



# Cardiology in the Young

© 2010 Cambridge University Press

*44th Annual Meeting of the Association for European Paediatric Cardiology,  
AEPC with joint sessions with the Japanese Society of Pediatric Cardiology and  
Cardiac Surgery Innsbruck, Austria May 26–29, 2010*

## O1-1

### **Pre-operative assessment for Total Cavo-pulmonary Connection in Hypoplastic Left Heart Syndrome with MRI alone: Evaluation of outcomes**

Bellsham-Revell H. (1,2), Witter T. (2), Razavi R. (1,2), Bell A.J. (2)

King's College London, UK (1); Evelina Children's Hospital, Guy's and St Thomas' NHS Foundation Trust, London, UK (2)

**Introduction:** Standard pre-operative assessment for total cavo-pulmonary connection (TCPC) in hypoplastic left heart syndrome (HLHS) includes cardiac catheterisation. From 2003 we have used only echocardiography and Magnetic Resonance Imaging (MRI) with central venous pressure (CVP) measurement from an internal jugular vein (representing downstream pulmonary artery pressure) for preoperative assessment. We evaluated the postoperative outcomes in these patients.

**Methods:** Retrospective analysis of medical notes and MRI scans was performed. Information was collected on mortality, duration of ventilation, length of intensive care and inpatient stay, chest drainage and the need for further procedures.

**Results:** Between 2003 and 2009, 47 patients with HLHS were solely investigated with this method and underwent lateral tunnel TCPC with a 4 mm fenestration. 6 (12.8%) patients had tricuspid valve surgery at the time of TCPC. Results are described as median (range). CVP at the time of MRI was 12.4 mmHg (6–16). The age of patients at operation was 3.2 years (2.3–5.6) and weight was 14.4 kg (9.1–19.8). Survival was 98% with only one death in the immediate post-operative period secondary to an intractable arrhythmia. Patients were ventilated for 6.1 hours (1.75–23.3) and days spent in intensive care was 3 (2–11). Duration of chest drainage was 9 days (4–38) with 9 patients (19.6%) requiring chest drainage for over 2 weeks. Inpatient stay was 13.5 days (6–44). Spontaneous occlusion of the fenestration occurred in 2 patients, one required stenting of the fenestration on day 5, the other re-operation on day 4. This patient also required catheter occlusion of aorto-pulmonary collaterals. 3 (6.4%) patients required additional pericardial drainage. 5 (10.6%) were readmitted post discharge for drainage of recurrent effusions. 4 (8.5%) developed late complications requiring further intervention. These were protein losing enteropathy (1 patient), recurrent atrial restriction despite atrial septectomy at TCPC

(2 patients) and bradycardia requiring permanent pacing (1 patient). No patients required take down of the TCPC.

**Conclusions:** Results from TCPC in this group are favourable. The low reintervention rate suggests that echocardiography combined with MRI and CVP measurement is sufficient for pre-operative assessment for TCPC in HLHS and can obviate the need for cardiac catheterisation.

## O1-2

### **Total Anomalous Pulmonary Venous Drainage: Outcomes of Post-Operative Pulmonary Venous Obstruction from a Multinational Population-Based Study**

Seale A.N. (1), Uemura H. (1), Webber S.A. (2), Partridge J. (3), Roughton M. (1), Ho S.Y. (1), McCarthy K.P. (1), Jones S. (1), Shaughnessy L. (1), Sunnegardh J. (4), Hanseus K. (5), Berggren H. (4), Johansson S. (5), Rigby M.L. (1), Keeton B.R. (6), Daubeney P.E.F. (1). On behalf of the British Congenital Cardiac Association. Royal Brompton Hospital & National Heart and Lung Institute, Imperial College, London (1); Pittsburgh Children's Hospital, Pittsburgh (2); Harefield Hospital, London (3); Queen Silvia Children's Hospital, Gothenburg (4); Lund University Hospital, Lund (5); Cardiothoracic Unit Southampton General Hospital, Southampton (6)

**Introduction:** Pulmonary venous obstruction (PVO) is an important cause of late mortality in total anomalous pulmonary venous connection (TAPVC). We aimed to describe current practices for the management of postoperative PVO and the efficacy of the different interventional procedures in a population-based multicenter international study.

