002$), and the

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highest plasma lactate level after the bidirectional Glenn operation ($p = 0.045$) were significantly associated with the Developmental Quotient.
Conclusions: At age 1 year, the level of development of children with univentricular heart was significantly lower than for controls only in motor skills, whereas children with hypoplastic left heart had a more widespread developmental delay. The diagnosis, a clinical seizure history and elevated plasma lactate levels following bidirectional Glenn operation emerged as risk factors.

P-111
Plasma levels of B-type natriuretic peptide after surgical repair of tetralogy of Fallot
Koch A., Zink S., Glöckler M., Dittrich S.
Paediatric Cardiology, Hospital for Children and Adolescents, University of Erlangen-Nürnberg, Germany

Objective: B-type natriuretic peptide (BNP) is an established diagnostic marker in congestive heart failure and left ventricular dysfunction. Recent reports suggest a role for BNP to detect right ventricular (RV) dysfunction, too. The aim of this study was to evaluate the diagnostic validity of plasma BNP on patients with surgically repaired tetralogy of Fallot.
Methods: From 2002 to 2007, plasma BNP concentration was measured (Triage BNP assay, Biosite®) in all patients who came to our outpatient clinic for follow-up or diagnostic work-up including venous puncture. 131 patients with surgically repaired tetralogy of Fallot (78 males, age 16.1+7.1 years) were analysed retrospectively. BNP levels were compared with age and gender-specific normal values, with clinical, electrocardiographic, and echocardiographic data, and with results of exercise testing.
Results: Plasma BNP levels were between 5 and 196 pg/ml (median BNP 26 pg/ml, interquartile range [IQR] 1–48 pg/ml) with slightly higher values in females (median 37 [IQR 21-57] vs. 16 [IQR 11–30] pg/ml) comparable to the sex related difference in healthy adolescents. According to age and gender 60% of the values were increased. BNP was significantly correlated with RV dilatation (ratio right to left ventricular diameter, $r = 0.29$, $p = 0.005$), severity of tricuspid regurgitation ($r = 0.25$, $p = 0.001$), and severity of pulmonary regurgitation ($r = 0.19$, $p = 0.01$). There was no correlation to age, maximum exercise capability, QRS duration, RV pressure, or RV outflow tract obstruction. Pulmonary valve replacement was performed in 14 patients during follow-up. BNP was significantly higher in this group of patients (72 [27–124] vs. 23 [12–39] pg/ml, $p = 0.001$), but decreased within 4–30 months after valve replacement from 64 [40–113] to 33 [21–53] pg/ml in all reassessed patients ($p = 0.04$).
Conclusion: In the majority of patients with surgically repaired tetralogy of Fallot BNP plasma concentration was slightly increased. There was a significant correlation between BNP and parameters reflecting RV volume load. However, there was a considerable overlap between patients with different severity of valve insufficiency or RV dilatation. No correlation was found between BNP and RV pressure load. Therefore, elevated or increasing plasma BNP levels can indicate RV volume load but a single normal BNP level does not exclude RV dilatation.

P-112
Evaluation of Right Ventricular Functions and It's Relation with Pulmonary Regurgitation and B-type Natriuretic Peptide Levels by Using Tissue Doppler Imaging in Patients After Repair of Tetralogy of Fallot
Çetin İ. (1), Tokel K. (1), Varan B. (1), Örün U.A. (1), Gökdemir M. (1), Cıýndık N. (1), Aþlamacı S. (2)
Baskent University, Pediatric Cardiology, Ankara, Turkey (1); Baskent University, Cardiovascular Surgery, Ankara, Turkey (2)

Objectives: The aim of this study was to assess the relation between plasma B-type natriuretic peptide (BNP) levels and global right ventricular functions evaluated by tissue Doppler imaging (TDI) in patients after repair of tetralogy of Fallot (ToF).
Methods: Twenty five patients with a mean age of 14.1 ± 4.4 years who underwent repair of ToF at a mean age of 4.9 ± 5.1 years and 29 age and sex matched healthy children at a mean age of 13.1 ± 2.8 years enrolled in this study. Plasma BNP levels were measured at rest. The right ventricle was assessed by two-dimensional echocardiography and color Doppler. TDI was performed at rest on the right ventricular basal segments to determine the myocardial performance index (MPI) by using right ventricular time intervals and myocardial velocities.
Results: Plasma BNP levels were significantly higher in patients than in controls (28.3 ± 24.1 vs. 7.4 ± 2.3 pg/ml, $p = 0.0001$). The right ventricular end-diastolic volume (RVEDV) (77.5 ± 15.2 vs. 39.1 ± 7.4 ml, $p = 0.0001$) and diameter (RVEDD) (43.1 ± 4.9 vs. 32.8 ± 3.4 mm, $p = 0.0001$) were significantly increased, right ventricular basal segments MPI (1.08 ± 0.35 vs. 0.58 ± 0.11, $p = 0.0001$) was higher and isovolumic acceleration (IVA) (3.1 ± 0.7 vs. 5.4 ± 1.0 m/s2, $p = 0.0001$) was lower in patients. There were significant correlations between the degree of PR and RVEDV ($r = 0.9$, $p = 0.0001$), RVEDD ($r = 0.7$, $p = 0.0001$), MPI ($r = 0.7$, $p = 0.0001$) and IVA ($r = -0.7$, $p = 0.0001$). The correlations were also significant between BNP level and RVEDV ($r = 0.7$, $p = 0.0001$), MPI ($r = 0.6$, $p = 0.0001$), IVA ($r = -0.4$, $p = 0.002$) and the degree of PR ($r = 0.6$, $p = 0.0001$).
Conclusions: As a result, the severity of PR has a negative influence on both systolic and diastolic right ventricular functions, and both MPI and IVA from the right ventricular basal segments might be used for the assessment of right ventricular functions.

P-113
Left ventricular hypertrophy and arterial hypertension after successful aortic coarctation repair in pediatric age: results from a 12-year follow-up
Bocelli A., Favilli S., Pollini I., Bini R.M.
Pediatric Cardiology Unit, A. Meyer Hospital, Firenze, Italy

Objectives: Systemic arterial hypertension is a quite common complication in the long-term follow-up of patients with successfully repaired aortic coarctation (AC). Furthermore, abnormal blood pressure during exercise stress test and/or 24-hours ambulatory blood pressure monitoring (ABPM) is reported in patients who are normotensive at rest and has been related to increased left ventricular mass. We aimed at exploring the prevalence and predictors of left ventricular hypertrophy (LVH) and late onset hypertension in patients submitted to successful AC surgery or percutaneous intervention in the pediatric age.
Methods: A total of 48 subjects (62.5% male, median age 15.1 ± 9.7 years) who underwent corrective surgery or angioplasty for AC between 1977 and 2007 (median [interquartile range] age at correction 2.1 [0.8–41.7] months) were followed at our Department for development of late complications. All patients underwent periodic clinical examination, electrocardiogram and echocardiogram. Supine bicycle exercise echocardiogram was performed in 22 subjects >12 years, and ABPM was performed in 20 patients >7 years.
Results: Over a mean follow up of 12.6 ± 9.1 years, the overall prevalence of LVH, as identified after normalization to height$^{2.7}$ using sex-specific pediatric criteria, was 39.6%, with concentric

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LVH representing approximately 31.6% of cases. Of note, 47.6% of patients with evidence of LVH were normotensive at rest. Late-onset hypertension was observed in 6 (16.7%) of the 36 patients with no evidence of residual postoperative hypertension. Abnormal hypertensive response to exercise was detected in one third of patients normotensive at rest, and was not related to either rest or effort residual transisthmic gradient. Recoarctation occurred in 11 (22.9%) of patients. At multivariate analysis, the risk of LVH was independently related to age at the time of AC repair and residual hypertension, irrespective of adequate medical treatment.
Conclusions: In our population of AC patients, the prevalence of LVH was higher than previously reported, being detectable even in subjects with a good late result that are normotensive at rest. In repaired AC, LVH may represent a simple and reliable echocardiographic marker of cardiac involvement that can be easily assessed, even in patients who are not able to perform effort test or ABPM.

P-114

The mid-term follow-up and ongoing morbidity during childhood and adolescence in surviving premature neonates with a prior patent ductus arteriosus

Dodge-Khatami A. (1), Tschuppert S. (1), Doell C. (1), Latal B. (2), Valsangiacomo Buechel E. (1), Bauersfeld U. (1), Prêtre R. (1)
University Children's Hospital (1), Child Development Center (2), Zurich, Switzerland

Objectives: To determine mid-term survival and eventual ongoing morbidity in children who were previously premature neonates and underwent a patent ductus arteriosus (PDA) closure.
Methods: From a consecutive series of 210 premature babies who underwent medical (MED; n = 168) and/or surgical (SURG; n = 42) treatment of a PDA between 1985 and 2005, a cohort of 180 hospital survivors was studied, with detailed questionnaires filled out by caring pediatricians/physicians. The groups were compared using a 2-tailed Pearson Chi-square test.
Results: Complete follow-up was obtained in 129/180 patients (72%), 84% in the surgical and 69% in the medical group. At a median follow up of 7 years (range 2–22 years), there were 3 late deaths (2.3%). At last follow-up, only 45% of patients were considered healthy by their physicians, with a significant difference between children having had prior medical treatment (56.1% healthy) versus those after surgery (9.7%; $p < 0.001$). Significant co-morbidities, mainly related to prematurity, were frequent in the entire cohort and comparable between the two groups: developmental delay (medium and severe; 11.2% MED vs. 13.0% SURG), pulmonary illnesses (12.2% MED vs. 12.9% SURG), gastro-intestinal disease (2.0% MED vs. 6.5% SURG), neurological impairment (15.3% MED vs. 12.9% SURG), and hearing impairment (2.0% MED vs. 9.7% SURG). Slight developmental delay was significantly more prevalent in the surgical group (48.4%) compared to the medical group (29.5%; $p = 0.012$). Thoracic deformity with scoliosis occurred in 2/31 (6.5%) patients, only after surgical PDA ligation.
Conclusion: After initial hospital mortality following closure of a hemodynamically significant PDA in premature neonates, survival is satisfactory until childhood and adolescence. Physical and mental co-morbidity, including developmental delay, are most likely related to prematurity, and not necessarily altered by successful medical or surgical closure of a PDA. Considering that only half of the patients are considered healthy at last follow-up despite successful PDA closure, parent counselling in premature neonates should be cautious, given the co-morbidities related to prematurity.

P-115

Renal function in patients with Eisenmengers Syndrome

Jensen A.S. (2), Recke P.V.D. (1), Sondergaard L. (2)
Department of Radiology [1], Department of Cardiology (2), Rigshospitalet, Copenhagen University Hospital, Denmark

Background: It is known from the literature that cyanotic heart disease can lead to impaired renal function and secondary systemic arterial hypertension. One explanation can be that these patients have an increased risk of thromboembolic events so that renal infarcts can explain the renal dysfunction.
The aim of this study was to examine the prevalence of diminished renal function and arterial hypertension, and to examine whether this could be explained by renal infarcts.
Methods: Eighteen outpatients with Eisenmenger's syndrome were examined. Blood samples were analysed for creatinine and uric acid, and Chrom EDTA determined glomerular filtration rate (GFR). Furthermore, 24 hours blood pressure recording and CT-scan of the kidneys with contrast were performed.
Results: The cohort consisted predominantly of females (n = 11/18, 61%). None of the patients were known with renal disease but two patients (11%) were known with well regulated systemic arterial hypertension. Eleven (61%) of the patients had previous thromboses in other organs.
Blood samples revealed that only one patient (6%) had a slightly increased creatinine level, whereas fourteen patients (78%) had increased uric acid. The Chrom EDTA revealed that 7 patients (39%) had moderately reduced GFR. The 24 hours blood pressure recording showed that all patients including the two patients with hypertension were normotensive. The CT-scans did only show signs of renal infarct in two of the patients, where only one of them had affected GFR.
Conclusion: Interestingly, this study showed that even though most of our patients had normal creatinine values, 39% had moderately reduced renal function. Renal infarction was only found in two patients, and only one of them had affected GFR so this does not seem to be the explanation. Other reasons have been mentioned such as chronically organ ischemia due to hypoxemia and/or hyperviscosity. This needs further investigation, but it is important for the physician to know, that even though the creatinine level is normal, renal function can be diminished and therefore maybe controlled regularly in these patients.

P-116

Limited Exercise Capacity After Coarctation Repair Even With Optimal Surgical Results

Hager A., Kanz S., Kaemmerer H., Hess J.
Department of Pediatric Cardiology and Congenital Heart Disease, Deutsches Herzzentrum München, Technische Universität München, Germany

Objective: There are contradictory reports whether exercise capacity is reduced in patients on long-term after coarctation repair. Data from unselected patient groups are missing.
Design: cross-sectional long-term follow-up study in a tertiary congenital cardiology referral centre.
Patients and methods: 260 patients (30.2 ± 11.4 years old, 84 female) after surgical repair for isolated aortic coarctation (age at surgery 11.5 ± 11.2 years) underwent a symptom-limited exercise test.
Results: Peak work load was 180 ± 52 Watt and significant less than the age and height related reference values ($p < 0.0005$). A peak work load under 80% of expected was found in 200 patients (76.9%). Exercise performance of the patients was independent

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from age at surgery, type of surgery or the systolic brachial-ankle blood pressure difference. The only exercise limiting factor found was the chronic administration of diuretics to treat hypertension ($r = -.174$, $p = .005$).
Exercise hypertension, defined as a systolic blood pressure > 2 SD above the load-dependent reference value, was found in 73 patients (28.1%). It was independently related to the systolic brachial-ankle blood pressure difference ($p < .0005$) and diuretics administration ($p = .037$).
Conclusions: Most patients after coarctation repair have a reduced exercise performance. This is not related to the surgical approach and results. Particularly, as these patients are at risk of early atherosclerosis, exercise should be promoted as primary prevention after restenosis, aortic or cerebral aneurysms and severe exercise hypertension are ruled out.

P-117
Fetal outcome in women with heart disease – a UK single centre experience at University College London Hospitals NHS Trust
Brooks R. (1), von Klemperer K. (1), Abdin S. (2), Peebles D. (2), O'Brien P. (2), Walker F. (1).
The Heart Hospital, UCLH, London, UK (1); Elizabeth Garret Anderson Hospital, UCLH, London, UK (2)

Introduction: The prevalence of maternal heart disease (MHD) remains constant at 1%, however its complexity is increasing due to improved survival of those with congenital and inherited heart disorders. There is limited fetal outcome data for those with such complex heart defects, and we therefore report our experience.
Method: Data on completed pregnancies between September 1st 2003–September 1st 2007 were collected with reference to complexity of MHD, gestation at delivery, birth weight and fetal outcome.
Results: Fetal outcome data was available for 77% of all babies ($n = 213$). There were 204 pregnancies in 183 women (twins [$n = 9$]). Complexity of MHD; simple (68), moderate (91), highly complex (45).
There were 111 deliveries at UCLH, 89 in local obstetric units (DGH) and 3 deliveries at the Heart hospital, UCLH.
Gestational age of live births; < 36/40 (33), term (151) >40/40 (20). Mean birth weight (kg) (singleton 2.95 [range 0.68 – 5.30], twins 2.49 [range 1.62- 3.26). 33 delivered < 36/40; premature labour (8), IUGR (5) obstetric (11), patient choice (1), cardiac indication (8) (congestive heart failure [5], aortic dissection [1], mechanical valve thrombosis [1] severe connective tissue disease [1]).
Four babies had congenital heart disease (CHD) (2%) (atrial septal defect [1], tetralogy of Fallot [TOF] [1], patent ductus arteriosus [1] and ventricular septal defect [1]. Only 1 required treatment (TOF). Other diagnoses (3); brain cystocoele, rectal cyst, talipes.
Fetal deaths (3) (unexplained intrauterine [2] at 33/40, 37/40 and 6 hrs postpartum at 40/40).
Conclusion: Fetal outcome in women with HD is good. The majority of babies were born at term with normal birth weight (96%). Mean birth weight of babies born at term was 3.063 kg. Those born preterm have no residual disability. Recurrence of CHD is low (2%) and IUGR secondary to MHD is uncommon (2%).

P-118
Biological valve replacement and pregnancy – is there a concern?
Cleuziou J. (1), Teodorowicz A. (1), Hörer J. (1), Prodan Z. (1), Kaemmerer H. (2), Vogt M. (2), Schreiber C. (1), Lange R. (1)
Department of Cardiovascular Surgery (1) and Department of Paediatric Cardiology and Congenital Heart Disease (2), German Heart Centre Munich at the Technical University

Objectives: Pregnancy in women with a biological heart valve prosthesis, might lead to a faster deterioration of the prosthesis. We sought to look at the outcome of pregnancies in young women with a biological valve prosthesis, focusing on the course of the pregnancies and the rate of prosthesis replacement.
Methods: At time of the study, 100 female patients who underwent a biological valve replacement between 1976–2006, were 18–40 years old. Eighty seven women were evaluated by a questionnaire at a mean interval of 10.8 ± 8 years after valve replacement.
Results: A biological prosthesis was implanted in 45 patients (51.7%), 42 patients (48.3%) had a homograft placement. The position of the valve was pulmonic in 50 (57.5%), aortic in 22 (25.3%), tricuspid in 8 (9.1%), mitral in 6 (6.9%) and combined aortic and mitral in one patient (1.1%). After valve replacement, 33 patients (37.9%) had a total of 56 pregnancies with a live birth rate of 76.8% ($n = 43$). There were 9 (16%) miscarriages, 4 (7.1%) abortions and no stillbirths. A caesarean section was performed in 19 (44.2%) patients, whereas 24 (55.8%) had a vaginal delivery. All but 3 newborns were healthy. Two have a congenital heart defect and one has a hearing and speaking disorder. Cardiac complications during the pregnancy were rhythm disturbances in 4 patients (12.1%). Thirty one (35.6%) patients required a replacement of the valve prosthesis at a mean time of 7.6 ± 5 years after implantation. Twelve (38.7%) of them had been pregnant. The freedom from prosthesis replacement at 5 years after implantation, was $96.4 \pm 3.5\%$ for patients after a pregnancy compared to $93.1 \pm 3.9\%$ for patients without a pregnancy ($p = 0.2$).
Conclusions: Our limited data point out that young women who plan to conceive later and require a heart valve replacement should be advised to opt for a biological prosthesis, even if reoperation is inevitable. The risk must be weighed against the increased risk of pregnancy with a mechanical valve and anticoagulation. A pregnancy does not increase the risk for the replacement of a biological prosthesis, particularly not during or short after pregnancy.

P-119
Evaluation of right heart failure in grown up patients after correction of congenital heart disease
Lemmer J. (1), Heise G. (1), Peters B. (3), Rentzsch A. (4), Boettler P. (2), Kuehne T. (1), Dubowy K.O. (5), Hager A. (6), Stiller B. (1,2) Supported by Competence Network for Congenital Heart Defects founded by the Federal Ministry of Education and Research (BMBF), FKZ 01G10210
German Heart Institute, Berlin, Germany (1), Albert-Ludwigs-University Freiburg, Germany (2), Otto-v-Guericke-University Magdeburg, Germany (3), Universität des Saarlandes. Homburg/Saar, Germany (4), Heart and Diabetes Center North Rhine-Westphalia, Bad Oeynhausen, Germany (5), German Heart Centre, Munich, Germany (6)

Introduction: The aim of the presented study was the identification of predictive parameters for early detection of right ventricular failure in patients with congenital heart disease (CHD) after corrective surgery.
Methods: We performed a prospectively designed assessment of clinical, neurohumoral and functional parameters in 104 GUCH patients with congenital right heart disease and corrective surgery (mean age 20.4 ± 7.3 years) and 54 sex- and age-matched healthy

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control subjects (mean age 21.2 ± 7.7 years). The study was conducted as multi-centre study with 9 university hospitals or cardiac centres recruiting patients and controls between October 2003 and June 2006.
Results: Compared to controls GUCH patients with right heart load had significant higher circulating levels of natriuretic peptides, even when they are still asymptomatic (tab.). But regarding the endogen catecholamine levels (noradrenaline 244.47 ± 89.33 vs. 251.77 ± 89.46, p = 0.716) as well as angiotensin, aldosterone and ET-1 (1.86 ± 1.06 vs. 1.99 ± 1.08, p = 0.994) plasma levels no differences between patients and healthy controls could be found. Plasma levels of natriuretic peptides increase according to NYHA classification (NTpro-BNP: NYHA I vs. NYHA II/III 85.6 ± 1.1 vs. 139.7 ± 1.2 pg/ml, p = 0.001). Besides that significant differences in maximal exercise capacity assessed by treadmill exercise testing (VO2 max 29.1 versus 37.5 ml/kg/min, p < 0.001), right ventricular ejection fraction (RVEF) and the enddiastolic right ventricular area (RVd) measured by ECHO and MRI (see tab.) could be found.

	Patients (n = 104)	Control subjects (n = 54)	p
NTpro-BNP (pg/ml)	101.29 ± 1.09	25.38 ± 1,12	< 0.001
BNP(pg/ml)	36.60 ± 2.1	17.57 ± 1.1	< 0.001
MidproANP (pg/ml)	71.66 ± 1.0	55.20 ± 1.0	< 0.001
RVEF (%) [ECHO]	40.8 ± 12.1	46.26 ± 10.54	0.039
RVd (cm^2) [ECHO]	27.63 ± 8.8	18.67 ± 4.6	< 0.01
RVEDV (ml) [MRI]	188.50 ± 65.91	162.27 ± 31.50	0.01
RVESV (ml) [MRI]	99.23 ± 45.75	67.42 ± 20.16	< 0.001

Conclusions: Even in asymptomatic patients (NYHA I) after correction of CHD and right ventricular load differences in natriuretic peptide plasma levels as well as in exercise capacity, echocardiographic and MRI parameters can be found. Electrocardiogram, echocardiographic and spiroergometric data correlate well with the plasma concentration of natriuretic peptides. Thereby NTpro-BNP seems to be the most sensitive parameter to show beginning right heart failure. On the basis of this parameter (in accordance with clinical parameters) it seems possible to identify those patients who may benefit from early (prophylactic) cardioprotective medication.