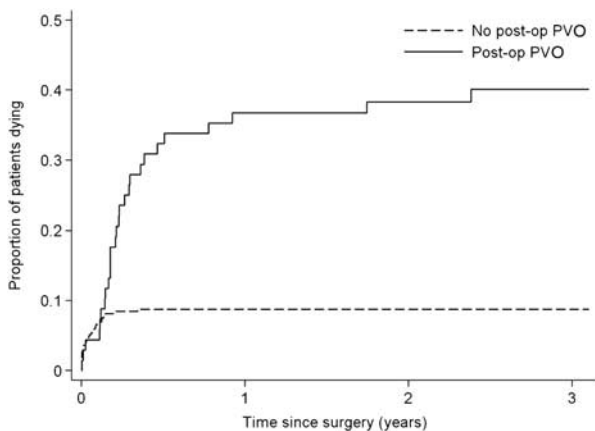
**Methods:** Retrospective international collaborative population-based study involving 19 paediatric cardiac centres in UK, Ireland and Sweden. Cases of TAPVC born between 1/1/1998 and 31/12/2004 were identified. Cases with functionally univentricular circulations or atrial isomerism were excluded. All available data and imaging were reviewed.

**Results:** Of 406 patients undergoing repair of TAPVC, 71 (17.5%) had postoperative PVO. Twenty-five had supracardiac, 25 had infracardiac, 11 mixed, 9 cardiac and 1 common pulmonary vein atresia. Diagnosis of postoperative PVO was made within 6 months of surgery in 83%. In 14, discrete stenosis

progressed to diffusely small pulmonary veins and sometimes atresia. Patients presenting after 6 months had less severe disease; all are alive at most recent follow-up.

Fifty-six (13.8%) underwent intervention: 44 surgical and 12 an initial catheter intervention. One half required re-intervention. There was no significant difference between interventional strategies. Three-year survival for patients with postoperative PVO was 60.4% (95% CI 47.7–70.9%), which is significantly worse (HR = 3.2  $p < 0.001$ , 95% CI 1.8–5.5) than those cases without postoperative PVO (Figure).

At diagnosis of postoperative PVO, diffusely small pulmonary veins (HR 7.8, 95% CI 2.3–2.8,  $p < 0.001$ ) and an increased number of lung segments affected (HR 1.8, 95% CI 1.1–3.1,  $p = 0.03$ ) were significant risk factors for death. Time between initial TAPVC repair and presentation with postoperative PVO was also important; patients presenting later with postoperative PVO did better (HR 0.964, 95% CI 0.95–0.98,  $p < 0.001$ ).



**Conclusions:** Postoperative PVO tends to present in the first 6 months after TAPVC repair and can be progressive. Close observation is required during this period, with further imaging being performed if there is any concern of postoperative PVO. Early intervention for PVO may be indicated before irreversible secondary changes occur.

### O1-3

#### Length of pleural effusions after Fontan completion is prolonged with extracardiac Fontan and Right ventricular morphology

Stumper O., Stickley J., Parkes S., Anderson B., Jones T., Barron D., Brawn WJ.

Birmingham Children's Hospital, Birmingham UK

**Introduction:** Prolonged pleural effusions are a common finding after Fontan completion. This study assesses whether the type of Fontan procedure or the underlying ventricular morphology has an impact on the length of pleural effusions [LPE].

**Methods:** Retrospective case review of 444 Fontan procedures carried out over a 25-year period. Analysis of pre and post op pulmonary artery pressures, time to chest drain removal, time to discharge (LOS), underlying ventricular morphology (RV = 211; LV = 233), and type of Fontan procedure (atriopulmonary [APC] = 130, lateral tunnel [LT] = 40, extracardiac [ECF] = 274). Postoperative management has evolved with a tendency towards earlier aggressive catheter intervention for residual haemodynamic lesions, the introduction of oral pulmonary

vasodilators and the cessation of pleurodesis, in principal favouring the later date Fontan population (=ECF).

**Results:** For the whole group LOS was significantly longer for ECF compared to APC/LT ( $p < 0.0001$ ). There was a significantly longer LOS for patients with RV versus LV morphology ( $p < 0.0001$ ). In the subgroup of ECF, LOS and LPE were significantly longer in patients with RV morphology ( $p < 0.05$ ). In the subgroups with RV morphology LOS and LPE were significantly longer in the ECF group compared to the APC/LT group ( $p < 0.05$ ).