P-120

Intermediate-term follow-up after end-to-end anastomosis for coarctation of the aorta - one center experience

Jagiellowicz D. (1), Kawalec W. (1), Brzezinska-Rajszys G. (2), Niemirska A. (3), Turska-Kmiec A. (1)
The Children's Memorial Health Institute, Warsaw, Poland; Cardiology Clinic (1), Heart Catheterization Laboratory (2), Nephrology Clinic (3)

Introduction: End-to-end operation is a standard treatment of coarctation of the aorta in children. The aim of the study was to evaluate the 6–12yrs results of end-to-end anastomosis performed in one center by evaluation of the blood pressure (BP), measurements of left ventricle mass by echocardiography and intima-media thickness (IMT) in relation to age at surgery.
Material/methods: In 50 patients operated between 1995–2000 ABPM, echocardiography, IMT measurements were performed. Patients were divided into two groups. Group I–15 pts operated below 3mths of age (range 0.3–2.5 mths, mean 1.2), the current age 6.4–11.2 yrs (mean 7.9 yrs), the follow-up 6.3–11.2 yrs (mean 7.8 yrs). Group II–35 pts operated after 3 mths of age (range 3.1–98.2 mths, mean 30.1 mths), the current age 6.65–18 yrs (mean 11.9 yrs), the follow-up 6.4–12.2 yrs (mean 9.3 yrs).
Results:

	Group I	Group II
Number of patients	15	35
Age at surgery (mths)	1.2+/-0.766	30.1+/-29.6
Follow-up (yrs)	7.8	9.3
Average day-time systolic pressure (mmHg)	118+/-9	120+/-10
Average night-time systolic pressure (mmHg)	102+/-9	105+/-11
24-hours pressure load	23+/-25	31+/-22
Day-time index	0.96	0.93
Night-time index	0.97	0.93
Arterial hypertension (pts)	5 (33%)	9 (26%)
IMT RCA (mm)	0.449 +/-0.04	0.421+/- 0.053
IMT LCA (mm)	0.407+/-0.066	0.436+/- 0.051
Desc Ao max gradient (mmHg)	27.79+/-5.62	27.73+/-7.32
LV mass (gram)	62.42	111.75
LVM/H 2.7	34.66	37.41

No significant differences were found with respect to pressure index and load in ABPM, left ventricle mass index and IMT between two groups. Multiple correlation coefficients between IMT RCA, IMT LCA and others variables were absent.
Conclusion: Age of children operated for coarctation of the aorta by end-to-end anastomosis has no significant influence on the IMT, the 24-hour pressure load and pressures indexes in ABPM, as well as the index of the mass of left ventricle.
There is a substantial incidence of arterial hypertension in inter-medium term follow-up in patients after end-to-end operation.

P-121

Transcatheter therapy of aortic coarctation has widely displaced surgery

Ewert P., Moysich A., Peters B., Kühne T., Ovroutsky S., Miera O., Nagdyman N., Berger F.
Department of Congenital Heart Diseases/Peadiatric Cardiology, German Heart Institute Berlin, Berlin, Germany

Introduction: The use of balloonexpandable stents and self expandable stent grafts have amplified the transcatheter possibilities of the treatment of coarctation in adults. How important is the intervention in comparison to surgery?
Methods: Retrospective analysis of all 106 adult patients with coarctation or re-coarctation who were treated during the last 10 years either by surgery or intervention at our institution. We compared the patients treated from 1997 to 2001 (group A, n = 49) with those treated from 2002 to 2006 (group B, n = 57) with regard to the frequency of operation versus intervention and analysed the causes which led to surgery.
Results: In group A 63% of the patients were operated, in group B only 18% (10 of 57). The causes of operation were in one patient an aneurysm five years after balloon dilatation and in the remaining 9 patients re-coarctations after surgery (3x small interponats; 2x aneurysms in immediate vicinity to carotid arteries; 2x gothic arch morphology of the anastomosis; 1x hypoplastic transverse arch; 1x not dilatable scar). The type of intervention were in group A 39% stent implantation in group B 60% and increased to 83% in the last year.

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Conclusion: The therapy of first choice of coarctation or re-coarctation in adulthood is the intervention. Surgery remains an option for difficult residual defects mainly after surgery. With the further expansion of transcatheter treatment already during childhood, those cases will probably become even less frequent.

P-122

Are adolescent and adult patients adequate informed about their congenital heart disease?

Toussaint-Götz.N (1), Vahsen N. (1), Kaestner M. (1), Muentjes C. (2), Neudorf U. (2), Schneider M (1)
German Pediatric Heart Center, Departement of Pediatric Cardiology ,Sankt Augustin (1); Departement of Pediatric Cardiology, University Essen (2), Germany

Introduction: The number of the adults with congenital heart disease constantly increases. Although surgical correction is performed in early childhood or even in neonates, lifelong follow-up investigations are necessary. For the close contact to the cardiologist on a regular basis a sound understanding of the own disease is crucial.The aim of the study is to assess the illness understanding of the adolescent and adult patients.
Patients and methods: Between February and July 2007 127 patients (w 70, m 57) at the age of 14 to 61 years (mean 19,4 years) answered a questionnaire during an outpatient visit. The questionnaire was based on the Leuven knowledge questionnaire for congenital heart disease. The patients were divided up into two groups according to their age: Group 1 14–18 years (n = 63) and group 2 18–61 years (n = 64).
Results: Only 58 % of the patients could name their cardiac defect, 50% were able to describe the defect or to mark it in a diagramm. 42% of the patients know about the necessity of an endocarditis prophylaxis but only 18% knows the term endocarditis.
In the female population 58% of does not know whether a pregnancy may detoriate the cardiac function.
37% of the people asked felt influenced by the cardiac defect in their choice of occupation and 33% during the everyday life.
The adult patient were significantly better informed and showed a better understanding of the heart disease than the adolescents. On the other hand the adult patients felt more restricted by their heart disease then the adolescent.
Conclusion: In adolescent and adult patients the illness understanding shows considerable gaps. Intensive and structured patient consultations are necessary and can contribute to reduce fears and uncertainties and consequently increases their quality of life. Additionally, from the medical point of view a good understanding of the own disease might increase patients discipline to keep to the medical follow up investigations on an regular basis.

P-123

Predictors of late outcome after surgery for congenital heart disease in the adult population

Padalino M.A. (1), Cogo P.E. (2), Speggiorin S. (1), Vida V.L. (1), Maschietto N. (2), Reffo E. (2), Daliento L. (3), Stellin G. (1)
Pediatric and Congenital Cardiac Surgery Unit, University of Padua, Medical School, Italy (1); Departments of Pediatrics, University of Padua, Medical School, Italy (2); Department of Cardiology, University of Padua, Medical School, Italy (3)

Aims: We report our experience with surgery for congenital heart disease in adults, to evaluate its effectiveness, safety and clinical predictors of late outcome.
Methods and Results: We collected data of 161 consecutive patients who underwent 230 procedures from January 1997 to December 2004. Patients were divided into two surgical groups: Repair (85.7%) and Reoperation (14.3).
There was only one in-hospital death (Fontan revision, in Reoperation group). At a mean follow up time of 56 ± 30 months, overall freedom from any kind of complication is 76.5%. Cox analysis showed that incremental risk factors were preoperative cyanosis, reoperation and length of ICU stay (HR = 4.47, 3.34 and 1.49 respectively; $p < 0.001$ in all), while decremental risk factors were preoperative NYHA class 1 (HR = 0.328, $p < 0.001$), surgery for "septal defect" (HR = 0.26 p = 0.02).
Conclusions: surgery for congenital heart disease in the adult age is an overall safe and a low risk treatment, especially when it is performed in patients with good preoperative clinical conditions, before cardiac decompensation occurs.

P-124

Sexuality and reproductive health related problems in women with congenital cardiac disease

Vigl M. (1), Bauer U. (1), Niggemeyer E. (1), Nagdyman N. (2), Rattenhuber J. (3), Trigas V. (4), Kaemmerer M. (4), Seifert-Klauss V. (3), Schneider K.T.M. (3), Berger F. (2), Hess J. (4), Kaemmerer H. (4)
Competence Network Congenital Heart Defects, Berlin, Germany (1); Deutsches Herzzentrum Berlin, Berlin, Germany (2); Frauenklinik der Technischen Universität München, Munich, Germany (3); Deutsches Herzzentrum München, Munich, Germany

Introduction: Due to the improved life expectancy a rising number of women are entering sexual maturity. Sexuality and reproductive health become important issues in the life of these patients.
Methods: Questionnaire-based observational survey over 12 months in two tertiary care centres, including 540 adult female patients with a median age of 29 years [18–75]. The study population was grouped into functional classes according to Perloff (functional class I: 51.0%; II: 40.8%; III/IV: 7.2%)
Results: The first menstrual bleeding took place at a mean age of 13.4 years and in patients with cyanotic cardiac anomalies at 14.4 years; this is later than the figures from the general population (12.9 years). 25.9% of the patients had at least once sought medical assistance for menstrual irregularities and 7.8% complained about intensified cardiac symptoms during the menses.This figures rose to 19.4% and 16.3% in the functional class III/IV and in the cyanotic group respectively (both $p < 0.05$). The risk of hypermenorrhoe increased in the group of patients under anticoagulation therapy (OR = 2.8; 1.6–5.1). The mean age at the first sexual intercourse was 17.7 years and 9.4% of the patients reported to have physical problems related to the heart defect during sexual activity. This rate was significantly increased in the functional class III/IV (27.3%, $p < 0.001$).
Conclusions: Congenital cardiac defects are interrelated to different aspects of the reproductive health of female patients. Information on sexual development and the risks of a possible pregnancy are crucial in this patient population and at present insufficiently addressed. These issues have to be proactively included in the care of female patients with congenital cardiac defects.

P-125

Coronary anatomy in patients with transposition of great arteries and Musturd procedure : a study with 64 multislice computed tomography.

Marcora S. (1), Di Renzi P. (2), Bellelli A. (2), Pierleoni M. (2), Mancini P. (2), Giannico S. (1), Sanders S.P. (1)
Bambino Gesù Paediatric Hospital, Rome, Italy (1); Fatebenefratelli Isola Tiberina Hospital, Rome, Italy(2)

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Introduction and Objective: It is well known the ability of cardiac computed tomography in performing a non invasive study of coronary vessels. This technique is also used to evaluate cardiac anatomy and function when magnetic resonance (MRI) is not possible. The aim of this study is to determine coronary anatomy in patients (pts) with transposition of great arteries (TGA) and Mustard procedure (Mustard pts) using 64-multislice computed tomography (64-MSCT).
Methods: A 64 MSCT examination was performed in 12 Mustard pts (age 21.3 ± 3.1years; 9 male and 3 female) who were not suitable to MRI because of a pacemaker and need a complete evaluation of biventricular function. A non- iodinate contrast agent was injected. Ecg-retrospective gating was used in order to reconstruct MIP, MPR and VR images in different phases of cardiac cycle. Two observers, a radiologist and a cardiologist, separately described origin and anatomy of coronaries .
Results: Seven pts were studied at 80–90%, 3 pts at 40–50% and 2 pts at 0% of cardiac cycle . In all pts the non coronary cusp was anterior and non facing the pulmonary valve. In 11/12 pts there was an origin of the coronaries usual for TGA. In 1/12 (8%) there was an "inverted right coronary artery (RCA) and circumflex coronary artery (CX)" with RCA and descending coronary artery (DA) from left coronary cusp and CX from right coronary cusp. In 1 pts a "intramiocardic bridge" of DA was observed. In all cases there was a relative hypoplasia of coronary vessels for non-systemic left ventricle (LV).
Conclusion: 64 MSCT showed a possible variability in the origin of coronary vessels in pts with TGA, as described in previous echocardiographic studies. Moreover this study demonstrated hypoplasia of left coronary vessels in Mustard pts in long term follow up. This hypoplasia could determine some possible ischemic effects on LV such as fibrosis and myocardial dysfunction with some important therapeutic consequences in term of LV retraining or biventricular resynchronization. 64-MSCT and other non invasive technique such as MRI , cardiac scintigraphy and stress echocardiography could be used to further investigate this fascinating hypotesis.

P-126
Right Ventricular Dilatation Index: An Echocardiographic Index to Evaluate The Both Systolic and Diastolic Right Ventricular Functions in Patients After Repair of Tetralogy of Fallot
Çetin İ., Örün U.A., Varan B., Gökdemir M., Cýndýk N., Tokel K.
Baskent University, Pediatric Cardiology, Ankara, Turkey

Objectives: It was aimed to build up a new and practical index to determine the critical level of the right ventricular dysfunction to decide the necessity of pulmonary valve replacement (PVR) after repair of tetralogy of Fallot (ToF).
Methods: 25 patients with a mean age of 14.1 ± 4.4 years who underwent repair of ToF at a mean age of 4.9 ± 5.1 years and 29 age and sex matched healthy children at a mean age of 13.1 ± 2.8 years enrolled in this study. The right ventricular dilatation index (RVDI) was calculated by using BNP level, QRS duration, myocardial performance index (MPI) and isovolumic acceleration (IVA) at right ventricular basal segments (RVDI: BNP level x QRS duration x RV basal segments MPI / RV basal segments IVA x 100).
Results: BNP levels were significantly higher in patients than in controls (28.3 ± 24.1 vs. 7.4 ± 2.3 pg/ml, $p = 0.0001$). QRS duration (127.2 ± 22.6 vs. 48.6 ± 13.8 ms, $p = 0.0001$) and MPI (1.08 ± 0.35 vs. 0.58 ± 0.11, $p = 0.0001$) were significantly higher, IVA (3.1 ± 0.7 vs. 5.4 ± 1.0 m/s2, $p = 0.0001$) was significantly lower in patients. It was observed that RVDI was significantly higher in patients with severe PR than in patients with moderate PR and also than in controls (86.6 ± 72.2, 26.3 ± 15.3 ve 3.1 ± 1.3, $p = 0.0001$). There were significant correlations between the degree of PR and QRS duration ($r = 0.8$, $p = 0.0001$), BNP levels ($r = 0.6$, $p = 0.0001$), MPI ($r = 0.7$, $p = 0.0001$), IVA ($r = -0.7$, $p = 0.0001$) and RVDI ($r = 0.7$, $p = 0.0001$).
Conclusions: The area below the ROC curve was 0.961 for RVDI to show right ventricular dysfunction. The confidence interval was between 0.915 and 0.999 ($p = 0.0001$). In addition, for the cut-off value of 24.9, the sensitivity was 91% and the specificity was 88%. These results showed that RVDI might be a useful index by evaluating the biochemical, electrical and physical properties of the myocard for the assessment of both systolic and diastolic right ventricular functions.

P-127
Restrictive Right Ventricle Improves Right Ventricular Function in Patients with Pulmonary Regurgitation after Tetralogy of Fallot Repair
Dewayani R., Harimurti G.M., Roebiono P.S., Rahayoe A.U., Atmosudigdo I.S.
Department of Cardiology and Vascular Medicine, Faculty of Medicine, University of Indonesia/National Cardiac Center Harapan Kita, Jakarta, Indonesia

Introduction: Pulmonary regurgitation (PR) is a common problem following tetralogy of Fallot (TOF) repair. PR is well correlated with decreased right ventricle (RV) global function and increasing the incidence of sudden cardiac death. Restrictive RV is present in about half of patients late after TOF repair and is related to good prognosis due to limitation of regurgitation time hence limit the dilation of RV.
Objective: We aimed to sought whether restrictive RV improves RV function in patients with pulmonary regurgitation after TOF repair.
Methods: Fifty (50) patients with repaired TOF were studied. We measured RV function derived from tissue Doppler imaging (TDI) Tei index. We compared the severity of PR with RV global function and the presence of restrictive RV.
Results: There were 50 patients after TOF with mean age when operated 2 years old (7 month–12 years old), mean age when studied 8.8 years old (1.5–29 years old) and operation to follow up interval was 4.8 year (6 month–13 year). Eighteen patients (36%) with restrictive diastolic RV function; RV TDI Tei index was 0.43 ± 0.082 (range 0.25–0.61) which was higher than normal children. There was a tendency of decreased RV global function in patients with severe PR ($p = 0.056$). In a subgroup analysis between moderate PR with and without restrictive RV, we found a strong relation between restrictive RV and better RV global function ($p < 0.005$).
Conclusion: Restrictive RV physiologic persist in 36% of patients after TOF repair. Restrictive RV improves RV function in patients with PR after TOF repair.
Keyword: Restrictive RV, repaired TOF, pulmonary regurgitation, RV global function

P-128
Surgical reconstruction of congenital mitral valve dysplasia: a Tissue Doppler Imaging study in long-term follow-up
Biffanti R. (1), Varotto E. (1), Reffo E. (1), Maschietto N. (1), Stellin G. (2), Padalino M. (2), Vida V., Milanesi O. (2)
Department of Paediatrics (1) and Pediatric Cardiovascular Surgery Unit, University of Padua-Medical School, Italy (2)

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Objectives: The policy of our center is to attempt a surgical valvuloplasty whenever possible. Aim of the study is to evaluate the functional results of such surgical procedure using 2D-Doppler echocardiography and Tissue Doppler Imaging (TDI).
Methods: Twenthyone patients randomly selected among those followed at our Institution, affected by MV stenosis (7) or insufficiency (14) who had surgical valvuloplasty at a mean age of 68,7mos (range 6–227mos), underwent echocardiographic evaluation at a mean follow-up of 139 mos (range 22–243mos). Excluded were patients with MV anomalies in the spectrum of A-V canal defects. All were in NYHA class I/II, on sinus rhythm and all but 2 taking no medication. We assesed the following indexes. Systolic function: FS%, EF%, EDVi, ESVi M-mode derived. Diastolic function: PV FLOW: peak J, peak K, peak Z, J/K,J. MITRAL FLOW: peak E, peak A, E/A, PFR/SV. TDI parameters: peak Em, peak Am, Em/Am, E/Em, peak Sm. Mitral regurgitation using continuity equation and proximal flow convergence method. Our results were compared to normal values from literatures, according to the age.
Results: Residual mild MS was present in 2/21 (0,9%), mitral regurgitation in 9/21(43%): severe in 11%, moderate in 11%, mild to moderate in 11%, mild in 22%, trivial in 44%. EF and SF were normal in all (mean EF 65,3%, mean SF 38,2%).
TDI study showed a significant reduction of Em in 15/21 pts (11.17 cm/sec vs. 15.1–20.7 cm/sec in pts <16yrs of age; 7.36 cm/sec vs. 10.4–14.9 cm/sec in pts >16yrs of age), increased E/E1 in 9/21pts (13 vs. 3.1–5.3), documenting an abnormal relaxation and increased filling pressure of the left ventricle. Sixteen out of 21 pts had a reduction of Sm (7.22 cm/sec vs. 8.8–12.4 cm/sec), documenting an impairment of systolic function, in spite of a normal EF. They were all the patients <16 yrs and 6/11 >16 yrs.
Conclusions: Our data suggest that functional results of MV plasty in pediatric patients are satisfying also at a long follow-up, however TDI shows persistent systolic and diastolic impairment of the LV function.

P-129
Coronary artery bypass in GUCH patients
Nagdyman N. (1), Ewert P. (1), Hetzer R. (2), Berger F. (1)
Department of Congenital Heart Defects and Pediatric Cardiology (1); Department of Cardiothoracic and Vascular Surgery (2); Deutsches Herzzentrum Berlin, Germany

Introduction: To review data from a unit for grown ups with congenital heart disease (GUCH) for coronary artery bypass.
Methods: Follow-up of 1594 GUCH patients in a period of 19 years. We excluded patients with arterial switch operations and patients who had anomalies at the coronary arteries like anomalous origin of the left coronary artery. Included were patients with coronary artery stenoses which were leading to a coronary artery bypass operation.
Results: 66 patients (42 m; 24 f; median age 63.5 years, range 39-89 years) needed a coronary bypass operation. Congenital heart defect diagnoses were : atrial septal defect and PAPVR ($n=52$); partial endocardial cushion defect ($n=4$); VSD ($n=5$), TOF ($n=2$), absent pulmonary valve ($n=1$); coarctation of aorta ($n=1$) und aortic stenosis ($n=1$). In 57 patients the coronary bypass operation was performed simultaneous with the operation of the congenital heart defect. 3 patients died in the early postoperative period.
Conclusions: The analysis demonstrates that GUCH patients develop with higher age additionally coronary artery stenoses which require surgery. The number GUCH patients will increase due to their better actuarial survival rate and therefore we have to count on a higher incidence of coronary artery problems in this patient group.

P-130
Evaluation of Right Ventricular Functions and B-type Natriuretic Peptide Levels by Cardiopulmonary Exercise Test in Patients with Pulmonary Regurgitation After Repair of Tetralogy of Fallot
Çetin İ. (1), Tokel K. (1), Varan B. (1), Örün U.A. (1), Gökdemir M. (1), Cındık N. (1), Eyüboğlu F. (2), Ulubay G. (2), Aşlamacı S. (3)
Baskent University, Pediatric Cardiology, Ankara, Turkey (1); Baskent University, Chest Diseases, Ankara, Turkey (2); Baskent University, Cardiovascular Surgery, Ankara, Turkey (3)

Objectives: Impairment of right ventricular functions especially due to chronic pulmonary regurgitation (PR) is a well known entity in patients with tetralogy of Fallot (ToF) after repair. The aim of this study was to examine the relation between BNP levels and right ventricular dysfunction by cardiopulmonary exercise test (ET) in patients after repair of ToF.
Methods: Twenty five patients with a mean age of 14.1 ± 4.4 years who underwent repair of ToF at a mean age of 4.9 ± 5.1 years and 29 age and sex matched healthy children at a mean age of 13.1 ± 2.8 years were enrolled in this study. BNP levels were measured at baseline and at maximal exercise. The volume of right ventricle (RV) and the degree of PR were assessed by two-dimensional echocardiography and color Doppler.
Results: BNP levels were significantly higher in patients with ToF than in controls (28.3 ± 24.1 vs. 7.4 ± 2.3 pg/ml, $p=0.0001$). Exercise was associated with increased plasma BNP levels in both groups. A greater increase in BNP was noted in patients than in controls (37.6 ± 27.5 vs. 11.3 ± 4.5 pg/ml, $p=0.0001$). Forced vital capacity (FVC%) (84.9 ± 16.9 vs. 98.4 ± 18.2, $p=0.01$) and forced expiratory volume during the 1st second (FEV1%) (91.5 ± 19.3 vs. 103.8 ± 16.1, $p=0.02$) were decreased, exercise duration (ED) (10.1 ± 1.9 vs. 11.4 ± 1.7 min, $p=0.02$), maximum heart rate (HRmax) (171.2 ± 18.9 vs. 186.4 ± 13.9/min, $p=0.004$) and maximum oxygen uptake (VO2max) (1.56 ± 0.53 vs. 2.1 ± 0.6 L/min, $p=0.007$) were lower in patients. There were significant correlations between the degree of PR and ED ($r=-0.3$, $p=0.009$), HRmax ($r=-0.4$, $p=0.001$) and VO2max ($r=-0.4$, $p=0.001$). The correlations were significant both before and after exercise, being more pronounced after exercise between BNP level and the degree of PR ($r=0.6$, $p=0.0001$).
Conclusions: As a result, the severity of PR has a negative influence on right ventricular functions and there is significant relation between right ventricular functions and exercise capacity.

P-131
Psychological and health factors affecting disease perception in Adults with Congenital Heart Defects
Callus E. (1), Quadri E. (1), Negura D. (1), Butera G. (1), Fesslova V. (1), Zerbetto R. (2), Carminati M. (1), Chessa M. (1)
IRCCS-Policlinico San Donato, San Donato M.se, Milan, Italy (1); Centro Studi di Terapia della Gestalt, Milan, Italy (2)

Objectives: To verify if there is a discrepancy between the actual disease severity as rated by the medical doctor, and the perception of the disease severity of the patients and what factors could influence these two ratings.
Methods: In this descriptive, cross sectional study, the disease severity of 100 adults with congenital heart defects (ACHD)

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patients (The following diagnoses were present in the sample: VSD, ASD, PFO, pulmonary and aortic stenosis, tetralogy of fallot, transposition of great arteries, atresia and other conditions) was evaluated by means of a numerical rating scale (NRS) 0–100 (0 being the least severe and 100 being the most severe) by both medical doctor and patient. In order to evaluate the severity of the condition the NYHA functional class and the number of previous interventions (both catheterisms and open heart surgeries) were also considered. The following two psychological questionnaires were also administered: PGWBI: Psychological General Wellbeing Index (measures quality of life) and IBQ: Illness Behavior Questionnaire (measures the perception the patients have of being ill).

Results: A high correlation was found between disease perception and evaluation, however the correlation is not perfect indicating that there are some differences. When correlating the various measures and subscales to the two evaluations, the doctor's evaluation was more correlated to the NYHA scales and the previous number of interventions, whereas the patients' evaluations correlated more with psychological constructs such as anxiety, disease perception and (absence of) self-control.

Conclusions: Even though most of the time the patients' perception of their illness severity is close to the one given by the doctors, some patients highly underestimate or overestimate their condition. It seems that the patients are also significantly affected by psychological factors when rating the severity of their condition.

P-132

Myocardial blood flow and viability in children with transpositon of the great arteries after arterial switch operation assessed with high field MRI

Rickers C. (1), Voges I. (1), Jerosch-Herold M. (2), Hart C. (1), Hansen T. (1), Helle M. (1), Heiner Kramer H. (1)
University Hospital Kiel, Dept. Ped. Card., Germany (1);
Harvard University, Brigham and Womens Hospital, Boston, USA (2)

Introduction: In children with transposition of the great arteries (TGA), arterial switch operation (ASO) with coronary reimplantation is accepted as the management of choice. However, myocardial ischemia may represent an important risk factor for sudden cardiac death and infarction late after ASO. Purpose: We hypothesized that high-field cardiac magnetic resonance imaging would allow more precise quantitative assessment of regional blood flow distribution and viability in these patients.

Methods: MRI first-pass perfusion imaging (0.03 mmol/kg Gd-DTPA; TR/TE/α = 2.6/1.1/20°) and cine MRI were performed in asymptomatic 12 TGA patients post ASO (age: 12.9 ± 5.4 yrs, range: 3 months-21 yrs) and 20 healthy volunteers (age: 24 ± 5 yrs) using a 3 Tesla MR-scanner (Philips, Achieva). Myocardial blood flow (ml/g/min) was calculated in 8 LV segments per slice (2–3 slices/pt). Quantitative blood flow at rest and stress (Adenosine 140 mcg/kg/min) was derived from signal intensity curves with a Fermi-model for constrained deconvolution of the tissue contrast-enhancement curves. Late enhancement studies (Gd 0.1 mmol/kg) unsing T1 weighted inversion recovery sequences were performed to detect myocardial necrosis and scarring. Furthermore cine MRI and 3 D coronary artery imaging was performed to assess ventricular function and coronary anatomy. All CMR results were compared to conventional x-ray guided coronary angiography.

Results: In 4 patients myocardial blood flow was significantly reduced at stress due to an occlusion of the left coronary artery in 2 and a hypoplastic left anterior descending artery in one patient. Another pt showed subendocardial ischemia but normal coronary arteries. Scar tissue was detected in the antero-septal region of one patient. Myocardial perfusion reserve (MPR = hyperemic: resting flow) in remaining non-ischemic pts (n = 7) was significantly reduced, compared to healthy volunteers (2.6 ± 0.8 vs. 4.1 ± 1.5; $p < 0.05$).

Conclusions: In asymptomatic children late after arterial switch operation stress-induced perfusion defects and scar tissue can be detected with high-field CMR. The global impairment of coronary flow reserve may indicate altered vasoreactivity following coronary reimplantation.

P-133

Novel Global Strain and Strain Rates Indices are Independent of Angle of Insonation

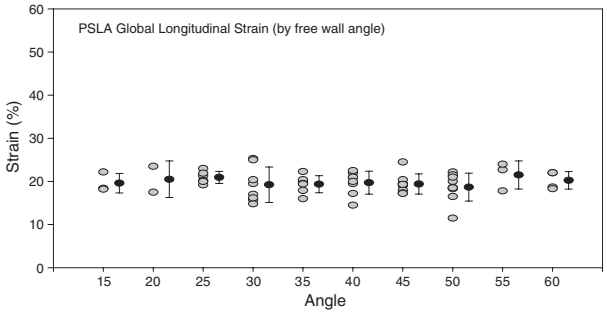
Golender J., Perles Z., Gavri S., El-Laham N., Ibrahim S., Ta-Shma A., Kashani H., Rein A.J.J.T.
Pediatric Cardiology Unit, Hadassah Hebrew University Medical Center, Jerusalem, Israel

Background: 2 Dimentional Strain Imaging (2DSI) is a novel technique for quantitative assessment of myocardial performance. 2DSI allows measurement of two dimensional velocity, strain and strain rates by tracking the digital signature of myocardial "speckles". Global longitudinal strain is a parameter that defines the deformation of ventricular perimeter length throughout a cardiac cycle and calculated as (EDL-ESL)/EDL (EDL and ESL = end diastolic and end systolic length respectively). Strain rate is a rate of deformation per unit of time (strain/Δt). 2DSI, a non Doppler technique, is assumed to be independent of the angle of insonation. Since echocardiographic windows in children allow a wide span of insonation angles of the same cardiac structures, we tested this assumption in pediatric population.

Methods: 30 digital scan-line raw data of echocardiographic studies of children (age 61 (0–219) months) with normal cardiac function were analyzed using 2DSI software at different angles of insonation (General Electric Medical Systems, Milwaukee, WI, USA). In apical four chamber view (A4CH) insonation angles for the septum ranged from 0° to 20° and for the free wall 15° to 40°. In parasternal long axis view (PSLA), insonation angles for the septum ranged from 5° to 45° and for the free wall 15° to 60°. We measured global left ventricular strain and strain rate at increment angle of insonation of 5°.

Results: There was no significant variability of global longitudinal strain and strain rate within described angle span (ANOVA p = ns).

Mean global left ventricular strain and strain rate values in our population were 18.6 ± 2.7% and 1.3 ± 0.25 sec^{-1} in A4CH and 19.9 ± 2.7% and 1.5 ± 0.3 sec^{-1} in PSLA respectively.



Conclusions: This *in-vivo* study confirms that global longitudinal strain and strain rate measured by 2DSI technique are not affected by the angle of insonation up to 60°.

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P-134
Right ventricle adaptation to chronic systemic afterload: a MRI study

Ladouceur M. (1), Redheuil A. (1), Iserin, L. (2), Sidi D. (2), Mousseaux E. (1)
Georges Pompidou European Hospital, Paris, France (1); Necker, Paris, France (2)

Background: Multiple mechanisms of the systemic right ventricle (RV) dysfunction have been suggested: myocardial ischemia, fibrosis or hypertrophy. Chronic increased afterload for morphological RV would be the main determinant of RV function impairment.
Purpose: Evaluation of RV adaptation to systemic afterload by cardiovascular magnetic resonance (CMR) analysis of function, geometry, remodelling and wall-stress index.
Methods: we performed CMR on 38 adult patients (mean age, 27 years) after atrial redirection surgery for transposition of the great arteries and compared them with 19 healthy volunteers. Indexed volumes, mass, ejection fractions were measured. To assess ventricular geometry, perpendicular ventricular diameters (height: Dh and width: Dw) in diastole and systole were measured and the ratio Dh/Dw defined a sphericity index. Wall-stress index was calculated by the formula: humeral systolic blood pressure x end systolic sub-aortic ventricular volume/sub aortic ventricular mass. Late gadolinium enhancement (LGE) study was performed and was classified as absent or present.
Results: 1/Adaptation: compared to normal RV and normal LV, a significant impairment of ejection fraction and a significant hypertrophy of systemic RV were observed. Compared to normal RV, end diastolic (ED) volume of systemic RV was decreased and end systolic (ES) volume did not changed, and compared to normal LV, ED volume of systemic RV was similar and ES volume was increased. In this manner, wall stress index was same in systemic RV and normal LV (table) 2/RV failure: in 11 symptomatic patients (NYHA>I) systemic RV was more hypertrophied, dilated and spherical than asymptomatic patients. Wall stress was also significantly greater and was correlated with RVEF ($R=0.682$, $p<0.0001$). LGE was not related to symptoms: it was found in 58% of patients, among them 41% were symptomatic. Fibrosis was more frequently associated with a decreased RVEF and diastolic sphericity, but not with mass, volumes or wall stress.
Conclusion: concentric hypertrophy is the main mechanism of systemic RV adaptation. Probably RV failure occurs when this mechanism is overstepped and cannot limit RV wall stress anymore, leading to RV dilatation and severe dysfunction. Role of fibrosis in this mechanism of adaptation is unclear but could be a consequence of it.

	asymptomatic patients RV	normal subjects RV	P value	normal subject LV	P value	symptomatic patients RV (NYHA>I)	P value
ED volume (ml/m²)	72±16	88±19	<0.006	79±20	0.2478	116±66	0.0029
ES volume (ml/m²)	33±9	37±7	0.1792	27±7	<0.0189	75±57	0.0007
EF (%)	54±6	57±4	0.0419	65±3	<0.0001	40±12	0.0029
Mass (g/m²)	87±22	47±11	<0.0001	65±17	0.0007	110±32	0.017
	1.78±0.36	2.0±0.3	0.0166	1.28±0.15	<0.0001	1.53±0.20	0.04
Systolic sphericity index	1.61±0.27	2.3±0.35	<0.0001	1.18±0.15	<0.0001	1.41±0.04	0.03
Wall Stress index	41±12	-	-	46±10	0.15	70±50	0.007

P-135
Echocardiographic Preoperative Risk Factors For Norwood Procedure In Hypoplastic Left Heart Syndrome (HLHS)

Mazurek-Kula A. (1), Sysa A. (1), Moll J.A. (1), Moll J.J. (2)
Cardiology Department Polish Mother's Memorial Hospital, Research Institute (1); Cardiosurgery Department Polish Mother's Memorial Hospital, Research Institute (2)

Background: Despite of spectacular improvement of outcome HLHS still remains one of the most difficult challenges in pediatric cardiosurgery.
Material and Methods: Echocardiograms of 110 patients with HLHS 78 boys (70.9%) and 32 girls (29.1%) were reviewed preoperatively (mean age of 5.3±3.97 days). We compared preoperative data such as anatomical subtype of HLHS, ascending aorta diameter, right ventricle function measured by myocardial performance index (RV-MPI), type of atrial communication, tricuspid insufficiency, pulmonary insufficiency in 54 children who survived Norwood procedure (group I) with 56 children who died within 30 days after operation (group II).
Results: The most frequent subtype of HLHS in group I was MS/AS (30.9%), in group II MA/ AA (37.5%). There was a significant difference ($p=0.003774$) in ascending aorta diameter 4.2±1.5mm (2–7.5mm) in group I versus 3.4±1.6mm (1–7.5mm) in group II. In 10.7% patients in group II aortic diameter was less than 2 mm, and there were no patients with so small aorta in group I. The difference in RV-MPI in survivors comparing with no-survivors (0.519±0.158 vs. 0.519±0.182) was not significant (NS). In both groups RV-MPI was significantly ($p=0{,}000$) higher in children with HLHS 0.519±0.17 (0.097–0.968) comparing with age-matched 20 healthy neonates 0.3±0.075 (0.183–0.445).
Restrictive atrial communication was diagnosed in 11 cases (10%); 9 (81.8%) patients in this group died after the operation. Restrictive atrial communication was found in 2 patients (3.7%) in group I and in 9 patients (16.1%) in group II. Severe tricuspid regurgitation was found in 4 patients (7.4%) in group I and in 11 patients (19.6%) in group II. Moderate pulmonary regurgitation was found in 5 patients (9.3%) in group I and in 7 patients (12.5%) in group II; differences were NS ($p>0.05$). There was no severe pulmonary regurgitation.
Conclusions: 1. The most important preoperative risk factor in HLHS is diminutive ascending aorta diameter.
2. Restrictive atrial communication and severe tricuspid regurgitation were more common in group of patients with bad outcome but statistic significance was not proved.

P-136
Early diagnosis of transplant coronary artery disease by intravascular ultrasound (IVUS) improves the outcome of pediatric heart transplant recipients

Tissot C. (1), Pietra B.A. (1), Boucek M.M. (2), Gilbert D.J. (1), Campbell D.N. (1), Mitchell M.B. (1), Ivy D.D. (1), Miyamoto S.D. (1)
The Children's Hospital, Denver, Colorado, US (1); Joe DiMaggio Children's Hospital, Hollywood, Florida, US (2)

Introduction: Transplant coronary artery disease (TCAD) represents the leading cause of late graft loss after pediatric heart transplantation and the major cause of retransplantation. The diffuse nature of the disease and coronary remodelling renders early diagnosis difficult. The incidence of TCAD is related to the method of surveillance.

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Intravascular ultrasound (IVUS) allows detection of TCAD at an earlier stage compared to angiography.
Methods: We retrospectively reviewed pediatric heart transplant recipients who had IVUS at the time of routine surveillance catheterization. IVUS was performed in the left anterior descending coronary artery in children whose weight was ≥25 kg. Abnormal IVUS was defined as intimal thickening ≥0.3 mm according to the Stanford classification. Patients with TCAD were separated into 2 groups: (1) angiography diagnosis of TCAD and (2) normal angiography with IVUS diagnosis of TCAD.
Results: 141 IVUS procedures were performed in 57 of 310 transplanted children. Of 36 patients diagnosed with TCAD, 10 (28%) had normal angiography but abnormal IVUS. There was no difference between the groups in ischemic time, age at transplantation, number of rejections and prior IVUS exams. Weight at transplant was lower in the angiography group (17 ± 17 vs. 29 ± 8.5kg, p ≤ 0.01). Weight at TCAD diagnosis was higher in the IVUS group (60 ± 20 vs. 40 ± 19kg, p ≤ 0.01), reflecting the limitation of the procedure in small patients. There was a trend towards shorter time from transplant to diagnosis of TCAD in the IVUS group (6.9 ± 3.9 vs. 7.6 ± 3.5 years, p = ns). Eight patients died or required retransplantation as a consequence of TCAD in the angiographic group (8/26 = 30%) as opposed to one death and no retransplantation in the IVUS group (1/10 = 10%). The time from TCAD diagnosis to death was shorter in the angiography group (1.1 ± 1.8 vs. 4.3 ± 3.7 years, p ≤ 0.01).
Conclusion: IVUS was able to detect early TCAD in 18% of patients with normal angiography in the population suitable for IVUS, the biggest limitation being the size of the patient. Shorter time to death in the angiography group may reflect TCAD diagnosis at a later stage of the disease. Early TCAD diagnosis by IVUS is associated with improved outcome and decreased need for retransplantation.

P-137

Increased rotation compensates for impaired longitudinal and radial deformation in children with hypertrophic cardiomyopathies

Laser K.T., Palacios Argueta J.R., Knyphausen E., Crespo-Martinez E., Görg R., Kretschmer A., Kececioglu D.
Clinic for congenital heart defects, Heart and Diabetes Center North Rhine-Westphalia Bad Oeynhausen, Germany

Background: The echocardiographic deformation pattern of patients with excentric hypertrophic cardiomyopathies (HCM) is an interesting field for echocardiographic tissue deformation studies in order to characterize their cardiac function. Until now a decrease in basal longitudinal deformation could be demonstrated in tissue doppler studies, however, there is less information about radial, circumferential deformation and rotation. We tested the hypothesis if rotation which is mostly caused by contraction of oblique muscle fibers is increased to preserve cardiac function in this patient group.
Methods: 10 patients with HCM (5 obstructive, 5 non-obstructive, 4y-17 y) were compared with body-surface area-matched healthy children. Global and regional longitudinal, radial and circumferential strain and strain-rate as well as rotation were calculated by 2d-strain speckle-tracking (Echopac, GE). The net difference between angular displacement of the apical and basal short axis segments was calculated. Maximal torsion (deg/cm) was determined as the maximal value normalized by LV-length. Further echocardiographic data including stroke volume, enddiastolic volume and muscular mass were measured to characterize control- and patient-group.
Results: Patients with HCM showed decreased longitudinal strain values in the basal, midseptal and basal lateral segments ($p < 0{,}05$). Regional analysis of the short axis views at papillary muscle level revealed decreased radial as well as increased circumferential strain and strain-rate values in the hypertrophic septal and anterior segments ($p < 0{,}05$).
We found significantly increased maximal torsion in patients with HCM (3.1 deg/cm ± 1.4 vs. 1.7 deg/cm ± 0.7, $p < 0{,}01$).
Conclusions: Patients with HCM have a decrease in longitudinal and radial deformation in the hypertrophic areas. Conversely maximal torsion that expresses rotational function is significantly increased. As the localized hypertrophy is an impediment to radial and longitudinal deformation, increased twisting might be a compensatory mechanism to preserve left ventricular function.

P-138

Reduced systolic right ventricular longitudinal and left ventricular radial deformation in children after arterial switch operation

Laser K.T. (1), Palacios Argueta J.R. (1), Görg R. (1), Kretschmer A. (1), Böttler P. (2), Schlensak C. (3), Kececioglu D. (1)
Clinic for congenital heart defects, Heart and Diabetes Center North Rhine-Westphalia, Bad Oeynhausen, Germany (1); Department of pediatric cardiology, University Freiburg, Germany (2); Department of cardio-thoracic surgery, University Freiburg, Germany (3)

Background: At present the arterial switch operation (ASO) is the treatment of choice for correction of transposition of the great arteries with good short and midterm results. In the longterm follow up the influence of coronary transfer on cardiac function is not yet entirely understood. As the assessment of systolic global and regional cardiac function of both ventricles is facilitated by echocardiographic speckle-tracking techniques we applied this method to our patients after ASO.
Methods: 22 patients with transposition of the great arteries without ventricular septal defect after ASO (age: 1.5-16.5, median 6.5 years) were examined by echocardiography and compared with age-matched controls. Left ventricular longitudinal, radial, circumferential and right ventricular longitudinal deformation were calculated by 2d-strain speckle-tracking (Echopac, GE) using 4 chamber and short axis views.
Results: Right ventricular global longitudinal strain (−19.4% ± 4.8 vs. −26.2% ± 3.7) and strain-rate (−1.1/s ± 0.35 vs. 1.65/s ± 0.34) were reduced in the patient group ($p < 0.001$). Left ventricular longitudinal and circumferential deformation parameters were not significantly different whereas global radial strain (30.2% ± 10.8 vs. 42.1% ± 14.9) and strain-rate (1.4/s ± 0.25 vs. 1.8/s ± 0.38) were reduced after ASO ($p < 0.05$).
Conclusions: Although most patients are doing extremely well after ASO a reduction of predominantly systolic right ventricular function can be demonstrated. The impairment of left ventricular function seems to be less significant and can be observed by a decline in radial function in this patient group. Longitudinal and circumferential deformation are not altered. Great deviations from normal parameters could be used to initiate further diagnostic steps as cardiac catheterization.