**Conclusion:** Despite advances in postoperative management, the latest type of Fontan completion (ECF) is burdened with the longest hospital stay and length of pleural effusions – this is irrespective of underlying ventricular morphology. Studies should be undertaken to determine whether a different conduit material or weave might reduce length of drainage.

### O1-4

#### Congenital Heart Defects in Europe 2000–2005

Garne E., Loane M., Dolk H.

Hospital Lillebaelt, Kolding, Denmark

University of Ulster, Belfast, UK.

**Objectives:** To describe the prevalence of congenital heart defects (CHD) diagnosed prenatally or in the first year of life in Europe, and perinatal mortality.

**Methods:** Data from 29 population-based EUROCAT registries of congenital anomalies covering 3.3 million births, 2000–2005. Cases included livebirths, fetal deaths and terminations of pregnancy for fetal anomaly (TOPFA) at any GA. Non-chromosomal cases were classified by the ICD10 codes into two severity groups. VSD, ASD and Pulmonary Valve Stenosis made up the less severe group. 11% of cases were not classified, including PDA in term infants. Cases with no major non-cardiac malformations were classified as “isolated”. Data on surgery was only available from 2 registries.

**Results:** There were 26598 CHD cases. Total CHD prevalence was 8.0 per 1,000 births. 12% of cases had chromosomal anomalies. CHD with Down Syndrome occurred in 0.46 livebirths per 1,000 births of whom 0.40 survived the first week. There was a seven-fold variation in liveborn prevalence of CHD with Down Syndrome between countries. Liveborn babies with non-chromosomal severe CHD occurred in 1.69 per 1,000 births of which 1.58 survived the first week of life. Severe CHD was associated with a perinatal mortality of 0.16 (8%), and a TOPFA rate of 0.28 per 1,000 births (14%). Perinatal mortality and TOPFA rates varied significantly between countries.

Liveborn babies with non-chromosomal less severe CHD occurred in 4.11 per 1,000 births of which 4.08 survived the first week of life. Less severe CHD was associated with a perinatal mortality rate of 0.05 (1%) and a TOPFA rate of 0.05 per 1,000. 18% of all non-chromosomal CHD cases (48% of TOPFA) had associated major non-cardiac defects. The surgery rate for isolated non-chromosomal CHD was 0.91 per 1,000 births for severe CHD, and 0.48 per 1,000 for less severe CHD.

**Conclusions:** CHD remains the most frequent group of major congenital anomalies, demonstrating the lack of successful primary prevention. Although perinatal deaths and TOPFA are infrequent compared to number of livebirths with CHD, they are a major cause of perinatal mortality, with significant variation between countries relating to difference in TOPFA rate as well as to treatment.

**O1-5****Results of the PAN study: Congenital heart defects in newborns in Germany – prevalence and association with demographic, genetic and peripartur parameters.**

Lindinger A. (1), Schwedler G. (2), Hense H.-W. (3)

(1) Clinic of Pediatric Cardiology, University of the Saarland, Homburg,

(2) Competence Network for Congenital Heart Defects, Berlin,

(3) Institute of Epidemiology & Social Medicine, University Muenster.

**Objective:** The PAN study (Praevalenz angeborener Herzfehler bei Neugeborenen in Deutschland) is a nationwide prospective registration of newborns diagnosed with congenital heart defects (CHD) in Germany. From July 2006 to June 2009 about 18.000 patients have been entered into the data base by 283 participating institutions.

**Results:** The overall prevalence of CHD was calculated to be 1.08% in the first registration year (patients born July 06–June 07). The most common lesion was the ventricular septal defect (all types) (48.9%), followed by the atrial septal defect (17.0%), valvular pulmonary stenoses (6.1%), persistent arterial duct (4.3%) and aortic coarctation (3.6%). The most common cyanotic lesions were tetralogy of Fallot (2.5%) and D-transposition of the great arteries (2.2%). A single ventricle (all types) was identified in 2.8%, half of them being a hypoplastic left heart syndrome. Female gender was more common among mild CHD (57.3%), while in severe lesions there was a predominance of male infants (58.4%). Prematurity (<37 wks of gestation: 18.3 vs 9.1%), low birth weight (<2.500 g: 16.6 vs 6.8%) and multiple pregnancy (5.7 vs 3.3%) were about 2–3 times more common in infants with CHD than in the national average of live births. Independent of gender, maturity and genetic disorders, multivariate regression analysis revealed a coincidence of low birth weight with specific cardiac lesions, such as TAC, PA/VSD, ToF and AS. Extracardiac anomalies were registered in 2.2% and genetic disorders in 5.3% of the patients. The cAVSD was associated with trisomy 21 in 65% and with extracardiac anomalies in 2.3%. CATCH 22 was associated with VSD (22%), IAA (20%), ToF (18%), TAC and PA/VSD (11% each), und DORV (7%). Diagnosis assessment was depending on the severity category of the cardiac lesion: severe defects have been detected earlier than mild cardiac defects, as well pre- and postnatal.

**Conclusion:** The nationwide PAN study revealed a CHD prevalence of newborns in Germany of 1.08%. Hemodynamically mild cardiovascular lesions accounted for about two third of all defects. CHD was associated with prematurity, low birth weight and multiple births.

*The study is supported by the German Federal Ministry of Education and Research, FKZ01GI0601.*

**O1-6****SWEDCON – The Swedish Registry of Congenital Heart Disease**

Björkhem G., Thilén U.

University Hospital, Lund, Sweden.

**Objectives:** To create a nationwide registry for children and adults with congenital heart disease in Sweden, also including surgery and catheter interventions. Data about diagnoses, operations, interventions, hospital care, out-patient visits, examinations and quality of life should be entered. Such a registry would provide information about the natural course for different cardiac malformations and results of treatment. It would make it possible to follow the patients from birth to adult age and also to compare the quality of care in different parts of Sweden.

**Methods:** The new registry SWEDCON was based on the previous GUCH registry. The registry is web-based and data

from the GUCH registry and from local registries at two regional centers for pediatric cardiology were imported. Special parts of the registry were created for registering cardiac surgery with specific details about the operations and catheter interventions also including any complications to the procedures. Quality of life is registered for children using DISABKIDS and for adults using EQ-5D. Every hospital has access to its own data and can compare data to the whole country's data but cannot see data from any other specific hospital.

**Results:** The registry was opened to use in February 2009. At the start SWEDCON included 25 588 imported patients, 6755 from the previous GUCH registry and the rest from the regional pediatric registries. 1500 patients were present both in the pediatric and the GUCH parts of the registry. After the first 9 months the registry is now routinely used by the 7 major GUCH centers and the 4 major pediatric cardiac centers including the 2 operating centers in the country. The registry is also used by 19 regional hospitals with pediatric cardiology units and this number is increasing rapidly. A total of 4157 new patients and 5631 out-patient visits have been registered in SWEDCON since the start. **Conclusions:** SWEDCON now covers all congenital cardiac surgery and all specialized GUCH and pediatric cardiology units in Sweden. It also covers most local pediatric cardiology clinics. Major success-factors have been the joint creation of the registry and the easy availability of data for each separate hospital.

**O1-7****What is the optimal dose of clopidogrel in children with heart disease?**

Naqvi N., Edwards S, Damda F, Maiya S, Jiggins M, Chubb H, Forrester S, Slavik Z.

Royal Brompton and Harefield Hospitals Foundation NHS Trust.

**Objective:** There is paucity of data on clopidogrel dosing in children. We compared efficacy and safety of high versus low dose clopidogrel administered as an alternative to warfarin, due to needle phobia, in a paediatric cardiac population.

**Method:** Single tertiary paediatric cardiac centre hospital record review and telephone questionnaire of parents of 64 children receiving clopidogrel treatment (Jan 2004–Dec 2009).