P-139

Normal values for aortic diameters in children and adolescents - in vivo assessment by contrast-enhanced MR angiography

Valsangiacomo Buechel E.R. (1), Kaiser T. (1), Albisetti M. (2), Bergsträsser E. (3), Kellenberger C.J. (4)

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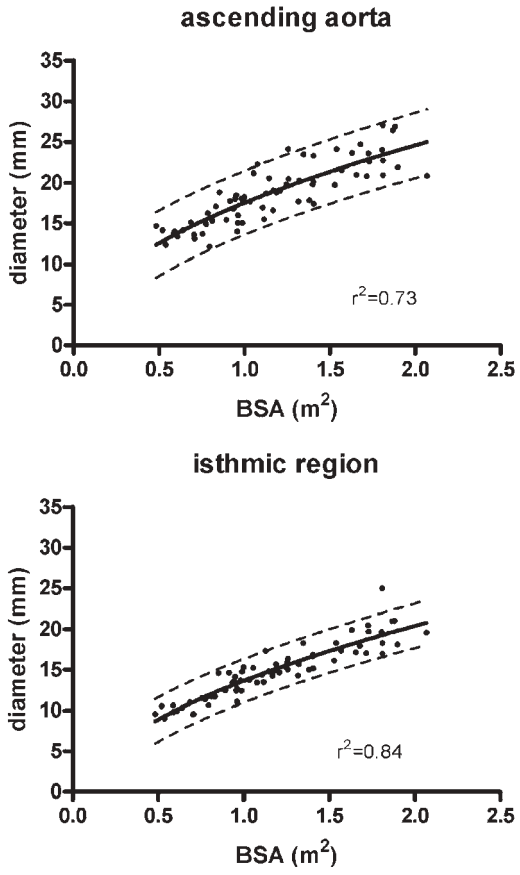
Division of Pediatric Cardiology, University Children's Hospital, Zurich, Switzerland (1); Division of Pediatrics, University Children's Hospital, Zurich, Switzerland (2); Division of Pediatric Oncology, University Children's Hospital, Zurich, Switzerland (3); Division of Diagnostic Imaging, University Children's Hospital, Zurich, Switzerland (4)

Introduction: Contrast-enhanced MR angiography (CEMRA) is being increasingly utilised for diagnosis, planning and follow-up after interventions in children with aortic arch anomalies. We sought to establish normal values of the diameters of the thoracic aorta and reference curves for body growth in children using CEMRA.
Methods: CEMRA was performed in 69 children without cardiovascular disease. Median age was 10 years (range 2–20 y), weight 32 kg (10–83), height 140 cm (81–188). Aortic diameters were measured at nine standardised sites (aortic sinus, sinotubular junction, ascending aorta at the level of right pulmonary artery, proximal to the brachiocephalic artery, proximal transverse arch, distal transverse arch, isthmus, proximal descending aorta and at the level of the diaphragm) on maximum-intensity projections images in longitudinal and cross-sectional views. Regression analysis in relation to body surface area (BSA) and height was performed and normative curves created. The limits of agreement between measurements from longitudinal and cross-sectional planes were calculated.
Results: Best fitting model was linear regression of the diameters to the square-root of BSA. Regression analysis curves with prediction bands for 95% confidence intervals are shown in Figure 1 for selected locations. Longitudinal measurements tended to minimally underestimate the diameters compared to cross-sectional measurements (mean difference −0.16 mm ± 0.34).



Conclusions: Normative curves for aortic diameters in children measured by CEMRA are presented. These are the first normal data available for CEMRA and can be used for diagnosis and planning treatment of aortic disease in children.

P-140

Automatic Functional Imaging : a new tool for assessment of myocardial function in children

Stos B. (1), Labombarda F. (2), Rein A. (3), Bonnet D. (2), Boudjemline Y. (2)
Centre Chirurgical Marie Lannelongue, Le Plessis Robinson, France (1), Hopital Necker Enfants Malades, Paris, France (2), Hadassah-Hebrew University Hospital, Jerusalem, Israel (3)

Background: Automatic Functional Imaging (AFI) is a novel echocardiographic modality, based on 2-dimensional gray scale images analysis, which allows the assessment of global and segmental myocardial function (strain and strain rate).
Objective: To compare AFI and Tissue velocity imaging (TVI) in children with normal heart.
Methods: Data from 43 healthy children who had an echocardiogram for innocent heart murmur were reviewed. Mean age was 6.7 years (0-13 years). Data could be analysed in all cases for left ventricular function and in 70% of cases for right ventricular function. Velocity, strain and strain rate were calculated for each myocardial segment and considering ventricles as single segment (global function). TVI was used to compare velocities measurements to those obtained by AFI.
Results: Mean AFI myocardial systolic velocity at the basal segment of left ventricle, septum and right ventricle was respectively 7.8, 5.1, and 10 cm/s. Same measurements by TVI were consistent for these segments (mean 6.5, 5.1, 9.8 cm/s, respectively). Segmental strain values showed an increasing gradient from the base to the apex with a mean of −17.2 ± 4% at the left basal segment to −26.3 ± 4.6% at the apex lateral wall, and a mean of − 29.2 ± 8.9 to −36.7 ± 8.6% at the right lateral wall. For global function, global strain and strain rate were −21.8 ± −2.6% and −1,35 ± 0.19 s-1 for the left ventricle and −33.7 ± 6.4% and −2.1 ± 1 s-1 for the right ventricle. None of AFI based parameter correlated with age.
Conclusion: AFI is a rapid and reproducible technique that provides a new method for assessment of segmental and global myocardial function. Strain and strain rate values are not correlated with age. This tool can be of great interest in paediatric cardiology, particularly when assessment of cardiac function is limited with conventional modalities, for example in the evaluation of right ventricular function after tetralogy of Fallot repair.

P-141

Intra- and interventricular synchrony in paediatric patients with single-site right versus left ventricular pacing

Tomaske M. (1), Breithardt O.A. (2), Bauersfeld U. (1)
Division of Paediatric Cardiology, University Children's Hospital Zurich, Switzerland (1); Department of Cardiology, Klinikum Coburg, Germany (2)

Introduction: Right ventricular (RV) pacing may cause dyssynchronous left ventricular (LV) contraction and dysfunction. Likewise, LV-based pacing could influence ventricular synchrony. We evaluated LV and RV synchrony with LV versus RV pacing in children.
Patients and Methods: Patients with normal cardiac anatomy and single-site epicardial RV apex (RV pacing, n = 9) or LV free wall

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pacing (LV pacing, n = 10) for congenital complete heart block were prospectively enrolled. Median age/ median pacing duration was 8.6/5.7 years for RV and 9.2/ 4.0 years for LV paced children. Conventional echocardiography, myocardial 2D strain (2Ds) analysis (RV-mechanical delay: longitudinal 2Ds in the apical 4-chamber; LV-mechanical delay: circumferential 2Ds in the parasternal short axis) as well as timing of systolic velocity peaks were obtained. Data are given as mean ± SD.

Results: Paced QRS duration did not differ between groups. Interventricular mechanical delay and intraventricular synchrony by the septal to posterior wall motion delay was preserved with LV pacing. Accordingly, LV-mechanical delay demonstrated LV synchrony with LV but not RV pacing. RV-mechanical delay did not differ. Timing for systolic velocity peaks in the basal segments differed between groups for the LV, but not for the RV or septum. Moreover, the maximum difference (Δ) of measured timing for systolic velocity peaks between LV and septum was narrow with LV pacing. LV ejection fraction was normal with LV but not RV pacing.

Conclusion: Conventional, 2Ds and SVP echocardiographic measurements indicate regular intra- and interventricular synchrony and LV function in paediatric patients with single-site LV, but not with RV pacing. LV pacing does not influence RV synchrony.

		LV pacing	RV pacing	p-value
Paced QRS duration (ms)		173 ± 18	170 ± 16	NS
Interventricular mechanical delay (ms)		17 ± 13	67 ± 12	p < 0.0001
Septal to posterior wall motion delay (ms)		56 ± 26	286 ± 93	p < 0.0001
LV-mechanical delay (ms)		84 ± 38	170 ± 41	p = 0.0003
RV-mechanical delay (ms)		79 ± 18	78 ± 43	NS
Systolic velocity peaks	LV (ms)	165 ± 39	247 ± 33	p = 0.0004
	RV (ms)	209 ± 52	194 ± 47	NS
	Septum (ms)	182 ± 35	214 ± 37	NS
ΔSystolic velocity peaks, LV -Septum (ms)		43 ± 17	59 ± 12	p = 0.039
LV ejection fraction (%)		55 ± 10	44 ± 6	p = 0.012

NS = not significant.

P-142

Right ventricular dilatation in severe pulmonary regurgitation late after repair of tetralogy of Fallot - How fast does it progress?

Valsangiacomo E.R. (1), Kaiser T. (1), Kellenberger C.J. (2), Dodge-Khatami A. (3), Bauersfeld U. (1)
Division of Pediatric Cardiology, University Children's Hospital, Zurich, Switzerland (1); Division of Diagnostic Imaging, University Children's Hospital, Zurich, Switzerland (2); Division of Congenital Cardiovascular Surgery, University Children's Hospital, Zurich, Switzerland (3)

Introduction: Pulmonary valve replacement (PVR) in severe pulmonary valve regurgitation after repair of Tetralogy of Fallot (TOF) is recognized as a crucial intervention for preserving right ventricular (RV) function. However the ideal timing for PVR is still subject of debate.

We sought to assess the natural course of progression of RV dilatation in patients with pulmonary regurgitation after TOF repair.

Methods: 22 patients underwent two magnetic resonance (MR) examinations for evaluation of RV volume and function within a median time interval of 25 m (9–45 m). TOF repair was performed at a median age of 20 m (range 2 m–9 y) and consisted of insertion of a transannular patch in 10, a monocusp homograft patch in 7, a valved conduit in 3, and a pulmonary valvotomy with supravalvular patch in one patient.

Age at first MR examination was 12 y 9 m (5–26 y); the time interval since surgical repair 11 y (4–23 y).

Ventricular volumes were measured on short-axis steady-state free precession images.

Results: RV volume was larger in patients with transannular patches (enddiastolic volume RVEDV 171 ± 32 ml/m2) than in patients with monocusp valved patches (145 ± 18 ml/m2) or valved conduits (144 ± 26 ml/m^2) (p < 0.05).

The overall mean ventricular volumes and ejection fractions did not change significantly over time (table 1).

Table 1: ventricular parameters

	MR 1	MR 2
RV EDV (ml/m^2)	155 (±30)	153 (±30)
RV ESV (ml/m^2)	73 (±14)	72 (±15)
RV EF%	48 (±6)	47 (±6)
LV EDV (ml/m^2)	73 (±13)	75 (±13)
LV ESV (ml/m^2)	34 (±7)	34 (±9)
LV EF%	54 (6)	55 (6)

Progression of RV dilatation occurred in 4/22 patients, while RVEDV remained unchanged (accepted variability 8%) in 11, and decreased in 7. In 11 patients with severe RV dilatation (RVEDV > 150ml/m^2), RVEDV increased in 3, remained unchanged in 4 and decreased in 4. The overall mean rate of RV volume change was 0.1 ± 12ml/m^2/year, without correlation neither to the technique of repair nor to the degree of initial dilatation.

Conclusions: During a follow up time of 2 years, there was no significant progression of RV dilatation in children, adolescents and young adults. Change rate in ventricular volume did not correlate to the technique of repair or to degree of RV dilatation. A more conservative approach to PVR may be discussed on the base of these results.

P-143

Dobutamine Stress MRI in post repair Tetralogy of Fallot with severe pulmonary regurgitation: Preliminary results.

Parish V. (1), Bell A. (1), Head C. (2), Rosenthal E. (2), Greil G. (1), Razavi R. (1), Beerbaum P. (1)
King's College, London, UK (1); Guy's and St Thomas' NHS Trust, London, UK (2)

Introduction: In Tetralogy of Fallot (TOF), surgical repair is undertaken in childhood but standard repair techniques result in pulmonary regurgitation (PR). Although pulmonary valve replacement has been shown to improve symptoms, appropriate timing for this procedure continues to be debated. This study aims to evaluate dobutamine stress magnetic resonance imaging (MRI) in the assessment of right ventricular (RV) contractile reserve in post repair TOF with significant PR.

Methods: Patients with repaired TOF and PR referred for cardiac MRI were invited to participate. Ventricular volumes (2D cine MRI) pulmonary artery and aortic flows were obtained at baseline and during dobutamine infusion at 10 and 20 mcg/kg/min. Data comparison was performed using the student t-test (p < 0.05).

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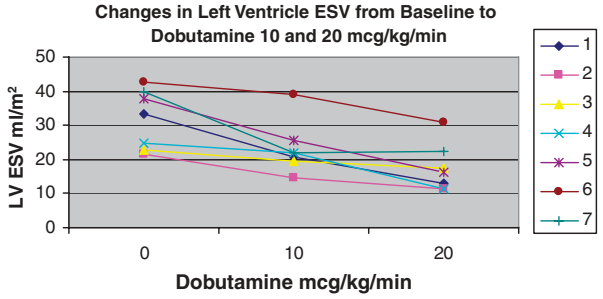
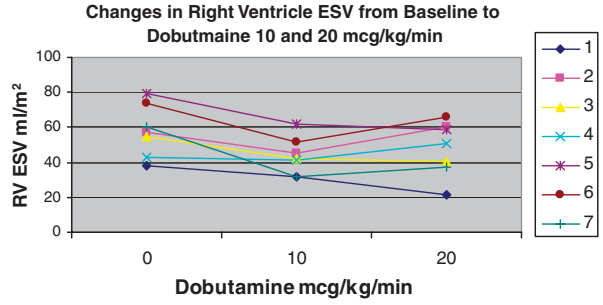
Results: To date 14 TOF patients were prospectively enrolled in the study (mean age at MRI 32.6 ± 15 years, mean age at surgical repair 6.1 ± 5.8 years). Preliminary data revealed at baseline, mean RV end diastolic volume (EDV) 133 ± 19 ml/m^2, mean RV ejection fraction (EF) 53 ± 6 % and mean pulmonary artery regurgitant fraction 45 ± 9 %. 10 patients completed 10 mcg and 7 completed 10 and 20 mcg dobutamine. During stress imaging heart rate (cardiac index) increased from 67 ± 9 bpm (2.9 ± 0.4 l/min/m²) at baseline to 89 ± 19 bpm (3.9 ± 1.1 l/min/m²) at 10mcg dobutamine and 120 ± 23 bpm (4.7 ± 1.1 l/min/m²) at 20mcg dobutamine ($p < 0.01$). In the left ventricle (LV), there is a clear decrease of LV EDV and LV end systolic volume (ESV) at 10 and 20mcg dobutamine. In contrast, for the RV there is less decrease of RV EDV and RV ESV from baseline to 10mcg dobutamine, and no significant further reduction of end systolic volume is seen at 20mcg dobutamine.
Conclusions: Our preliminary data demonstrates a diagnostic potential of dobutamine stress MRI in patients with TOF and severe PR. Whereas the LV clearly reduced EDV and ESV at each dobutamine level, the RV showed much less response at 10mcg dobutamine and failed to reduce or even increased RV ESV at 20mcg dobutamine. RV end systolic volume under stress may thus become a discriminative parameter in post repair TOF patients with relevant PR if confirmed in a larger population.





P-144

Normal values of biventricular function, volumes, and mass in children aged 8 to 17 years. A magnetic resonance study using steady-state free precession

Robbers-Visser, D. (1), Boersma H. (1), Helbing W.A. (1)
Erasmus MC, Rotterdam, The Netherlands

Introduction: Normal values for biventricular function, volumes, and mass assessed with current cardiovascular magnetic resonance (CMR) imaging sequences are lacking for the pediatric population. This limits optimal use of CMR imaging in the follow-up of pediatric cardiac patients.
Methods: 60 healthy children were included and divided in 3 groups of 20 children (10 boys) according to age: 8–11, 12–14, and 15–17 years. A short axis set of contiguous slices was acquired with CMR imaging employing a steady-state free precession sequence. End-diastolic volume (EDV), end-systolic volume (ESV), ejection fraction (EF), and mass of the left ventricle (LV) and right ventricle (RV) were determined. Uni- and multivariable linear regression analyses were performed to study the interrelation of age, sex, and body surface area (BSA) on biventricular EDV, ESV, and mass. We report regression coefficients and percentage explained variance (R2). A coefficient of variation was calculated for intra- and interobserver variability.
Results: EF did not differ between boys and girls in all age groups (mean LV-EF 69 ± 5 (SD)%, mean RV-EF 65 ± 5%). BSA had good (EDV, mass) and modest (ESV) correlation with biventricular measurements (see table). Sex appeared a significant modifier of these relations, whereas age had no independent contribution. Intra-observer variability was between 2.1 and 6.2% for these variables. Interobserver variability was between 3.4 and 13.9%.

	boys	girls	R^2 BSA only	R^2 full model
LV-EDV (ml)	= 112.7 * BSA – 49.4	= 78.3 * BSA – 10.2	0.729	0.792
LV-ESV (ml)	= 38.9 * BSA – 20.1	= 18.7 * BSA – 5.3	0.486	0.568
LV-mass (gr)	= 127.6 * BSA – 83.2	= 60.5 * BSA – 3.4	0.654	0.814
RV-EDV (ml)	= 125.0 * BSA – 59.4	= 83.9 * BSA – 15.2	0.683	0.782
RV-ESV (ml)	= 30.7 * BSA – 31.0	= 24.6 * BSA – 0.7	0.498	0.655
RV-mass (gr)	= 38.2 * BSA – 22.6	= 19.8 * BSA – 0.6	0.534	0.636

Conclusions: assessment of biventricular volumes, function, and mass with CMR imaging employing steady-state free precession, reveals adequate gender-specific normative data in children aged 8-17 years. These data can be used as reference data in the follow-up of pediatric cardiac patients.

P-145

Differential analysis of pulmonary vascular resistance, endsystolic- and enddiastolic ressure-volume relations in Fontan patients: Its diastolic dysfunction that limits cardiac reserve

Falkenberg J., Ovrouski S., Ewert P., Rahmanzadeh P., Maarouf N., Schmitt B., Riesenkampff E., Bodhey N.K., Berger F., Kuehne T.
Dept. for Congenital Heart Disease and Pediatric Cardiology, German Heart Institute Berlin, Germany

Background: Parameters of myocardial contractility (ESPVR), diastolic compliance (EDPVR) and pulmonary vascular resistance (PVR) are essential determents of cardiac function. MRI was shown to accurately measure PVR and ESPVR. In this study we validated a new MRI method for EDPVR acquesition. Subsequently ESPVR, EDPVR and PVR were measured in patients with Fontan circulation for differential analyis of cardiac function.
Methods: The MRI method for assessment of EDPVR, ESPVR and PVR was based on ventricular pressure recording synchronized with ventricular volume and/or pulmonary/aortic blood flow measurement. Validation part: In 6 pigs MRI and conductance-catheter derived EDPVR (gold standard) was compared using Bland-Altman test. Clinical part: Parameters of global pump function as well as ESPVR, EDPVR, PVR were measured with MRI at rest and under dobutamine stress in 13 patients with univentricular physiology and Fontan circulation as well as in 5 controls (patients with patent foramen ovale, healthy hearts).
Results: Bland-Altman-Analysis showed good agreement between conductance-catheter and MRI method for EDPVR assessment. In Fontan patients diastolic compliance was substantially lower compared to controls ($p < 0.05$). Dobutamin-stress further increased

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the EDPVR (0.07 ± 0.02 to 0.1 ± 0.07, $p < 0.01$). In contrast, PVR slightly decreased during stress (4.9 ± 3.4 to 3.2 ± 1.8, $p < 0.05$). Further comparison of rest versus dobutamine showed increase of cardiac index, which however was mainly due to higher heart rates but not augmented stroke volumes. In conjunction, the ESPVR did not significantly increase during stress.
Discussion/Findings: The presented MRI method enables a reliable measurement of EDPVR as adjunctive to global parameters of pump function, ESPVR and PVR. The application of this technique in Fontan patients evidenced impaired ventricular adaption to stress (in terms of the Starling mechanism). This was mainly due to limited inotropic competance (myocontractility) and diastolic dysfunction due to reduced (compliance. A raise of PVR as possible reason for impaired diastolic filling was excluded.