**Results:** The median age at commencement of treatment was 4.7 years (range 0.1–17.2 years). Sixty children underwent cardiac surgery (including 31 patients after TCPC, 11 patients post systemic-pulmonary shunt or RV-PA conduit, 11 patients with additional stent or intracardiac device, and 7 miscellaneous complex cardiac surgeries), 3 had dilated cardiomyopathy and one had intracardiac thrombus. Given the relatively high incidence of side effects (56.5%) on a dose of  $\geq 0.5$  mg/kg/day between 2004–2007 (46 children, mean dose of 1.4 mg/kg/day [range 0.5–4.4 mg/kg/day]), we changed dosing regime in Jan 2008 to  $< 0.5$  mg/kg/day (21 children, mean dose of 0.2 mg/kg/day [range 0.15–0.4 mg/kg/day]). The intention was to convert those still on high dose to low dose and to start all new patients on lower dose. Age range and indication for treatment were similar in both groups. Side effects were experienced by 56.5% of the high dose group patients, with spontaneous excessive bruising (31%) and epistaxis (9%), haemoptysis (1 child), excessive bleeding (2 children, undiagnosed underlying clotting abnormality, 1 required transfusion post minor trauma). One set of parents were mistakenly accused of child abuse for excessive bruising. Clopidogrel was discontinued due to unacceptable side effects in 11%.

Side effects occurred in 20% of those receiving clopidogrel at  $< 0.5$  mg/kg/day, haemoptysis (1 child), epistaxis (1 child) and

distressing bleeding occurred in a child with eczema. Only 1 child stopped clopidogrel due to side effects. Importantly 1 child receiving clopidogrel at 0.3 mg/kg suffered an embolic stroke whilst there were no thrombotic complications in the high dose group.

**Conclusions:** Clopidogrel prevents thrombosis at doses >0.5 mg/kg but has a high incidence of unpleasant side effects. At lower dose the side effects are less common but this benefit may be outweighed by increased risk of thrombosis including stroke. Optimal dose remains elusive.

## O2-1

### **Assessment of ventricular function and dyssynchrony in anatomic subtypes of hypoplastic left heart syndrome after Fontan surgery using 2D speckle tracking**

Petko C., Hoffmann U., Möller P., Kramer H.-H., Uebing A.

Klinik für Kinderkardiologie, Universitätsklinikum Schleswig-Holstein, Kiel, Germany.

**Background:** There is evidence that the presence of a significant left ventricle (LV) in patients with hypoplastic left heart syndrome (HLHS) negatively affects systemic right ventricular (RV) function and outcome. The purpose of our study was to investigate differences in global and regional RV function as well as intraventricular dyssynchrony between HLHS patients after Fontan surgery who did or did not have a significant LV.

**Methods:** A total of 29 HLHS patients after Fontan surgery (mean age  $7.6 \pm 2.1$  years) were studied by echocardiography. The patient cohort was divided into two groups. Group 1 consisted of 15 patients with mitral and aortic atresia and a negligible LV, group 2 of 14 patients with a significant LV remnant. In addition to global strain (S) and strain rate (SR), we calculated regional peak systolic longitudinal S, SR and velocity in the 6 RV segments (basal septal, mid septal, apical septal, apical lateral, mid lateral and basal lateral) from the apical 4-chamber view using 2D speckle tracking. To assess dyssynchrony, we calculated the 3 standard deviations of the time interval from the onset of the QRS interval to peak systolic event (S, SR and velocity) of the 6 RV segments.

**Results:** Global S ( $-19.5 \pm 2.8$  vs.  $-17.4 \pm 3.9\%$ ) and global SR ( $-1.0 \pm 0.2$  vs.  $-0.9 \pm 0.3$  s<sup>-1</sup>) were not different between groups. Group 2 had significantly lower peak S in the basal septal ( $p = 0.003$ ) and mid septal ( $p = 0.009$ ) segments and lower peak SR in the mid septal segment ( $p = 0.047$ ). Peak velocities were similar in all segments among groups. The standard deviation of time to peak S was significantly higher in group 2 than group 1 ( $53 \pm 64$  vs.  $13 \pm 7$  ms,  $p = 0.02$ ) indicating greater dyssynchrony in patients with a significant LV.

**Conclusions:** The presence of a significant LV in HLHS after Fontan surgery did not impact global RV function in our cohort. It was associated with reduced strain and strain rate of the septal myocardium adjacent to the LV. In addition, intraventricular dyssynchrony was more pronounced. Whether this has any impact on RV function in the long term or influences outcome warrants longer follow-up.