P-146
Teleradiologic network for central analysis and archiving of MR images: A three-years experience of a cardiovascular science-association
Kuehne T. (1), Mueller S. (2), Sarikouch S. (3), Beerbaum P. (3), Franke D.(1), Lange P. (4) , Sax U. (2)
Abteilung für Angeborene Herzfehler - Kinderkardiologie, Deutsches Herzzentrum Berlin, Germany (1); Abteilung Medizinische Informatik, CIOffice Medizinische Forschungsnetze, Universitätsmedizin Göttingen, Germany (2);Klinik für Angeborene Herzfehler, Ruhr-Universität, Herz- und Diabetes-Zentrum Bad Oeynhausen, Germany (3); Kompetenznetz Angeborene Herzfehler, Zentrale, Berlin, Germany (4)

Background: Analysis of cardiovascular MR images requires increasingly high standards and produces more and more costs for specially trained medical personal and technical equipment. In the German network of competence a science-association was founded in which MRI data were acquired in peripheral intitutes and than send by teleradiologic means to a core-labaratory for central data analysis and archiving. The first three years of operation of this network were evaluated regarding its technical course of activity, accuracy of MRI data analysis and costs.
Methods: 16 cardiovascular institutes form part of the network. MR images are acquired in these institutes (Philips, Siemens, GE scanners) using a standardized protocol. Images are send by teleradiologic means (pseudonomyzed via internet) to a central core-laboratory. There, images are analysed for parameters of cardiac function using custom-made software. Variability of the measured parameters is continuously determined (quality assessment). Thereafter, analysed data and original MR images are archived and are accessible to the participating institutes by remote-data-entry-system (RDE). Economic evaluation of the network was done by cost-analysis and ex post-contemplation under consideration of investments, labor- and consumption-costs.
Results: Data transmission speed was approximately 300 kbit/s upstream. MR images arrive at the core-laboratory as pseudonymized DICOM-data. After the pilot phase, no errors in image transmission (e.g. incomplete data sets) were observed. Therefore, all MRI scans that were acquired using standardized protocols could be analysed in the core-labaratory, so far more than 900 scans. Interobserver variability of quantitative function parameters was significantly lower when analysis was done in the core-labaratory, compared to individual analysis in the peripheral institutes. MR images were ubiquitously accessible in the RDE-system and via the internet at any time. Ex-post evaluation showed an expenditure of 55 EUR per MRI data set when 552 sets per year are enrolled under scientific (non economic) conditions to the core-labaratory.
Conclusion: A teleradiologic network was successfully established in a science-association of congenital heart diseases and optimized for primary health care and scientific purposes. The first three-year experiences showed that this network is attractive by reducing labor-, investment- and consumption costs as well as for efficient and accurate data management.

P-147
The utility of enalapril treatement in anthracyclines-induced cardiac injury in children with malignancies-preliminary study
Mandric C. (1), Dimitriu A.G. (1), Miron I. (1) Dimitriu L. (2)
University of Medicine and Pharmacy, Iasi, Romania (1); Medical Center MEDEX, Iasi, Romania (2)

Background: Treatement with angiotensin converting enzyme (ACE) inhibitors in children with malignancies who received high doses of anthracyclines have been shown useful in order to reduce manifestations of anthracyclines-induced cardiotoxicity .
Purpose: study of effects of enalapril treatment on anthracyclines-induced echocardiographic change in children with malignancies.
Methods: we performed, a prospective clinical study comparing enalapril to placebo treatement in 30 survivors of pediatric cancer (aged beetwen 6–14 years) from the Department of Pediatric Hemato-Oncology from "Saint Mary" Children Hospital Iasi, treated with certain therapeutical protocols for hematological malignancies, who included anthracyclines- doxorubicin or adriamicin; all of this patients had at least one cardiac abnormality identified at any time after anthracyclines exposure; those chidren were devided in two groups: group A - 10 children treated enalapril (dose range beetwen 0.2–0.5 mg/kgc/d); it was performed regular echocardiographic examinations during enalapril therapy, mean age at cancer diagnosis - 6 years, mean median follow-up since the start of enalapril, 16 month; group B placebo - 20 children treated with similar doses of anthracyclines, without enalapril treatment, after completion of doxorubicin therapy. Cardiological evaluation was performed at baseline and at 3, 6, 12, 16 month after initiation of enalapril/placebo therapy.
Results: Over the 16 month of enalapril therapy, there was progressive improvement in left ventricular(LV) dimension end-systolic and end-diastolic, fractional shortening, LV mass, LV per cent posterior wall thickening, interventricular per cent septal thickening, Tei index, comparative with evolution of the same ecocardiographic parameters of the controller group, who was constant or it worsened in course of follow-up
Conclusions: Enalapril treatment improved anthracyclines-induced ecocardiographic changes in patient included in study, justifing precocious introduction of enalapril in children with cardiotoxocity anthracyclines induced.

P-148
MRI evaluation of biophysical proprieties of ascending aorta in bicuspid aortic valve
Ait-Ali L. (1,2), Aquaro G. (2), Bernabei M. (1), Recla S. (1), Kull S. (1), Foffa I. (1), Mariani M. (1), Festa P. (1,2)
"G.Pasquinucci" Hospital CNR, Massa,Italy (1); MRI Lab Institute of Clinical Physiology- CNR, Pisa, Italy (2)

Introduction: Abnormal biophysical proprieties of ascending aorta in bicuspid aortic valve (BAV) have already been demonstrated also by magnetic resonance (MRI). However MRI findings show large variations between different series.
Aim: to detect aortic wall biophysical properties in young subjects with BAV, by new MRI indexes referred as Relative Velocities of

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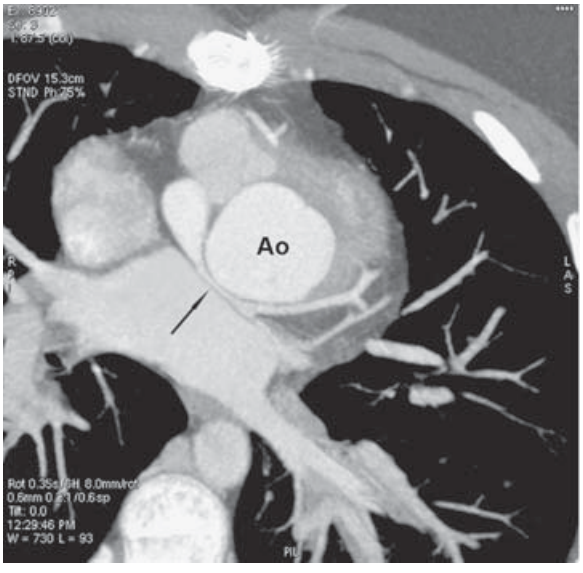
Maximal Systolic Distension (RVSD) and Relative Velocities of Maximal Diastolic Recoil (RVDR).
Methods: we enrolled 23 consecutive young subjects with BAV, without significant aortic valve dysfunction, aged from 9 to 21 years old (mean 17 ± 5 y.o.), and ten healthy volunteers age and sex matched as control group. All the subjects underwent MRI exam comprehensive of phase-velocity-mapping acquisition in order to assess aortic flow and cross sectional area (CSA) excursion with high temporal resolution (approximately 10–3 sec), at several aortic level Systemic pressure was non-invasively measured at each acquisition. CSA, systemic vascular resistance (SVR), aortic wall distensibility and flow wave velocity propagation (FVP) have been evaluated at each location as previously described. The variation in percentile of maximal CSA was measured by the difference CSA between each cardiac phase and the preceding (percentile of maximal area/10–3 sec). RVSD was defined as the highest ascending aorta CSA systolic augmentation in percentile and RVDR as the highest diastolic reduction CSA in percentile.
Results: the ascending aortic CSA was significantly larger in BAV compared to control (796 ± 312 vs. 514 ± 146 mm2 $p = 0.01$) with a significantly lower distensibility (5.3 ± 2.8 vs. 8.3 ± 3.8 103 mmHg-1 $p = 0.01$). FVP resulted significantly higher in BAV than in control (1.8 ± 0.75 vs. 1.26 ± 0.4 mm/msec. $p < 0.05$). RVSD was significantly lower than in control (4.2 ± 1.12 vs. 8.9 ± 2 %tile of maximal area/10-3 sec $p < 0.0001$, as well as RVDR (−4.39 ± 1.3 vs. -8 ± 3 %tile of maximal area/10-3 sec $p < 0.0001$) independently of SVR and without overlapping between BAV and control.
Conclusion: RVSD and RVDR assessed by MRI, have high sensitivity and specificity to detect aortic wall abnormalities in young patients with BAV. Further longitudinal study could help to early identify BAV subjects prone to develop progressive ascending aortic dilatation.

P-149
Afterload reduction in children and young adults with left ventricular obstructive lesions by interventional cardiac catheterization is followed by a decrease in rotation
Laser K.T., Haas N.A., Schäffler R., Palacios Argueta J.R., Görg R., Kretschmer A., Kececioglu D.
Clinic for congenital heart defects, Heart and Diabetes Center North Rhine-Westphalia, Bad Oeynhausen, Germany

Background: In addition to longitudinal and radial deformation rotation is another mechanism that contributes to left ventricular function. It is predominantly influenced by the contraction of oblique muscle fibers. Torsion describes the maximal difference of angular rotation between basis and apex of the heart. We tested the acute effect of afterload reduction by relief of left ventricular obstructions on torsion.
Methods: 17 patients (4–20 years, 5 female, 12 male) with re-coarctation (13) and valvular aortic stenosis (4) were examined by echocardiography before and 4-6 hours after successive interventional therapy. Left ventricular longitudinal, radial, circumferential deformation as well as rotation were calculated by 2d-strain speckle-tracking (Echopac, GE). Maximal torsion (deg/cm) was determined by the peak value of the difference between apical and basal rotational values normalized to left ventricular size. The indication of therapy considering re-coarctation was a gradient of more than 20 mmHg under medical provocation with orciprenalin.
Results: Invasively measured systolic gradients at rest dropped from 27.8 mmHg ± 17.4 to 8.5 mmHg ± 15.2 ($p < 0.001$) during interventional therapy. We did not see significant changes in longitudinal, radial and circumferential strain and strain-rate values. Maximal torsion dropped significantly after therapy (1.89 deg/cm ± 0.76 vs. 1.68 deg/cm ± 0.75, $p < 0.05$) and correlated with a decrease in values ($p < 0.001$, $r = 0.91$). The effect could not be explained by a mild increase in cardiac frequencies (73 bpm ± 19 vs. 81 bpm ± 23).
Conclusions: Acute reduction of afterload is followed by a decrease in rotation of the left ventricle despite of only moderately elevated resting gradients before interventional therapy. The high load-dependency of maximal torsion contrasts the lack of changes in longitudinal and radial deformation parameters in this group of children. Thus an increase in rotation seems to be a compensatory mechanism in left obstructive lesions.

P-150
Interest of 64-slice CT in the understanding of the mechanisms of the coronary lesions after arterial switch operation for transposition of the great arteries
Bonnet D., Khraiche D., Bajolle F., Agnoletti G., Vouhé P., Raisky O., Boudjemline Y., Sidi D., Ou P.
M3C-Pediatric Cardiology, Necker-Enfants Malades, Paris, France

Background: Late death after arterial switch (ASO) for transposition of the great arteries (TGA) is mainly due to myocardial ischemia caused by severe lesions of transferred coronary arteries.
Lesions that appears secondarily are often due to extrinsic compression or stretching of arteries at ostial or proximal segments by the surrounding structures. Aim of study: to assess if there is any correlation between the reimplantation site and the occurrence of coronary lesions after ASO.
Methods and Results: We performed 64-slice CT coronary to 147 children who underwent an ASO. This examination enabled us to assess the relative position of the reimplantated ostia in relation to the great arteries: angle between reimplantated ostia and the axe formed by great arteries annuli and distance between each ostium and pulmonary bifurcation. Left ostium anterior location favors occurrence of left coronary artery lesions (62 degrees in the patient group without coronary artery lesion versus 80 degrees in the patient group with coronary artery lesion; $p = 0{,}026$). Long distance between right ostium and pulmonary bifurcation (that is to say: low implantation of the ostium of the retro aortic circumflex in the right sinus) increases the occurrence of retro-aortic



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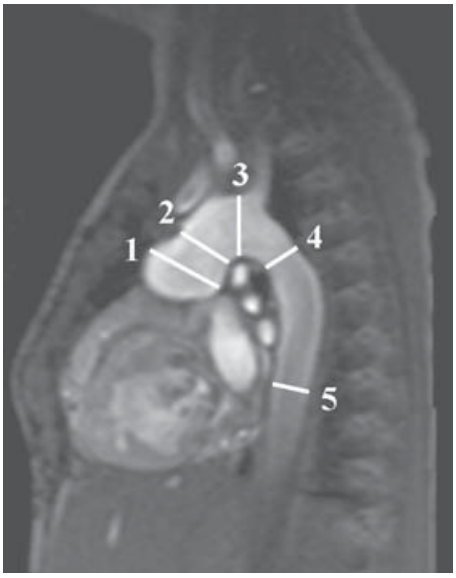
circumflex artery lesion (mid-distance of 16 mm in the group of patient without coronary artery lesion versus mid- distance of 24 mm in the group of patient with retro-aortic circumflex artery lesion; $p = 0.031$).
Conclusion: several ostia locations are associated with a higher rate of coronary artery lesion. According to growth, these coronary anomalies might progress during life.

P-151
Quantification of Mitral Valve Regurgitation in a Pediatric Population by Real-Time Three-Dimensional Echocardiography
Acar P., Latcu G., Paranon S., Dulac Y., Abadir S.
Pediatric Cardiology, Children Hospital, Toulouse France

Purpose: Real-time three-dimensional echocardiography (RT3DE) allows accurate left ventricular (LV) volumetric measurements without any geometric assumptions in adult patients. Our purpose is to validate RT3DE by measuring the stroke volume in a normal pediatric population and to calculate regurgitant volume in patients with mitral regurgitation (MR).
Methods: Thirty-nine pediatric patients (aged one week to 16 years, median 6 years), with normal left ventricular outflow tracts and no ventricular septal defects (29 without MR and 10 with MR) had 2D echocardiography coupled with a RT3DE volumetric acquisition of the LV. Stroke volume was calculated by the Doppler method at the aortic annulus (SVD). End-systolic and end-diastolic volumes of the LV were measured with the semi-automated method (Qlab, Philips). Three-dimensional stroke volume (SV3D) was calculated as their difference. Mean time for measuring SV3D was 1 minute in patients with good endocardial detection and 3 minutes when manual corrections were needed. In the MR group regurgitant volume was calculated by the PISA method (VRPISA) and as the difference between SV3D and SVD (VR3D). Regurgitant fraction was evaluated by the two methods (RFPISA and RF3D respectively).
Results: Measurements feasibility was 89 %. In the normal pediatric patients group, SV3D (27.9 ± 18.1 ml) was highly correlated with SVD (30.7 ± 19.6 ml): $r = 0.98$, $p < 0.0001$, $y = 0.90x + 0.08$). Mean difference was 2.8 ± 3.8 ml. The correlation was highly significant in both subgroups of patients with good endocardial detection (12 patients) and in those needing manual correction (13 patients), but was slightly better when no endocardial contour correction was needed ($r = 0.97$ and 0.94 respectively). In the MR group, VRPISA (20.7 ± 16.9 ml) and VR3D (12.9 ± 11.1 ml) are well correlated ($r = 0.92$, $p < 0.001$). Regurgitant fraction values are as also well correlated by the two methods: RFPISA = 39.9 ± 16.8, RF3D = 32.8 ± 17.0, $r = 0.79$, $p = 0.006$. Conclusions. RT3DE is a simple, rapid and reliable method for evaluating stroke volume in children. Hence, its use may be of particular interest in evaluating regurgitant volume and fraction in MR. A larger population with volumetric overload (MR or ventricular shunt) is needed to reliably assess feasibility in this group.

P-152
Aortic anatomy and aortic distensibility in children with hypoplastic left heart syndrome studied by cardiac magnetic resonance imaging (MRI) at 3 tesla.
Voges I. (1), Hart C. (1), Jerosch-Herold M. (2), Helle M. (1), Hansen T. (1), Kramer H.-H. (1), Rickers C. (1)
Department of Paediatric Cardiology, University Hospital, Kiel, Germany (1); Harvard Medical School, Boston, USA (2)

Introduction: The Norwood operation is the standard procedure of palliation in patients with hypoplastic left heart syndrome (HLHS), but little is known about the anatomy and distensibilty of the neo-aorta and its effects on right ventricular (RV) function in later life. Echocardiography and cardiac catheterisation have been the standard diagnostic modalities. However, the sensitivity of echocardiography for detection of stenosis of the neo-aorta is low and catheterisation is associated with considerable morbidity. Therefore we thought to evaluate aortic anatomy, aortic distensibility and RV ejection fraction (EF) in children with HLHS using MRI.
Methods: 32 patients (mean age 66.1 ± 34.7 months, age range 2 month - 14 years) with HLHS after Norwood-operation underwent an MRI study at 3 Tesla. Cine-MRI (TR/TE/$\alpha = 4.4/2.5/15°$) and high resolution contrast enhanced time-resolved MR angiography using a 3D keyhole technique with centric re-ordering (TR/TE/$\alpha = 2.4/0.93/15°$) were performed in all patients.



The MRI images were used for measuring diameters of the neo-aorta (at 5 levels, figure) and for determination of aortic distensibility (at 4 levels). RV-EF was assessed with a gradient-echo sequence in the short-axis plane. The measured diameters were compared to normal values generated by echocardiography and cardiac catheterisation, and distensibility was compared to normal values acquired by MRI.
Results: Diameters of the aortic root (mean 25.0 ± 4.6 mm), the ascending aorta (mean 21.5 ± 4.0 mm), the aortic arch (mean 17.3 ± 4.0 mm) and the descending aorta (mean 12.7 ± 3.8 mm) were significantly increased in HLHS patients. The aortic isthmus (mean 9.8 ± 3.1 mm) was significantly smaller. Distensibility was decreased in the ascending aorta (4.3 ± 2.8 vs. 8.6 ± 2.2 10-3 mmHg-1) and the aortic arch. Furthermore reduced distensibility in the ascending aorta was significantly correlated with decreased RV-EF ($r = 0.47$, $p < 0.05$).
Conclusions: 1) With MRI we were able to demonstrate that in HLHS patients the aortic root, ascending aorta, aortic arch and descending aorta are significantly dilated and the aortic isthmus is significantly smaller as compared to normal values. 2) The distensibility of the neo-aorta as a parameter of aortic function is inversively correlated to RV-EF.

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P-153

Improved decision making for ASD closure by surgery or catheter intervention with routine use of real time 3D echocardiography (RT3DE) – 2.5 years experience.

Razek V. (1), Schneider P. (1), Dähnert I. (1), Riede F.T., Kinzel P. (1), Kostelka M. (2), Janousek J. (1)
Department of Pediatric Cardiology, University of Leipzig, Heart Centre, Leipzig, Germany (1); Department of Cardiac Surgery, University of Leipzig, Heart Centre, Leipzig, Germany (2)

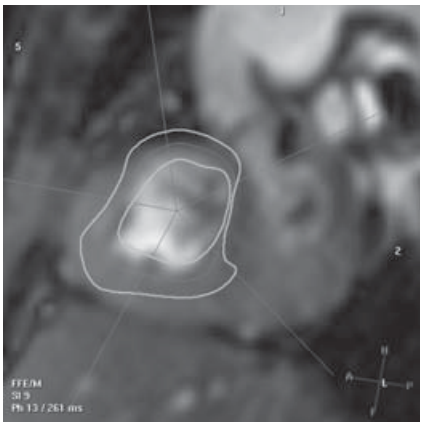
Introduction: The current routine method for decision on feasibility of secundum type ASD (ASDII) device closure is transoesophageal echocardiography (TEE). We hypothesized that transthoracic RT3DE is a sufficient alternative to TEE.
Methods: Between 07/2005 and 12/2007 RT3DE evaluation was performed in 89 consecutive pts referred with the diagnosis of ASDII. Indication for device closure was based on confirmation of ASDII anatomy, presence of the so-called posteroinferior margin (PM) and lack of ASDII size and patient's weight mismatch. Accuracy of the RT3DE diagnosis was confirmed by interventional closure feasibility and surgical findings, resp.
Results: RT3DE selected 47 pts for interventional closure and 33 pts for surgery. 9/89 pts had an intact interatrial septum or very small communication. Reasons for referral to surgery were sinus venosus superior defect (N = 10), ASDII with posterorinferior extension and missing margin (N = 21) and multiple ASDII (N = 2). In all 47 pts selected for interventional procedure RT3DE findings were in accordance with TEE. Device closure was successful in 40/47. In 6/47 pts with "good" PM interventional closure was not successful because of ASDII size and patient's weight mismatch and in 1/47 pts the occluder dislocated. These 7 pts went to surgery. RT3DE findings were confirmed intraoperatively in all 40 pts treated surgically. Thus, RT3DE was 100% positive and negative predictive of the presence of PM and avoided 21/68 (31%) otherwise necessary TEE evaluations by routing patients without PM directly to surgery.
Conclusion: RT3DE is highly accurate for correct ASD diagnosis and delineation of PM which's presence is essential for successful device closure. RT3DE improves pre-interventional decision making and reduces the number of necessary TEE evaluations. Mismatch of defect size and child's weight remains a separate problem to be defined irrespective of the PM.

P-154

Assessment of right ventricular function and regional wall motion by high-field magnetic resonance imaging in children with hypoplastic left heart syndrome

Hart C. (1), Voges I. (1), Jerosch-Herold M. (2), Scheewe J. (1), Hansen T. (1), Helle M. (1), Kramer H.-H. (1), Rickers C. (1)
Kinderherzzentrum, Universität Kiel, Germany (1); Harvard University, Boston, USA (2)

Introduction: In HLHS right ventricular function is considered to be an important determinant of long term survival and morbidity. Cardiac magnetic resonance imaging (MRI) allows precise non-invasive assessment of ventricular function, volume and mass independent from geometrical assumptions. Therefore, the aim of the study is to evaluate global and regional ventricular function in children with hypoplastic left heart syndrome (HLHS) using MRI with respect to a remaining rudimentary left ventricle (LV).
Methods: 20 children (3.2–14.0 yrs) with HLHS after completion of total cavopulmonary anastomosis were studied with high field MRI (Achiva, 3 Tesla, Philips). Short axis fast gradient-echo-cine-sequences (TR/TE/α = 1.1/1.6/60; 260 × 280; 30 phases/R-R) were used to obtain tomographical views of the right ventricle (RV) and the rudimentary LV. Global and regional function, volumes and mass have been quantitatively analysed with dedicated software (ViewForum, Philips).
Results: Systemic RV-function of patients with HLHS was significantly reduced as compared to LV-function of healthy children (EF = 46.3 ± 10.4% vs. 61.4 ± 9.3%, $p < 0.05$; cardiac index = 2.5 ± 0.88 vs. 3.2 ± 0.5 l/m²/min, $p < 0.05$). Wall motion analysis showed that greater LV mass and RV-function were inversely related ($y = 2.4 * x^{-1} + 1.8$; $p < 0.05$). Regional wall motion abnormalities were most prominent in the septal area (see fig.).