## O2-2

### **Present results after correction of Tetralogy of Fallot in Germany – Prospective analysis of 407 patients for realistic expectations in the results of exercise testing and cardiac MRI**

Sarikouch S. (1), Dubowy K.O. (2), Burkhardt B. (3), Mir T.S. (4), Hager A. (5), Peters B. (6), Kuehne T. (7), Beerbaum P. (8) for the German Competence Network Congenital Heart Defects-Investigators

Department of Heart-, Thoracic-, Transplantation-and Vascular Surgery, Hannover Medical School (1); Department for Congenital Heart Disease, Heart and Diabetes Centre Bad Oeynhausen (2); Department for Congenital Heart Disease; University of Freiburg (3); Pediatric Cardiology, University Heart Center Hamburg (4); Department for Pediatric Cardiology, Deutsches Herzzentrum München, TU München (5); Institute for Biometry and Medical Informatics, University of Magdeburg (6); Unit of Cardiovascular Imaging – Congenital Heart Diseases, German Heart Centre Berlin (7); Division of Imaging Sciences, King's College, Guy's & St Thomas' Hospital, London, United Kingdom (8)

**Introduction:** Knowledge of common present postoperative sequelae is essential for realistic expectations in the results of imaging modalities, exercise testing or re-interventions. It is for this reason the “Competence Network for Congenital Heart Defects” in Germany initiated a nationwide study in Tetralogy of Fallot (TOF) to determine the prevalence and extension of postoperative restrictions.

**Methods:** Prospective, longitudinal study in repaired TOF, age >8 yrs including physical examination, ECG, Holter, echocardiography (ECHO), tissue-doppler-imaging (TDI), magnetic resonance imaging (MRI), cardiopulmonary exercise test and age adjusted quality of life questionnaire. Repetition after 12 months. Standardisation for all diagnostic modalities, central core laboratory evaluation of ECHO, TDI, MRI and exercise tests. Results were related to age matched healthy reference populations as investigated within the Network using identical methodology.

**Results:** 407 patients were included in 14 sites, mean age  $17.87 \pm 8.31$  yrs, weight at corrective surgery  $11.55 \pm 6.57$  kg. Transvalvular patch in 50.9%. 67.2% were in NYHA class I, 30.8% NYHA II and 2% in NYHA III. 90.9% were in sinus rhythm, heart rate  $73.3 \pm 14.5$  bpm, max. QRS  $145.6 \pm 23.6$  msec. Right ventricular enddiastolic volume  $120 \pm 33$  ml/m<sup>2</sup>, right ventricular ejection fraction  $50 \pm 9\%$ , mean pulmonary regurgitation  $26 \pm 18\%$ . Correlations between MRI and exercise tolerance revealed highly significant correlations, e.g. left ventricular ejection fraction and peak oxygen uptake. Whereas the quality of life of pediatric patients did not differ in any dimension from the quality of life of healthy comparison groups, there were highly significant differences for cardiac MRI and exercise testing. E.g. the 50% percentile of patients with corrected TOF (using the LMS-method by Cole) for exercise testing parameters is in the range of the 3% percentile of healthy controls. For right ventricular volume analysed by MRI, the 50% TOF-percentile is in the range of the 97% percentile of healthy women or men.

**Conclusions:** A large data base for a contemporary cohort after correction of Tetralogy of Fallot was established for robust statistical analysis and further longitudinal studies. The prevalence of common present postoperative sequelae for TOF in Germany was determined, the results were used to compute heart defect (TOF) specific reference values for exercise testing and MRI.

## O2-3

### **Pulmonary vascular resistance and systolic-diastolic ventricular function in Fontan patients at rest and during dobutamine stress**

Kuehne T. (1), Schmitt B. (1), Steendijk P. (2), Ovroutski S. (1), Lunze K. (1), Rahmzadeh P. (1), Maarouf N. (1), Ewert P. (1), Berger F. (1)

German Heart Institute Berlin, Berlin, Germany (1); Leids Universitair Medisch Centrum, Leiden, The Netherlands (2).

**Background:** The role, interplay, and relative importance of the multifactorial hemodynamic and myocardial mechanisms causing dysfunction of the Fontan circulation remain incompletely understood.

**Methods:** Using a MRI catheterization technique, we performed a differential analysis of pulmonary vascular resistance (PVR) and aorto-pulmonary collateral blood flow in conjunction with global ventricular pump function, myocontractility (ESPVR), and diastolic compliance (EDPVR) in 10 Fontan patients at rest and during dobutamine stress. Pulmonary and ventricular pressures were measured invasively and synchronized with velocity encoded (VEC) MRI derived pulmonary and aortic blood flows and cine MRI derived ventricular volumes. PVR, ESPVR and EDPVR were determined from these pressure-volume data. Aorto-pulmonary collateral flow was calculated as the difference between aortic and pulmonary flow.