Conclusion: In patients with HLHS systemic RV-function is significantly reduced as compared to LV-function of healthy children. Regional and global RV-function is inversely related to the mass of a rudimentary LV. Follow up MR-studies are necessary to assess long term outcome.

P-155

Single vs. Double Balloon for Percutaneous Mitral Valve Dilatation: a Finite Element Model and an Experimental Test

Schievano S. (1), Nicosia M.A. (2), Khambadkone S. (1), Cochran R. (3), Einstein D. (4), Kunzelman K. (3), Bonhoeffer P. (1)
UCL Institute of Child Health & Great Ormond Street Hospital for Children, London, UK (1); Department of Mechanical Engineering, Widener University, Chester PA, USA (2); Central Maine Heart and Vascular Institute, Lewiston ME, USA (3); Biological Monitoring and Modeling, Pacific Northwest National Laboratory, Olympia WA, USA (4)

Introduction: Percutaneous mitral valve (MV) stenosis dilatation is performed with either a single balloon (SB) or a double balloon (DB) technique. This study aimed to compare the results of these two techniques, using a finite element (FE) model and a purposely designed experimental test.
Methods: An established FE model was modified by fusing the leaflet edges at commissure level to simulate a stenotic MV. FE models of a 30mm SB (low-pressure, elastomeric balloon) and an 18mm DB system (high-pressure, non-elastic balloon) were created and inflated into the MV model. An experimental test was designed to validate the results from the FE analysis: two elastic strips were sewn in two points with thread to simulate the welded commissures. A 30mm Inoue balloon and an 18mm DB system were inflated inside the sewn strips to break the stitches. Different combinations of strips and sewing threads were used (Table).
Results: Both SB and DB simulations resulted in full balloon deployment (Figure), splitting of the commissures and consequent

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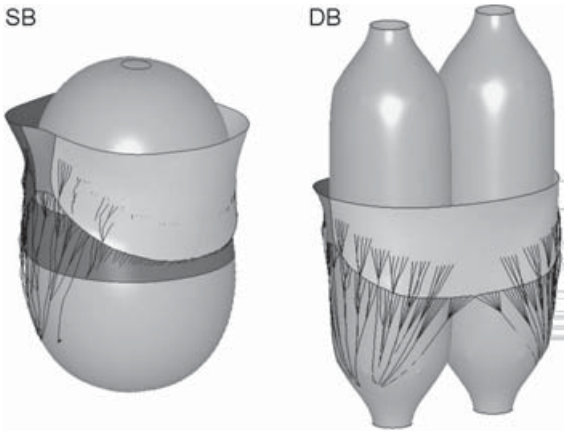
relief of the stenosis. Stresses induced by the two methods varied across the valve. At full inflation, SB showed higher stress in the central part of the leaflets and at the commissures compared to DB simulation, which demonstrated a more uniform stress distribution. This was due to the mismatch of the round shape of the SB within an oval mitral orifice. Due to its high compliance, commissural split was not easily accomplished with the SB. Conversely, the DB guaranteed commissural split even when a high force was required to break the commissure welds. The experiments showed that, depending on the strip/thread combination (Table), the SB could not break the thread, while the DB could.



Conclusions: FE analysis demonstrated that MV stenosis dilatation can be accomplished by both SB and DB techniques. However, the DB method resulted in lower stress values and a more even stress distribution, and therefore a lower risk of MV damage caused by overstretching. Furthermore, DB has a higher probability of success in splitting the commissures, which was confirmed by the experimental test results.

Break of the thread by balloon inflation		STRIP		
		high elasticity	medium elasticity	low elasticity
THREAD	weak	SB: no DB: no	SB: no DB: yes	SB: yes DB: yes
	strong	SB: no DB: no	SB: no DB: no	SB: no DB: yes

P-156

Early and mid- term results of stent implantation and re-dilation for native and recurrent coarctation of the aorta

Krasemann T., Bano M., Rosenthal E., Qureshi S.A.
Evelina Children's Hospital, Guy's & St Thomas NHS Foundation Trust, Westminster Bridge Road, London SE1 7EH, United Kingdom

Since 1993 stenting of native or recurrent coarctation of the aorta has been carried out in patients of more than 30kg in our institution.
Methods: 68 patients (24 female, weight 65.6+18.8 kg, <6 years: 1, 7–12 years: 7, 12–16 years 14, >17 years: 46) were treated with a coarctation stent for native coarctation (40/68) or recurrent coarctation (28/68, post surgery: 12, previous balloon-angioplasty: 4, post surgery + balloon-angioplasty: 24). In 23 patients a covered stent was implanted.
Results: After stent implantation the systolic blood pressure at the right arm decreased from 152.92 (+24.29) mmHg to 128.97 (+18.20) mmHg ($P < 0.0005$). The pressure drop estimated on echocardiography decreAsed from 47.19(+19.94) mmHg to 20.36 (+9.15) mmHg ($P < 0.0005$), the invasive gradient from 24.7 (+15.31) to 4.9 (+5.39) mmHg ($P < 0.0005$). The invasive poststenotic systolic blood pressure increased from 80.3 (+14.62) mmHg to 100.6 (+17.68) mmHg.
Reinterventions to dilate the stent were carried out in 26 patients 20.28 (range 2.6–85.7) months after primary intervention) and the results were sustained. Complications during the procedures were rhythm disturbances (2), inguinal haematoma (2), aneurysm of the femoral artery (1), stent migration (2) and stroke (1) in a patient with known bilateral stenosis of the carotid arteries. There were no late aneurysms detected. 59 patients were followed in the outpatients (mean 16.5 months, 3–59.7 months), 20 after reintervention (mean 37.5 months, 7–91.2 months). 51% remained clinically hypertensive, but less than prior to the intervention. The echo gradient was lower than before the (re) stent implantation.
Discussion: Stents are not fully dilated during the initial procedure to avoid complications. This explains the relatively high re-intervention rate. Covered stents are used to avoid aneurysms. Possible reasons for the persisting arterial hypertension are discussed.

P-157

Transcatheter Patch Correction of Non-Secundum Atrial Septal Defects: Patient Selection and Clinical Results

Sideris E.B., Chamie F., Macuil B., Haddad J., Kalahanis M., Toumanides S.
Athenian Institute of Pediatric Cardiology, Athens, Greece

Non secundum atrial septal defects (ASDs) include sinus venosus and ostium primum ASDs; they are traditionally corrected by surgery. Sinus venosus (SV) defects have deficient superior rim associated with partial anomalous veins in over 50% of cases; ostium primum (OP) defects have deficient inferior rim and various degrees of mitral insufficiency. The Transcatheter Patch (TP) could conceivably repair SV and OP ASDs providing that pulmonary veins are normal and there is no significant mitral insufficiency.
In 2 patients with SV ASD, 10 and 70 years old, test balloon occlusion of the ASD and simultaneous right pulmonary artery angiography were performed. Normal pulmonary venous return and SV ASD sizes of 23 and 25 mm, respectively, were found.
Two patients with OP ASD and insignificant mitral regurgitation, 7 and 26 year old, underwent test balloon occlusion of their defects. Both the sizing balloon and TP supporting balloon were placed over a wire. In the SV cases and the first OP case, the wire was placed through the ASD into the pulmonary vein. In the second OP case, for improved stability, the wire was placed through the ASD into the left atrium, the left ventricle, and the aorta, essentially forming a loop. Both defects measured 20 mm.
The 2 SV and the 2 OP ASD patients underwent successful TP occlusion under transesophageal echocardiography and fluoroscopy. The patches were released in 48 hours in 3 patients. In the fourth case a rapid release method using surgical adhesives allowed for release in 20 hours.
All defects were effectively occluded without complications. No mitral insufficiency or pulmonary vein abnormalities were noticed. All patients were doing well on follow up (range: 1 month – 4 years).
In conclusion TP occlusion of non secundum ASDs is feasible in selected cases. Trans-esophageal echocardiography, test balloon occlusion and angiography are important for the patient selection. The procedure has been simplified by over the wire placement of the patch and through the use of surgical adhesives.

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P-158

Redilation of ePTFE covered CP stents

Butera G., Gaio G., Chessa M., Piazza L., Negura D., Arcidiacono C., Micheletti A., Carminati M.
Pediatric Cardiology and Guch Unit, Policlinico San Donato IRCCS, San Donato Milanese - Italy

Introduction and Objectives: There are no data in the literature about the redilation of ePTFE covered CP stents. We aimed to evaluate the possibility to redilate covered Cheatham-Platinum stents during follow-up, in particular in growing children with aortic coarctation.
There are no data in the literature about the redilation of ePTFE covered CP stents.
Methods: Sixty covered CP stents were implanted in patients with aortic coarctation or re-coarctation between January 2004 and October 2007. Thirty subjects were aged less than 18 years. Seven patients needed to repeat the hemodynam study due to somatic growth and increase of aortic gradient with the occurrence of systemic hypertension. Two had near-atretic aortic coarctation, three had post-surgical recoarctation and aneurysm formation, one had native aortic coarctation associated with aneurysm of the arterial wall, one had severe native aortic coarctation.
Results: Procedures were performed a median of 24 months (range 12-24 months) after the primary stent implantation. Fluoroscopy time ranged between 7 and 15 minutes (median 10 minutes) while procedure time ranged between 60 and 75 minutes (median 65 minutes). After re-dilation the gradient across the stenosis decreased from a median value of 35 mmHg to a median value of 5 mmHg. The stent diameter increased of 20 to 50 % the pre-dilation value. No complications occurred and angiographic controls showed that the stenoses have been relieved.
Follow-up: During a median follow-up of 12 months (6-30 months) the results were stable without complications.
Conclusion: ePTFE Covered Cheatham-Platinum stents can be easily redilated during follow-up.

P-159

Late results of balloon pulmonary valvuloplasty in the management of pulmonary valve stenosis depending on its severity in children.

Rudzinski A., Werynski P., Krol-Jawen W. Kordon Z., Kuzma J.
Department of Paediatric Cardiology Polish-American Children's Hospital Collegium Medicum, Jagiellonian University Cracow, Poland

Since 1982, balloon pulmonary valvuloplasty (BPV) has been the major method of pulmonary valve stenosis treatment (PVS) in children. The objective of the study was to assess BPV results depending on PVS severity, especially with respect to restenosis, tricuspid and pulmonary valve regurgitation (TVR, PVR) in late follow-up. The material consisted of 137 patients (76 M and 61 F) subjected to BPV when $x = 5.3 \pm 4.8$ years old in 1988-2004. Newborns with critical PVS were excluded. ECG, CXR and echocardiography were performed in all the children and repeated in follow-up, which was $x = 6.1 \pm 3.4$ years. All the patients subjected to hemodynamic and angiocardiographic studies were divided into three groups depending on relation of right ventricular systolic to systemic pressure: 1/. (N = 58) - ≤0.75, 2/. (N = 41) - 0.76≤1.0, and 3/. (N = 38) - >1.0. Four Group 2 and six Group 3 pts showed dysplastic PVS, in which the ratio of balloon diameter to pulmonary annulus was significantly higher than in pts without these lesions (1.42 ± 0.1 vs. 1.31 ± 0.1, $p < 0.002$). The table presents results of BPV evaluation before, immediately after the procedure and in late follow-up.

Group	Before/after BPV hemodynamic ECHO					Late follow-up ECHO		
	RVSP	∇PVS	PASP	TVR >II°	PVR >II°	∇PVS	TVR >II°	PVR >II°
	mmHg x/±					mmHgx/±		
1	65.3±10.3	49.3±11.1	15.8±1.1					
	28.6±7.6	12.5±7.6	16.8±0.9	0	3.4%	13.4±6.9	5.1%	17.2%
2	91.7±11.6	75.6±12.3	15.8±1.2					
	35.0±14	17.0±13.0	17.8±1.3	7.3%	0	16.9±12.1	4.8%	24.4%
3	133.3±27.3	117.3±28.0	14.5±1.3					
	38.4±19.2	38.4±19.2	19.4±2.1	23.6%	2.6%	17.1±12.2	13.1%	39.5%

Only one dysplastic PVS patient from Group 2 required surgery. Restenosis was noted in seven pts (5.1%), $x = 6.1$ years after the first procedure. Repeated BPV was performed in five pts, two were operated on. Conclusions: BPV may be effective in PVS also when dysplastic lesions are present and in rare (~5%) restenosis cases. The incidence of TVR decreases, while PVR significantly increases in Group 3 pts ($p < 0.007$), what may be important in late follow-up.

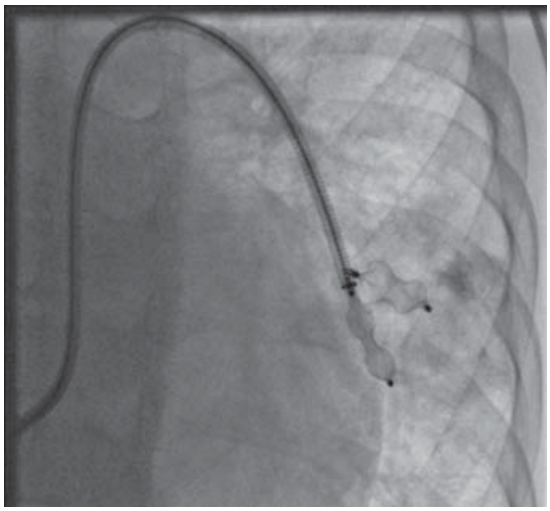
P-160

Amplatzer vascular plug application in cardiovascular lesion in children

Zunzunegui J.L., Ballesteros F., Alvarez T., Siles A, Medrano C., Camino C., Maroto E., Brochet G.
Gregorio Marañon Hospital, Madrid. Spain

Introduction or Basis or Objectives: Evaluation of efficacy of the Amplatzer Vascular Plug (AVP). We describe our experience in wide range of vascular closure indications in children Methods: From Janury 2005-December 2007 76 patients (age 50 days-17 years) underwent embolization using AVP in our center. A total of 109 devices (range diameter 4–16 mm) were implanted using 5-6 f. Guide catheters in 7 diferents patients group:
1. Aortopulmonary collaterals ; 43 patients.
2. Veno-Veno/Atrial Collaterals in Glenn physiology; 12 patients.
3. Bilateral Pulmonary Arteriovenous fistula; 1 patient.
4. Hepatic tract after transhepatic access; 1 patient.
5. Pulmonary Sequestration; 1 patient.
6. Blalock-Taussig fistula; 1 patient, asociated with coils.
7. Spermatic vein embolization; 17 patients.

Results: No major complications arose during the procedures. A complete closure of target vessel was obtained within 20 minutes.



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During the first 6 months we found recovered patency through the device in 3 re-evaluated patients, all of them were on aspirin and/or clopidogrel therapy.
Conclusions: The AVP is an effective transcatheter occlusion device in the embolization of a wide variety of vascular lesions. It can be implanted through 5–6 guide catheter, this low profile is really useful in small patients and tortuous vessels. It could be used like a "coil constrainer" decreasing the risk of coil embolization (BT Shunts). Spontaneous recanalization is possible in arteries, specially in children on antiplatelet therapy.
Two devices in arteriovenous fistula

P-161
Catheter interventional closure of atrial septal defects in small children
Daehnert I., Razek V., Weidenbach M., Hambsch J., Riede F.T.
Herzzentrum, University of Leipzig, Germany

Introduction: Device closure of secundum type atrial septal defects (DC-ASD) is a routine procedure in (mostly asymptomatic) older children. However, little is known about the safety and outcome in symptomatic younger children. Therefore, we evaluated results of DC-ASD in this subset of patients.
Methods: Retrospective analysis of hospital/outpatient records 2001 to 2007. All children indicated for DC-ASD with a body weight of less than 10kg and/or less than 2 years of age were included.
Results: DC-ASD was attempted in 16 children. Median age, weigth and length were 11 months (range 3–28), 7kg (range 3.5–12.0), and 67cm (range 48-87), respectively. Indications for intervention were failure to thrive (5), respiratory distress (6), pulmonary hypertension (3) and cyanosis (2). Concomitant problems were: restrictive ventricular septal defect (3), ductus arteriosus (PDA) (2), cardiomyopathy (1), palliated pulmonary atresia/intact ventricular septum (1), scimitar syndrome with functional single lung due to pulmonary sequestration (1), bronchopulmonary dysplasia (3), bronchial stenosis (1), tracheomalacia following esophagotracheal fistula (1), omphalocele (1), hydronephrosis (1), cerebral malformation (1), anal atresia (1), trisomy 21 (2) and unclassified syndrome (1).
Results: Defect size ranged from 5 to 19mm (median 8). The intervention was performed under fluoroscopic and echocardiographic guidance. Successful deployment of Amplatzer septal occuders was feasible in all patients (device size median 9mm (range 5–20). There were 4 additional interventions: PDA-closure (2), closure of surgical shunt (1), occlusion of sequester-feeding arteries (1). Median procedure and fluoroscopy time were 114 min (range 58–215) and 19.9 min (range 0.8–29.2), respectively. Early complications: iliac vein occlusion (1) and temporary femoral artery obstruction requiring prolonged heparinization (2). At discharge small residual interatrial shunts were seen in 3 (19%). Follow-up is median 46 months (range 1-85). There was no residual shunt at one-year follow-up (0/13) nor late complications. All children treated for failure to thrive showed improved growth, patients treated for respiratory distress showed improvement of symptoms. Two of three patients with pulmonary hypertension are still on treatment for elevated pulmonary resistance.
Conclusion: Indications for DC-ASD in small children include a wide spectrum of clinical symptoms and associated morbidities. DC-ASD offers a safe and effective alternative to surgery for selected patients.

P-162
Chronical Testing of a New Hybrid Technique for Closure of mVSDs in a Pig Model
Kozlik-Feldmann R. (1), Lang N. (1), Lehner A. (1), Aumann R. (1), Sodian R. (2), Rassoulian D. (2), Schmitz C. (2), Wintersperger B. (3), Sigler M. (4), Freudenthal F. (5), Hinterseer M. (6), Vasilyev N. (7), Del Nido P. (7), Netz H. (1)
Dpt. of Pediatric Cardiology, University of Munich, Germany (1); Dpt. of Cardiac Surgery, University of Munich, Germany (2); Institute of Clinical Radiology, University of Munich, Germany (3); Dpt. of Pediatric Cardiology, Georg-August University Goettingen, Germany (4); Kardiozentrum, La Paz, Bolivia (5); Internal Medicine I, University of Munich, Germany (6); Dpt. of Cardiac Surgery, Harvard Medical School, Boston, USA (7)

Objective: Therapy for muscular ventricular septal defects (mVSD) beyond the moderator band has always been challenging for surgeons and cardiologists, especially in newborns and infants. Hybrid therapy is a new innovative approach, but there are still some limitations. Recently, we established a new hybrid therapy for patch closure of mVSDs. Here, we tested our new hybrid therapy in a chronical design.
Methods: 12 pigs underwent anterolateral thoracotomy to expose the left ventricle. mVSDs were created under 2D and 3D echocardiographic guidance (Philips IE 33 Utrasound Systems) with a 7.5 mm sharp punch instrument, which was forwarded via a left-ventricular incision. In the same session patch closure of the created mVSDs was undertaken with our patch system in hybrid technique. After chest closure all animals were extubated and observed for at least three months. All pigs underwent echocardiography and cardiac catheterization after 1 and 3 months to detect residual shunts and to assess pulmonary artery pressure (PAP) and left-ventricular function. One animal received an MRI scan after 3 months. Finally all hearts were explanted and a gross pathologic evaluation was done.
Results: Creation of mVSDs was successful in all 12 animals. Location of the defects was midmuscular (n = 9), apical (n = 1), inlet (n = 1) and anterior muscular (n = 1). Closure of mVSDs was successful in 9 out of 12 animals. One animal died after a pneumothorax at postinterventional day 14. The other 8 animals survived for 3 months without postoperative complications. Echocardiographic controls showed no damage to the valves. Development of left-ventricular dysfunction was not seen confirmed by echocardiography. Hemodynamic measurements revealed a slight increase in the PAP after 3 months. There were no residual shunts after 3 months except in one animal. MRI revealed septum dyskinesia in the area of the patch. Gross pathologic examination showed complete endothelization of the patch and the nitinol anchors without protruding parts of the system.
Conclusions: Our new hybrid therapy is efficacious in closing mVSDs. The patch system as well as the nitinol anchors showed a good integration into the septum. Cardiac function was not significantly reduced. Further development of the system seems to be worthwhile.

P-163
The Helex Septal occluder for atrial septal defects closure: a single center experience
Spadoni I., Lunardini A., Ait-Ali L., Cantinotti M., De Lucia V., Rocca E., Giusti S.
Pediatric Cardiology and GUCH Unit, CNR, "G. Pasquinucci" Hospital, Massa, Italy

Objectives: We retrospectively reviewed our experience of atrial septal defects (ASD) and patent foramen ovale (PFO) closure with the HELEX Septal occluder
Methods: Between June 2003 and October 2007, 56 pts (mean age 39 ± 13, range 6–73 years, mean weight: 67 ± 16, range 26–111 Kg),

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underwent percutaneous implantation of Helex devices for closure of atrial septal defects. The standard technique of atrial septal defects closure was used. During the follow-up period, patients were followed with tranthoracic echo, with contrastography (ce-TTE) in cases of PFO, at discharge, 1 month, 6 months, and every year after the procedure. In pts with PFO and residual shunt, ce-TTE and Transcranial Doppler was repeated every 6 months.
Results: Among 65 cases scheduled for implantation of the Helex occluder, in 9 cases (14%) the anatomy and/or the size of the defect was judged unsuitable for the Helex, and were treated with Amplatzer devices. The implantation of an Helex occluder was attempted in 56 pts, 41 with PFO, associated with fenestrated/multifenestrated aneurysm in 18 cases , and in 15 cases of ASD, 6 of which with multiple defects. Fluoroscopy time was 21 ± 11 (8–77) min. The procedure was successful in 52 pts (93%) and failed in 4: in 2 cases of PFO it was impossible to cross the defect with the first generation device) and in 2 with ASD a proper position/configuration of the device was not achieved. In 7 cases (12%), additional attempts were necessary. In 2 pts with PFO (4%) the device was interventionally removed for unwanted lock release and malposition.
In the PFO group the closure rate at ≥18 mths of follow-up (mean 2 ± 1.3 years, range 1 day- 4.3 years) was 96%. Only 1 pt with multiple ASDs has a small residual shunt at mid-term follow-up.
Conclusions: Despite some disadvantages (long learning curve, time-consuming procedure, fragile structure and increased cost), the Helex occluder has some important advantages,that is its low profile and compliant design. Moreover, in our experience the device demonstrates a high rate of complete closure even in patients with complex atrial septal anatomy.

P-164
Development and implementation of a pulsatile circulation model to train catheter based interventions in paediatric cardiology
Schill M., Schaeffler R., Sarikouch S., Kececioglu D., Haas N.A.
Department for Congenital Heart Defects, Heart and Diabetes-Centre Nordrhein Westfalen, Ruhr-University Bochum, Bad Oeynhausen, Germany

Introduction: Cardiac catheterization and catheter based interventions require extensive training and experience. Usually learning of new techniques is achieved by patient oriented training under the guidance of skilled supervisors with a variable learning curve. In technical and industrial areas however, computer or model based simulators are widely used to train and prepare new staff (i.e. flight simulators, ship simulators, etc.). Simulation of medical circumstances for training purposes or for updating specific skills is more and more accepted for medical purposes too (i.e. resuscitation training, anaesthetic setups, etc.). We have developed a model to simulate and train interventional therapy of aortic coarctations.
Methods: We used a Medos® device to achieve a pulsatile flow with realistic pressures and waveforms. The circuit was created by the use of silicone and polyvinylchloride tubing with various diameters. A realistic model of the aortic arch and various different forms of aortic coarctation based on catheter and MRI models was then built. We choose a simple membranous coarctation, a coarctation with a longer segment of narrowing, a coarctation close to the subclavian artery, and a coarctation between the left common carotid artery and the left subclavian artery with arch hypoplasia as anatomical variances in our model. Then balloon angioplasty as well as stenting of the stenotic area was performed by junior doctors in training.
Results: We could simulate a realistic circulatory model of aortic coarctation for training purposes. Junior doctors were able to train catheter based therapy of various forms of coarctations on multiple occasions. Especially the practical manual steps such as wire exchange, sizing, positioning of the balloons, stent preparation, positioning and deployment could be trained in this pulsatile model under realistic simulation.
Conclusion: This model may be a helpful tool to train interventional techniques. The simulation of other congenital heart defects (i.e. PDA, pulmonary and aortic valve stenosis, ASD) is planned and currently in development.

P-165
Results of stent implantation in patients with aortic coarctation
Brzezinska-Rajszys G. (1),, Zubrzycka M. (1), Jagiellowicz D. (2), Kosciesza A. (3), Ksiązyk J. (1), Rewers B. (1), Kansy A. (4), Kawalec W.(2)
The Children's Memorial Health Institute, Warsaw, Poland
Heart Catheterization Laboratory (1), Cardiology Clinic (2), Computed Tomography (3), Cardiosurgery Clinic (4)

Introduction: Stent implantation for coarctation of the aorta has been widely used for the last years. The aim of the study was to analyse the results in patients with native and postoperative coarctation of the aorta performed in The Children's Memorial Health Institute in Warsaw in the period 1998–2007.
Methods: Sixty eight patients with different narrowings of the aorta (native coarctation 36 pts, postoperative recoarctation 17 pts, dissection after balloon angioplasty 1, middle aortic syndrome 14 pts) were treated with stent implantation. The group of 50 pts with native (35 pts) and postoperative (15 pts) coarctation was analysed. One neonate with complex congenital heart defect and coarctation of the aorta and 2 neonates with severe recoarctation of the aorta after operation of interruption of the aortic arch were excluded form the analysis.
Results: The mean patient age was 13.6+/-3.1 yrs (range 5–20, median 14) respectively. The diameter of the coarctation increased from a mean 5.28±2 mm (range 2–11, median 5) to 16.2±2,44 mm (range 12–22, median 16) ($p < 0.001$). The systolic gradient across the coarctation decreased from a mean 32.6±12.29 mmHg (range 10–84, median 30) before to a mean of 2.9±5.07 mmHg (range 0–18, median 0) after the procedure ($p < 0,000001$). During mean follow-up period 3,06±3,1 yrs (range 0.25–9, median 3), 22 pts (44%) did not need antihypertensive treatment and arterial systolic pressure mean 124±13,08 mmHg (range 85–146, median 125), diastolic pressure mean 66,5±9,68 mmHg (range 40–80, median 66) were recorded in whole group. Stent redilation was performed in 5 pts during follow up period due to neointimal hyperplasia or patients somatic growth. Covered stents were additionally implanted in 3 pts due to stent fracture and in-stent stenosis, in 1 pt due to small aneurysm detected 4 yrs after stent implantation and in 1 pt due to aortic arch hypoplasia.
Conclusions: Stent implantation in native and postoperative coarctation of the aorta has good immediate and early results. Arterial hypertension therapy can be easily modified after stent implantation. During follow-up period indications for stents redilation and additional covered stent implantations may become apparent.

P-166
The Cp Stent For The Treatment Of Congenital Heart Diseases In Children
Agnoletti G., Ba H., Marini D., Boudjemline Y., Bonnet D.
Necker Enfants Malades, Paris, France

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Background: The aim of this work was to evaluate the effectiveness and safety of the Cheatham Platinum (CP) stent compared to Palmaz stent in treatment of native and postoperative lesions of congenital heart disease (CHD) patients.
Methods: From January 2003 to December 2007, 89 CP stents (34 covered and 55 bare) and 85 Palmaz stents were implanted in 77 and 75 patients, respectively. The number of stent implanted per patients was 1.15 in the CP group and 1.13 in the Palmaz group. CP and Palmaz stents were used to treat right ventricular outflow and pulmonary arteries in 52 and 62% of patients, systemic veins in 31 and 25%, aorta in 17 and 13%. The percentage of native lesions was 13% in the CP group and 18% in the Palmaz group.
Results: Patients were significantly older in the CP group (median age 13 years, range 4–18 and 8 years, range 1–18, $p < 0.01$). All stents could be deployed. Pre-dilatation was performed in 56% and 62% and post-dilatation in 29% and 13% ($p < 0.05$), respectively. Success of the procedure was achieved in 93% and 95% of patients.
Complications (including vascular dissection, stent migration and balloon burst) occurred in 11 and 24% of patients ($p < 0.01$). Age at catheterization was not related to the occurrence of complications. The incidence of dissection was significantly lower with CP than with Palmaz stent (6 and 1%, $p < 0.01$), the incidence of stent migration was similar in the two groups (8 and 9%). Balloon burst was never observed in the CP group. There were no procedural deaths. Urgent surgery was needed in 3 patients of each group.
Conclusions: The CP stent is a valuable tool to treat native and post-surgical lesions of CHD patients. Complication rate is low; in particular, vascular dissection is rare and balloon burst was never observed.

P-167
Use of a microcatheter in a telescopic system to reach difficult targets in complex congenital heart disease.
Gewillig M., Boshoff D.E., Mertens L., Eyskens B., Budts W.
Pediatric & Congenital Cardiology, Leuven, Belgium.

Backgroud: Some lesions can be very difficult to reach, especially if an initial angle exceeds 90°, and/or if multiple turns "protect" access to the target. Once reached, a guiding system needs to give sufficient support for balloons or stents to be deployed.
Methods: A "telescopic" system was created:

- a microcatheter system (Progreat® Terumo, Leuven, BE) consists of a 0.021" highly steerable 130cm Terumo guide wire within a 2.9Fr catheter. This system fits into any catheter allowing a 0.035" wire.
- the 2.9Fr was mostly used though a 4F catheter Ri coronary, Cobra or Mammaria.
- if an initial angle of >90° was required, the 4Fr catheter was deployed through a 6Fr (or larger)(Ri coronary or Mammaria) guiding sheath, cut-off 15 cm out of the groin but revalved with a short 6Fr introducer sheath;

During the approach and certainly once the target was reached, both the 2.9Fr and 4Fr catheter were advanced as much as possible; the guiding wire was replaced by a stiffer 0.014"wire though the 2.9Fr, and/or a stiffer 0.035"wire through the 4Fr catheter.
Patients & results: Since 2004, 75 microcather telescopic systems were used in 958 procedures (7.8%). The technique allowed to probe with a floppy steerable & exchangeable guide wire, to secure any gained position, and to exchange with a stiffer guide wire. Frequently 4–6 experienced "hands" were required to steer the system. Lesions/procedures where the telescopic system was highly appreciated included: complex peripheral pulmonary stenosis especially MAPCA's(14), coronary fistulae or side-branches (5), tortuous ducts with cumulated angulation of >270°(7), anterograde dilation of critical aortic stenosis (9), transvenous anterograde crossing of critical coarctation (2), crossing critical stenosed shunt for stent implantation (3), entering Amplatzer prosthesis cavity to coil embolise residual shunt (2). Once in place, the telescopic system allowed introduction of medication (2), embolic material (33), balloon dilation (14) or stent deployment (20).
The interventionalists felt that using the telescopic system had reduced procedure and scopy time; the technique made some "impossible" targets reachable in a safe way (5patients were referred after multiple failed catheterisations with conventional catheters).
Conclusion: The microcatheter telescopic system has become an invaluable tool to reach difficult targets, and to exchange for adequate guide-wires.

P-168
End-diastolic pulmonary artery forward flow in right ventricular outflow tract obstruction – intrinsic right ventricular restriction or reflection of underlying haemodynamics?
Lurz P., Nordmeyer J., Puranik R., Khambadkone S., Coats L., Derrick G., Cullen S., Bonhoeffer P., Taylor A.M.
UCL Institute of Child Health and Great Ormond Street Hospital, London for Children, UK

Objectives: Pulmonary artery forward flow (PAFF) in late diastole has been described in patients after surgical repair of Tetralogy of Fallot in the context of pulmonary regurgitation. This parameter is commonly believed to be a marker for right ventricular restriction. Here we describe our finding in patients with PAFF and pulmonary stenosis and the impact of PAFF on outcome after percutaneous pulmonary valve insertion.
Methods: Patients underwent percutaneous pulmonary valve implantation (PPVI) from 2002 to 2007. Patients were only included, if they had a predominant stenotic lesion (peak gradient of > 50 mmHg and mild to moderate regurgitation on echocardiography). PAFF was defined by an antegrade late diastolic forward flow throughout the respiratory cycle, as assessed on echocardiography. Invasive pressure measurements and cardiopulmonary exercise testing were performed in all patients before and after PPVI.
Results: Out of 52 patients with significant right ventricular outflow tract obstruction, PAFF was presented in 11 patients (21%). Patients with PAFF had higher invasively measured RVOT gradients (62.3 ± 23.0 vs. 42.2 ± 11.9 mmHg, $p < 0.001$), right ventricular end-diastolic pressures (14.1 ± 3.6 vs. 10.8 ± 4.0 mmHg, $p = 0.019$) and decreased exercise performance 20.3 ± 4.4 vs. 25.3 ± 7.4 ml/kg/min, $p = 0.052$) compared with patients without PAFF. There was a significant inverse correlation between mean PAFF velocity and exercise capacity ($r = -0.84$, $p = 0.004$). After PPVI, the phenomenon of PAFF disappeared in all subjects. In addition, on comparison of patients with and without PAFF before valve deployment, there were no longer statistically differences present in RVOT gradients (19.0 ± 11.5 vs. 19.2 ± 11.2 mmHg, $p = 0.95$), right ventricular end-diastolic pressures (9.9 ± 4.2 vs. 9.7 ± 4.7 mmHg, $p = 0.91$) and exercise performance (26.4 ± 8.3 vs. 25.8 ± 7.4 mmHg, $p = 0.84$) after PPVI.
Conclusion: PAFF in the context of stenosis is associated with higher RV filling pressures and impaired exercise performance. However, these findings are reversible after relief of pulmonary stenosis. We speculate, that PAFF is a reflection of underlying haemodynamics rather than a parameter of intrinsic RV restriction.

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P-169

10- to 17-Year Follow-up After Aortic Balloon Valvuloplasty

Radtke W. (1), Fischer G. (2), Bandisode V. (3)
Nemours Cardiac Center (1), Wilmington, DE, USA; Kinderkardiologie Uni-Klinikum SH (2), Kiel, Germany; MUSC Children's Hospital (3), Charleston, SC, USA

Balloon valvuloplasty has been the preferred treatment for aortic valve stenosis for 15 years but equivalent long-term follow-up data are not available. Previous studies revealed a high rate of re-interventions particularly early post procedure. It is unclear if the re-intervention rate changes during later follow-up. We have therefore retrospectively reviewed the records of all patients undergoing aortic balloon valvuloplasty between 1987 and 2002. No patients were excluded. Procedural information was collected from catheterization reports, follow-up information from outpatient and inpatient records, operative reports, echocardiographic and recatheterization reports. Of the total cohort of 137 patients undergoing valvuloplasty during this period 20 patients (14.6%) were lost to follow-up, 10 patients (8.5%) died 1 day to 3.5 months (median 18.7 days) after the procedure mostly secondary to severe endocardial fibroelastosis, and left ventricular dysfunction during the initial period when the Norwood operation was unavailable. 42 patients (35.9%) underwent surgery a median of 3.6 years after the initial procedure (16 d–9.4 yrs): Norwood (3), transplant (2), valvotomy (1), valve repair (10), replacement (5) and Ross (21). Repeat balloon valvuloplasty was performed in 11 patients (9.4%) a median of 2.1 years after the procedure (1 d–7.3 yrs). The cohort with more than 10 year follow-up consisted of 18 patients. Age and weight at the time of the procedure was not significantly different from the total cohort: 12.3 months (7 d–11.0 yrs) versus 9.8 months (0 d–29.0 yrs) and 8.5 kg (4.6–27.0 kg) versus 6.0 kg (1.6–94.6 kg). Similar to the total cohort, valvuloplasty reduced the pressure gradient from 75.9 mmHg (25–190 mmHg) to 28.6 mmHg (8–50 mmHg). Follow-up ranged from 10.3 to 17.2 years (median 13.5 years). At the latest follow-up, the peak Doppler gradient was 39.0 mmHg (0–111 mmHg). Aortic regurgitation was mild or less in 12/18 patients (67%). Three patients (17%) were on afterload reduction. There were no surgical or catheter interventions in the follow-up interval between 10 and 17 years.

The 10- to 17-year follow-up period after aortic balloon valvuloplasty appears to have a lower re-intervention rate probably because of the preceding negative selection.

P-170

Diminishing a shunt with a self-fabricated fenestrated Amplatzer device – various indications and modifications

Kretschmar O. (1), Roffi M. (2), Knirsch W. (1)
University Children's Hospital, Department for Pediatric Cardiology (1); University Hospital, Department for Cardiology (2), Zurich, Switzerland

Background: In specific high-risk patients with congenital heart disease a complete closure of an existing intracardiac defect is not possible for a variety of reasons. We report our experiences with an interventional approach for shunt reduction using various modifications of a self-fabricated Amplatzer device since 09/05 in our institution.

Results:

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Diagnosis	ASD II, PHT	ASD II, CoA	ASD II, PHT	AVSD, HLHS, s/p Fontan + fenestration	Criss-cross-heart, RV hypoplasia, s/p Fontan + shunt-fenestration
Age [years]	3.4	3.9	66	2.7	17
weight [kg]	18	14	102	14.1	34
Reason to close	Volume overload	Volume overload	Volume overload, LR- and RL-shunt	Cyanosis (SpO2 75%)	Cyanosis (SpO2 80%)
Reason to close partially	Systemic PHT	Restrictive LV	2/3 systemic PHT	PAP >= 15 mmHg	PAP >= 15 mmHg, PLE
Device modification	ASO (14mm) + dilated fenestration	ASO (10mm) + dilated fenestration	ASO (24mm) + sutured fenestr.	ASO (4mm) + stent (5mm)	AVP (10mm) + stent (4mm)
Immediate result	Fenestration open, restrictive LV	Fenestration open, normal LV function	Fenestration open, good RV and LV function	SpO2 90%, PAP 15mmHg	SpO2 91%, PAP 15mmHg
Follow-up [months]	9	28	1	20	28
Mid term result	Normal PAP, fenestration closed	Normal LV function, fenestration closed	-	SpO2 87%, fenestration open	SpO2 90%, fenestration open

ASD: Atrial septal defect; PHT: Pulmonary hypertension; CoA: aortic coarctation; AVSD: Atrioventricular septal defect; HLHS: Hypoplastic left heart syndrome; RV: Right ventricle; LV: Left ventricle; PAP: Pulmonary artery pressure; PLE: protein-losing enteropathy; ASO: Amplatzer Septal Occluder; AVP: Amplatzer Vascular Plug

Conclusions: In patients with an ASD and significant PHT and/or restrictive LV physiology and in Fontan patients with a large surgically created fenestration but failing Fontan circulation a fenestrated closure with a self-fabricated fenestrated Amplatzer device can be a feasible and successful therapy option. Balloon dilated fenestrations in the Amplatzer device tend to close spontaneously during follow-up. To ensure patency of the created holes non-resorbable sutures or stenting can be recommended.

P-171

Catheter interventions in the management of early postoperative Fontan problems

Bhole V., Stickley J., Reinhardt Z., Wright J.G.C., DeGiovanni J.G., Dhillon R., Miller P., Chikermane A., Desai T., Brawn W.J., Barron D.J., Jones T., Stumper O.
Birmingham Children's Hospital NHS Foundation Turst, Birmingham, United Kindom

Introduction: Results after the Fontan procedure have improved consistently in the past. However frequent early postoperative complications include prolonged pleural drainage, low cardiac output and systemic desaturation . We postulated that in such patients catheter interventions can be performed safely to improve haemodynamics and to prevent take-down or death after Fontan. The objective of this study was to **e**valuate the effectiveness and safety of transcatheter interventions in the early postoperative period after the Fontan procedure.

Patients and Methods: Over a 7year period (1.2000–10.2007) 227 patients underwent completion of the Fontan procedure. There were 2 immediate Fontan takedowns (0.8%) and two early deaths (0.8%). Catheter interventions were performed in 15 patients (6.6%) in the early postoperative period defines as 30 days from surgery.

Results: Fifteen patients underwent 18 catheter interventions within 30days (Median 12, range 2–29days) of the Fontan completion.

Eleven patients had catheter interventions for persisting chylothorax 14–27 (median 18 days) days post surgery. In 7 of these patients

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significant pulmonary artery stenosis was identified and was treated with a stent implantation. 3 patients had stenting of fenestration including one patient who had creation of fenestration. One patient underwent device occlusion of residual antegrade flow to the pulmonary artery. In all patients the chest drainage ceased 6-14 (median 7) days after intervention.
Stenting of fenestration was performed in 2 patients for low cardiac output within 72hours of surgery. One of these underwent additional pulmonary artery angioplasty. Both had improved cardiac output and survived.
One patient underwent pulmonary artery stenting and fenestration device closure within 72 hours after surgery for profound desaturation. There was marked improvement in saturations post intervention. There was no catheter related mortality. One patient suffered thromboembolic event post fenestration stenting but fully recovered.
Conclusions: Catheter interventions can be performed effectively and safely in the management of early postoperative Fontan complications. Stents can be used safely to treat early pathway obstruction. Catheter modification of fenestration can be achieved in patients with prolonged pleural effusions or low cardiac output state. The early application of these techniques has contributed to improve the overall success of the Fontan procedure.

P-172

Transcatheter therapy of transverse arch coarctation of the aorta with covered stents

Sadiq M. (1), Rehman A. (1), Qureshi S.A. (2)
Punjab Institute of Cardiology, Lahore. Pakistan (1); Evelina Children's Hospital, Guys trust, London. UK (2)

Background: Transverse arch coarctation of the aorta (Co A) in adults poses a special challenge of management. The risk of residual coarctation & aneurysm formation following surgery and risk of dissection or rupture following angioplasty are major concerns. Covered stents may be of value in this sub-group of patients.
Objectives: To evaluate the use of covered Cheatham-Platinum (CP) stents in the management of transverse arch Co A in adolescents & adults.
Methods: 27 covered CP stents were implanted as primary treatment (May03-Nov07) in at risk patients with complex anatomy Co A at a single tertiary center. Four such stents (4/27-15%) were implanted in 4 pts with transverse arch Co A. Three of these had coarctation involving the origin of left subclavian artery. In 3 pts the lesion was crossed via the femoral artery while in one, additional access via right radial artery was used. The balloon & stent size was selected by measuring the proximal arch and the isthmus.
Results: The patient ages were 14, 21, 25 & 28 yrs. The mean weight was 55 (45-75) kg. The systolic gradient across the CoA decreased from 49 ± 16 mmHg before, to 6 ± 3 mmHg after the procedure ($p < 0.0001$). The diameter of the CoA increased from 4.6- ± 2.9 mm to 14 ± 3 mm ($p < 0.0001$). It was possible to avoid covering the innominate & left common carotid artery in all but left subclavian artery was covered in 3 where it was part of the coarctation. This led to diminished flow & loss of pulse in left arm but well tolerated with no functional deficit. No other vascular complications or thromboembolism occurred.
At a mean follow up of 27 (3-47) months, no patient has needed re-dilatation or has shown evidence of chronic functional deficit in left arm. All stents were patent & in good position on CT performed 3-6 months later.
Conclusions: Covered CP stents may be used as the therapy of choice in pts with transverse arch Co A. Occlusion of left subclavian artery is well tolerated with no acute/chronic functional deficit. Covered stents provide a safe alternative to surgery, balloon dilatation or conventional stenting in these pts.