**Results:** Compared to rest, dobutamine caused a small increase in mean pulmonary pressures ( $p < 0.05$ ). Collateral flow was significantly augmented ( $p < 0.001$ ) and contributed importantly to an increase in pulmonary flow ( $p < 0.01$ ). PVR decreased significantly ( $p < 0.01$ ). Dobutamine failed to increase significantly stroke volumes despite slightly enhanced contractility (ESPVR). Active early relaxation (Tau,  $dP/dt_{Min}$ ) was inconspicuous but the EDPVR shifted upwards indicating reduced compliance.

**Conclusions:** In Fontan patients, aorto-pulmonary collateral flow contributes substantially to enhanced pulmonary flow during stress. Pulmonary vascular response to augmented cardiac output was adequate but decreased diastolic compliance was identified as an important component of ventricular dysfunction.

## O2-4

### Experience with dobutamine stress echocardiography in the pediatric population

Joly H. (1), Ducreux C. (1), Henaine R. (2), Sassolas F. (1), Gouton M. (1), Ninet J. (2), Bozio A. (1), Di Filippo S. (1)

Pediatric Cardiology, Cardiovascular hospital Louis Pradel, Lyon, France (1); Cardiac Surgery, Cardiovascular hospital Louis Pradel, Lyon, France (2)

The ability of dobutamine stress echocardiography (DSE) to detect myocardial ischemia and to assess viability, has been demonstrated in adults. To date, the value of this technique is still a matter of debate in the pediatric population. The objective of this study is to report our experience with DSE in children, to assess safety and reliability of this technique.

**Material and methods:** 137 DSE studies were performed, 84 (group1: 47%) in heart transplant patients and 53 (group2: 33%) after arterial switch operation.

**Results:** Age at DSE was  $12.5 \pm 5$  years (median 11.7), higher in group1 (14.1 vs 9.9 years,  $p < 0.0001$ ). Time from surgery was  $7.7 \pm 4$  years (median 7.1): mean 6.7 years in group 1 and 9.4 years in group 2 ( $p = 0.0001$ ). Maximum dose of dobutamine was reached in 93.4% of cases, with no difference between groups. No significant side effect occurred. Maximal heart rate was  $153 \pm 19$  bpm, i.e.  $74 \pm 9$  of theoretical maximal. Increase in heart rate was 71% in group1 and 81% in group2 ( $p = 0.02$ ). Maximal blood pressure was  $151 \pm 20$  mmHg. Increase in blood pressure was 39% in group1 and 52% in group2 ( $p = 0.06$ ). Echocardiographic scores were abnormal in 17 cases (12.4%), 6 (7.1%) in group1 and 11 (20.7%) in group2 ( $p = 0.01$ ). These anomalies were significant at peak and post-infusion stages. Compared to coronary angiography or coroscanner, estimated sensitivity and specificity of DSE technique were respectively 88.8% and 94.5%.

**Conclusion:** Dobutamine stress echocardiography in children is safe and well tolerated. This technique can help to detect myocardial ischemia in cases with significant coronary stenosis and can assess hemodynamic profile of patients during exercise.

## O2-5

### Dobutamine Stress MR in Tetralogy of Fallot with severe pulmonary regurgitation: Is RV-ESV response important?

Parish V., Valverde I. (1), Head C. (2), Greil G. (1), Rosenthal E. (2), Quershi S. (2), Schaeffter T. (1), Sarikouch S. (3), Razavi R. (1), Beerbaum P. (1)

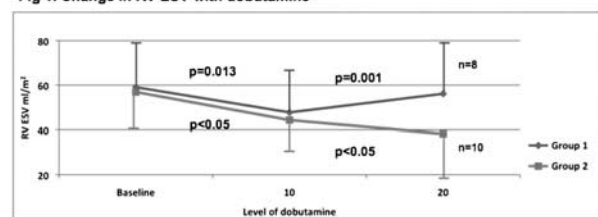
Imaging Sciences, King's College, London, UK (1); Guy's and St Thomas' NHS Trust, London, UK (2); Hannover Medical School, Department of Cardiac Surgery, Hannover, Germany (3)