P-173

Reduction of Irradiation in Cardiac Catheterisation of Children with Congenital Heart Disease using New X-ray Technology

D'Orsogna L., Tiang T., Harle Y., Scott B.
Children's Cardiac Service, Princess Margaret Hospital, Subiaco, Western Australia

Introduction: X-ray generated imaging is routinely used in cardiac catheterisation and may be associated with prolonged irradiation exposure to children with congenital heart disease, especially during interventional procedures. New technology with flat panel detectors claims to provide optimum imaging with lower x-ray dosage including acquisition with fluoroscopy only.
Objective: To ascertain the total dosage of irradiation used in children undergoing selected interventional cardiac catheterisation procedures prior to and after the introduction of the Siemens Axiom Artis dBC biplane flat detector angiocardiography system.
Method: A comparative retrospective review of all the records of patients undergoing selective cardiac catheterisation interventional procedures at Princess Margaret Hospital since the introduction of the new Siemens system in February 2007. Total dosage of x-ray (cgy cm2) was recorded in each patient who had (Group 1) coil occlusion of patent ductus arteriosus (PDA), Amplatzer Duct occlusion of PDA (Group 2) and percutaneous balloon pulmonary valvuloplasty (Group 3) and compared to patients in each group who had procedures performed in 2006 using the older Siemens bi-plane imaging intensifier system.
Results: In 2006, 4 patients in Group 1 had mean dosage of 394 (range: 218-746) cgy cm2 compared to 4 patients in 2007 with mean dose 83.7 (71-99) cgy cm2. Eight patients in Group 2 in 2006 had mean dose 315 (101-990) cgy cm2 compared to 8 in 2007 with mean dose 188.9 (80-460) cgy cm2. There were 4 patients in 2006 from Group 3 with mean dose 203 (63-508) cgy cm2 compared to 7 patients in 2007 with mean dose 97.3 (48-248) cgy cm2
Conclusion: New technology in x-ray imaging for interventional cardiac catheterisation procedures in children with congenital heart disease is associated with significant reduction in irradiation. This dose reduction not only has potential health and safety benefits for the patient but also for the operator and staff involved in the procedures.

P-174

Potts anastomosis for severe pulmonary arterial hypertension in children

Serraf A., Petit J. (1), Belli E.(1), Humbert M. (2), Simonneau G. (2), Bonnet D. (3), Sidi D. (3), Vouhé P. (3).
Marie-Lannelongue Hospital, Le Plessis-Robinson, France (1); Hôpital Antoine Béclère, Clamart, France (2); Hôpital Necker Enfants Malades, Paris, France (3)

Introduction: Severe pulmonary arterial hypertension (PAH) has a very poor prognosis, particularly in symptomatic children. Functional capacity and survival have improved by novel medical therapies. However, refractory PAH remains a major problem in children. Eisenmenger's syndrome represents the end-stage of a uncorrected left-to-right shunt where PAH prohibits intracardiac repair. Survival in these patients is much better than that of

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idiopathic PAH. Converting normal heart physiology features to features typical of Eisenmenger's syndrome by means of atrial septostomy has been proposed, but patients with severe right heart failure and markedly elevated pulmonary vascular resistance do not tolerate atrial septostomy, because massive right-to-left shunting may result in insufficient pulmonary blood flow and severe hypoxemia.

Methods: In an attempt to transform children with severe PH to an Eisenmenger circulation type, avoiding suprasystemic right ventricular and pulmonary artery pressures (PAP), a Potts anastomosis, namely a direct anastomosis between left pulmonary artery and descending aorta was performed in 10 children (6 boys and 4 girls, mean age 107 ± 50 months). Three were born with transposition of the great arteries and had undergone successful neonatal surgery. All had suprasystemic PH with a systolic PAP of 134 ± 30 mmHg, and PVR of 24.0 ± 0.9UI/m2). All but one received at least 2 PAH specific drugs including epoprostenol, bosentan and sildenafil. Six-min walk distance was 170 ± 110m and all were in NYHA functional class III or IV. All procedures were performed through left thoracotomy without cardio-pulmonary bypass.

Results: There was 2 early deaths from multiple organ failure and sepsis. Post-operative course in the remaining 8 patients was uneventful and showed a SaO2 gradient between upper and lower limbs (97 ± 3 vs. 80 ± 9%). A mean follow-up of 40 ± 22 months was achieved. All 8 children improved by at least 1 NYHA functional class ($p < 0.05$), six-min walk distance raised dramatically to 502 ± 109 meters ($p < 0.05$) and medical therapy was simplified (monotherapy or no medication). SaO2 gradient remained in the same ranges.

Conclusion: Potts anastomosis for severe PAH in children is a life-saving procedure. Longer follow-up remains necessary to confirm these initial observations.

P-175

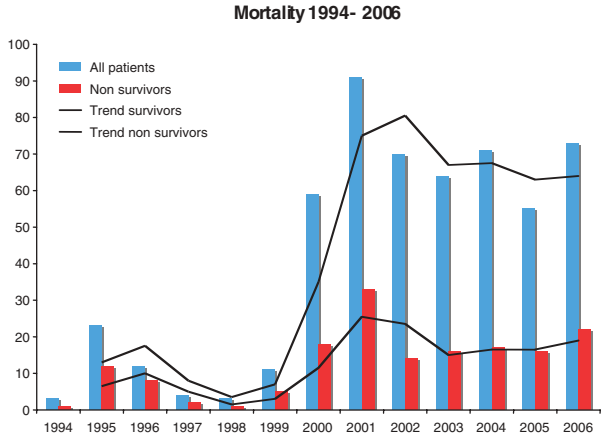
The Use of inhaled Nitric Oxide in the Critical Care Unit, Hospital for Sick Children, Toronto. A Retrospective Review 1994 – 2006

Hutter D., Sambamoorthi S., Humpl T.
Hospital for Sick Children, Department of Critical Care Medicine and Cardiology, Hospital for Sick Children, Toronto, Canada

Introduction: Since its discovery inhaled nitric oxide (iNO) has become standard treatment for a wide range of clinical conditions in pediatric critical care medicine. This retrospective review evaluates the use of iNO in a single institution over a 13 year period.

Methods: All patients who required endotracheal ventilation for ≥4 days between 01/1994 and 11/2006 in the Critical Care Unit, Hospital for Sick Children, Toronto, were identified and included into the study (n = 1701). Data were extracted from the paper chart or from the electronic charting system and consist of diagnosis, date of admission/discharge, age, gender, weight, height, duration of endotracheal intubation, mode of ventilation, ventilation score, maximum iNO delivered, duration of iNO treatment, catecholamine score, arterial blood gas and lactate. Pre-/during and post iNO treatment data were also investigated.

Results: A total of 539 patients (females = 251/male n = 288) were identified. Indications for iNO for both cardiac (n = 355) and medical surgical patients (n = 184) were: (1) congenital heart disease (CHD) with perceived pulmonary hypertension pre cardiac surgery (26%), (2) CHD s/p cardiac surgery (27%), (3) hypoxemic respiratory failure (18%), (4) diaphragmatic hernia (8%), (5) pulmonary hypertension (6%), (6) s/p HTx (3%), (7) cardiomopathy (2%), (8) s/p LTx (1%), other (9%). There was a significant increase in iNO use comparing the periods 1994 to 1999 (A) (n = 65) and 2000 to 2006 (B) (n = 474) and a decrease in mortality (A = 51% and B = 28%, Figure). Higher doses of iNO were administered during period A (29.9ppm ± 25) compared to period B (23.7ppm ± 21) while iNO treatment was longer in patients of period B (89.6h ± 18) compared to period A (71h ± 21). An identified predictor for survival was the patients condition prior to iNO therapy (Catecholamine Score A = 2.12 ± 0.34, B = 1.21 ± 0.21; pH A = 7.14 ± 0.18, B = 7.35 ± 0.14, lactate A = 5.39 ± 1.2, B = 2.95 ± 1.1).



Conclusion: This study represents the largest retrospective review of iNO use at a single centre. iNO was used more frequently in cardiac patients, with similar distribution before and after cardiac surgery. Lower dosing (used in the second timeframe) seems to be similarly effective compared to higher doses (used in the first timeframe). Clear indications for treatment should be established, especially with recently increased costs.

P-176

Cardiology services in Paediatric Intensive Care Unit in UK – Is it time to revise the provisions?

Thyaga Rajan S. (1), Al-Sabbagh A (2), Latifi. S(3), Kelsall W. (3)
Evelina Children's Hospital, London, UK (1); James Paget Hospital, Gt Yarmouth, UK (2); Addenbrookes Hospital, Cambridge, UK (3)

Aim: To audit the impact of cardiology services for critically ill children admitted to a primarily non-cardiac Paediatric Intensive Care Unit and to determine the availability of cardiology services for critically ill children in various non-cardiac Paediatric intensive care units in UK.

Methods: Data collected prospectively including all admissions to PICU at Addenbrooke's Hospital (8 bedded primarily non-cardiac PICU) over an 18 months period (Jan 2006–June 2007) were retrospectively analysed. The indications and outcomes of critically ill children referred for cardiology assessments were audited. A telephonic survey of all the Paediatric intensive care units within UK was conducted to determine the availability of cardiology services for children admitted to PICU.

Results: 716 admissions to the PICU over the 18 months study period were analysed. 40 children were referred for cardiology assessments. 20 patients had abnormal echocardiographic findings. Of these 20 patients, 7 were diagnosed with a critical cardiac condition that required urgent transfer to a cardiac intensive care unit for surgical intervention. A significant change in medical

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management following the echocardiogram occurred in a further 5 patients. 14 primarily non-cardiac PICUs were identified from the PICAnet database and contacted for the survey. ECHO expertise for emergencies were provided by paediatric cardiologist in 3 units, paediatrician with cardiology interest (when possible) in 5 units, by visiting paediatric cardiologist in 2 units, by trained paediatric intensivists in 1 unit, radiologist in 1 unit and children transferred out in 2 units. 5 units had 1 or more trained paediatric intensivists with ECHO expertise. None of the PICUs had tele-link facilities available or utilised.
Conclusions: The input from the paediatric cardiologist in a non-cardiac PICU cannot be underestimated. Majority of the primarily non-cardiac Paediatric intensive care units in UK are supported with cardiology expertise either by paediatric cardiologists or paediatricians with specific interest and available within 24 hours of referral. Very few paediatric intensivists are skilled with such expertise. Tele-link facilities appear under-utilised.

P-177
Gender differentuated minimally invasive surgical approaches for repair of simple congenital heart disease.
Padalino M.A., Veshti A, Vida V.L., Butticci R., Stellin G.
Pediatric and Congenital Cardiac Surgery Unit, University of Padova, Padova, Italy

Introduction: Interest in minimally invasive procedures has recently increased because it results in minor surgical trauma, reduced patient discomfort, shorter hospital stay, reduced costs and better aesthetic results. We analyzed our experience with gender differentiated surgical approach for repair of congenital heart diseases (CHD).
Methods: From January 1 st , 1998 to December 31st, 2004, a gender differentiated minimally invasive surgical approach was used for 308 patients affected by CHD (mean age: 8.9 yrs ± 10.3 yrs, median 5 yrs). Patients were divided in 2 groups, according to the chosen surgical approach: Mini-thoracotomy was performed in 147 patients (Group 1, M/F1/146), while a mini-sternotomy in 161 patients (Group2, M/F: 116/45). Both groups underwent bicaval venous and aortic cannulations through the chest incision. We routinously used assisted venous drainage during cardiopulmonary bypass and electrically induced ventricular fibrillation. Results were compared with a group of patients who underwent repair for ASD with full sternotomy (Group 3, 30 pts, mean age 16.1 yrs ± 20.3 yrs, median 5.5 yrs).
Results: There were no hospital deaths or major complications. No procedure required conversion to a full sternotomy in Group 1 or 2. Cross clamp, ventricular fibrillation and cardiopulmonary bypass times were equivalent in both groups. Hospital stay ranged between 4 to 17days in Group 1(G1) (mean 6 days ± 1.8 days), between 4 to 34 days in Group 2 (G2) (mean 6.7 days ± 2.9 days,). At a mean follow up of 5,6 yrs there were no residual cardiac defects, reoperations or interventional cardiac procedures. Among Group 1 and 2 patients, 96% were drug free, in NYHA Class I. In Group 1, there were no asymmetric breast development, or lactation problems at follow up.
Conclusions: Gender differerentiated aesthetic approach is a safe and effective procedure, with no additional costs or residual lesions. The majority of our patients are satisfied with aesthetic and functional results.

P-178
30 years experience with surgical correction of atrioventricular septal defects
Hoohenkerk G.J.F., Rijlaarsdam M., Schoof P.H., Koolbergen D.R., Bruggemans E.F., Hazekamp M.G.
Departments of Cardiothoracic Surgery and Pediatric Cardiology Leiden University, Medical Center, Leiden, The Netherlands

Objective: To identify risk factors for mortality and reoperation we analysed our experience with AVSD repair.
Methods: Between 1977 and 2006, 307 consecutive patients (males 152) underwent surgery for complete AVSD (n = 207; 67%), intermediate AVSD (n = 25; 8.2%) and, partial AVSD (n = 75; 24.8%). Patients with tetralogy of Fallot (n = 23), aortic coarctation (n = 8), and left ventricular outflow tract obstruction (n = 2) were included. All other associated cardiac anomalies were excluded. Median age was 6 months (range: 23 days -18 years), 199 (64%) patients were younger than 6 months. 157 Patients had Down's syndrome.
Preoperatively, 197 (64%) patients had moderate to severe AV valve regurgitation. In 31 (10%) patients, dysplasia of the AV valves was recorded from the surgeon's notes and operative reports. In 234 (76%) patients cleft closure was performed.
Results: There were 27 (9%) in-hospital deaths and three (1%) late deaths. Actuarial survival of the whole group after one year was 91%, after 10 years 90%, and after 20 and 30 years 87% In univariate analyses, risk factors for early mortality were age ($p < 0.001$), complete-AVSD ($p = 0.039$), AV valve regurgitation ($p = 0.047$) and associated cardiac anomalies ($p = 0.036$). In a multivariate analysis c-AVSD ($p = 0.038$) and AV valve regurgitation ($p = 0.004$) were risk factors for early mortality. Of the hospital survivors, 48 patients required reoperation. In univariate analyses risk factors for reoperation were AV valve regurgitation ($p = 0.001$), AV valve dysplasia ($p < 0.001$) and non cleft closure ($p = 0.045$). In a multivariate analysis, AV valve dysplasia ($p < 0.001$) and non cleft closure ($p = 0.045$) were risk factors for reoperation.
Follow- up was complete for all patients (median follow-up: 127 months; range, 13 to 360 months. At last follow-up all survivors were in good clinical condition and in NYHA functional class I (n = 258) or class II (n = 19).
Conclusion: AVSD repair can be accomplished with good longterm results. In our experience over a period of 30 years, risk factors for mortality, were complete-AVSD and AV valve regurgitation. Risk factors for reoperation were AV valve dysplasia and non cleft closure. Over the past two decades there was a decline in the age of AVSD repair.

P-179
Volume requirements after delayed sternal closure in infants after surgery for complex congenital heart defects
Lambrecht A., Weidenbach M., Hambsch J., Richter J., Kostelka M., Janousek J.
Heart Centre Leipzig, Leipzig, Germany

Introduction: Delayed sternal closure (DSC) is an accepted strategy for neonates and infants after complex congenital heart surgery associated with low cardiac output. Sternal closure changes intrathoracic volume and may have major impact on filling properties of the heart. Volume need may be altered due to temporary hemodynamic instability or recurring capillary leak. We prospectively investigated the fluid balance from 16 h before to 48 h after DSC.
Methods: Cristalloid and colloid balance was measured in 8 h intervals 16 h before until 48 h after DSC in 17 infants (median age 13 days) after complex congenital cardiac repair (TGA = 5; PA/VSD = 3; TAC = 3; HLHS = 2; IAA = 1, ALCAPA = 1, DILV = 1, ccTGA = 1). Indication for DSC was hemodynamic instability during sternal closure in the operating room or anticipated

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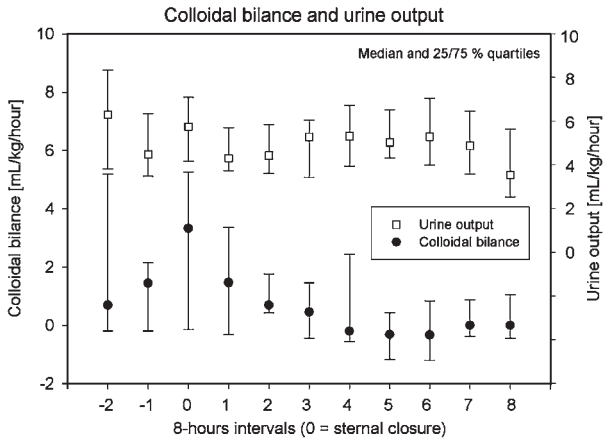

instability. Sternal closure was performed at a mean time of 1 day (range 1-5 days) after repair. Sternal closure was done when the patient was hemodynamically stable with either an equalised or negative fluid balance. Volume replacement was conducted based on clinical decision of the attending physician with colloidal fluids to achieve stable mean arterial pressure. DSC did not fail in any child, nor was there any major complications during DSC.

Results: Data are expressed as median and 25% / 75% quartile. For statistical analysis Friedman repeated measures analysis of variance on ranks was used with a p-value <0.05 as statistical significant. The maximal need for colloidal input was immediately after DSC with 4.6 [2.6/7.3] ml/kg/h ($p<0,05$ compared to the 8h intervall before) resulting in a positive colloidal balance of 3.3 [0.03/4.5] ml/kg/h. Colloidal input at the time of DSC was significantly greater than the input after 32 h ($p=0.05$). Colloidal balance was equalised 24 after DSC. Urine output did not changed significantly over the observed period (Figure).

Conclusion: To achieve stable hemodynamics in the immediate period after DSC infants need a significant increase in colloidal input. Colloidal need decreases significantly after 32 hours.

P-180

Novel prosthetic reinforcement technique for the prevention of autograft dilatation after the Ross procedure – early results

Kadner A. (1), Schwerzmann M. (2), Pavlovic M. (3), Pfammatter J.P. (3), Carrel T. (1)
Department of Cardiovascular Surgery (1); Department of Cardiology (2); Department of Pediatric Cardiology , University Hospital Bern (3)

Background: The Ross procedure remains a subject of debate, especially concerns exist regarding excessive dilatation of the neoaortic root leading to progressive regurgitation of the autograft valve. Here we present our early experience with an additional technical step, which consists of the inclusion of the pulmonary autograft in a prosthetic graft to prevent neoaortic root dilatation.

Material and methods: Between 2006 to 2007, 12 patients (mean age 23 ± 7 years; range: 15 –38 years) underwent the Ross procedure using the prosthetic reinforcement by a vascular graft with a prosthetic root configuration. Indications were aortic regurgitation ($n=2$), aortic stenosis ($n=5$), combined aortic stenosis and insufficiency ($n=5$). A bicuspid aortic valve was present in 9 patients. Balloon valvuloplasty had been performed in 7 patients. Follow up was performed by clinical and echocardiographic examinations.

Results: Hospital mortality and freedom from reoperation are 100%. Echocardiographic follow-up confirmed absence of aortic insufficiency in 11 patients after a mean of 12 months (range: 2 to 24 months). One patient still shows an asymmetric regurgitant jet, already observed at discharge echocardiography. As expected, no neoaortic root dilatation was observed during follow-up. All patients are in NYHA class I.

Conclusions: The present technique has the advantages of being technically simple, reproducible, and it does not require significant additional time. Furthermore, not only a singular structure, such as the aortic annulus or the sinotubular junction is supported, but the full autograft. The elasticity of the prosthesis should allow for the synchronous dilatation of the autograft root, while preventing the dilatation of autograft when exposed to systemic pressure in the long term.

P-181

Development of left ventricular systolic function of premature infant. By use of new echocardiographic tissue doppler imaging of strain and strain rate

Kikuchi Y.
Haga redcross hospital

Purpose. Left ventricular cardiac function of premature infant is gradually developed after birth. In this period, it is difficult to assess their cardiac function itself because of pulmonary hypertension. We assessed their cardiac function by use of new echocardiographic technique strain and strain rate.

Study design. We assessed 10 premature infant (33 to 35 weeks gestation). We performed echocardiographic assessment by use of Philips iE33 at within 7 days (early phase) and 21 to 28 days (late phase).

Parameters. Mean velocity, mean strain and mean strain rate (Tissue Doppler imaging; TDI). EF, Tei index (M mode).

Results. We could not find difference EF (58 ± 13 vs 62 ± 20; early phase vs late phase), Tei index (0.53 ± 0.07 vs 0.61 ± 0.34) between early and late phase. While we could find left ventricular dysfunction in early phase of premature infant in strain (36.9 ± 10.9 vs 48.9 ± 9.80 and strain rate (0.55 ± 0.065s vs 0.67 ± 0.089s).

Conclusion. We assessed cardiac function of premature infants by use of new TDI and M mode echocardiography. We could find left ventricular dysfunction in early phase of premature infant by use of new technique of strain and strain rate (TDI).

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