**Introduction:** In repaired Tetralogy of Fallot (r-TOF), chronic severe pulmonary regurgitation (PR) is associated with progressive right ventricular failure, exercise intolerance and life-threatening arrhythmia. Dobutamine stress MR (DS-MR) may improve detection of early RV dysfunction and thus help with timing of pulmonary valve replacement. Right ventricular endsystolic volume (RV-ESV) response at DS-MR is thought to be potentially discriminative but this could not yet be demonstrated at low-dose DS-MR (Dobutamine 7.5 mcg/kg/min) as commonly used. We hypothesized that higher doses of Dobutamine may provoke different RV-ESV responses in r-TOF with PR.

**Methods:** We prospectively recruited 28 patients with r-TOF, RV dilatation and chronic PR (age  $32 \pm 15$ , pulmonary RF  $40 \pm 7\%$ ), but without arrhythmia, impaired LV function, or severe pulmonary stenosis. Ventricular volumes (cine-MRI) and pulmonary/aortic flows (phase-contrast) were obtained at baseline and during Dobutamine infusion of 10 and 20 mcg/kg/min. DS-MR was stopped at systolic blood pressure  $>190$  mmHg, heart rate  $>75\%$  predicted ( $220 - \text{age}$ ) or significant side effects.

**Results:** Of 28 patients, 24 completed Dobutamine at 10 mcg/kg/min (2 had claustrophobia and 2 had frequent ventricular ectopics). Twenty patients went on to successfully complete Dobutamine at 20 mcg/kg/min. Two data sets at Dobutamine-20 had to be excluded due to incomplete volumetry.

Fig 1. Change in RV-ESV with dobutamine



Group 1: No response or increase in RV-ESV with increase from 10 to 20 dobutamine  
Group 2: Reduction in RV-ESV with increase in dobutamine concentration (10-20)

The heart rate (cardiac index) increased from  $66 \pm 6$  bpm ( $2.9 \pm 0.41$  min/m<sup>2</sup>) at baseline to  $91 \pm 18$  bpm ( $3.9 \pm 0.91$  min/m<sup>2</sup>) at Dobutamine-10 and  $117 \pm 14$  bpm ( $4.5 \pm 1.1$  min/m<sup>2</sup>) at Dobutamine-20 ( $p < 0.01$ ). Observer variance remained low at DS-MR. Significant reduction in LV enddiastolic (EDV) and endsystolic volumes (ESV) is seen at Dobutamine-10 and 20 ( $p < 0.01$ ). RV showed less marked reduction in volumes at Dobutamine-10. Further increase from Dobutamine-10 to Dobutamine-20 identified a subgroup in which RV-ESV increased or remained unchanged compared to Dobutamine-10 values ( $n = 8$ ,  $p < 0.01$ ), whilst in an otherwise identical subgroup the RV-ESV further decreased significantly ( $n = 10$ ,  $p < 0.05$ ; see Figure 1).

**Conclusions:** In r-TOF with chronic severe PR, DS-MR appears safe up to 20 mcg/kg/min Dobutamine. At this higher dose we observed a sub-group with no further reduction or even increase of RV-ESV, compared to Dobutamine-10. The incremental response in RV-ESV at higher-dose DS-MR may reflect early systolic RV dysfunction if confirmed in outcome studies.

### O3-1

#### Rotation of the truncus arteriosus – established surgical procedure

Ovroutski S., Miera O., Peters B., Ewert P., Berger F., Hetzer R., Hübner M.

German Heart Institute Berlin

**Background:** Rotation of the truncus arteriosus enables the native pulmonary valve to be preserved during correction of transposition of the great arteries (TGA) with ventricular septal defect (VSD) and left ventricular outflow tract (LVOT) obstruction.

**Methods:** In a retrospective analysis we studied seven patients who have undergone surgery with rotation of the truncus arteriosus since 2006, with age at operation of 5–40 months and weight of 5–20 kg. In all patients the base of the heart with the great vessels was resected, rotated by 180° and then reimplanted, so that the aortic valve was positioned above the LVOT. The LVOT obstruction was relieved by resection of the muscular conus, the VSD was closed and the coronary arteries were reimplanted. In reconstructing the right ventricular outflow tract either the native pulmonary valve was preserved (n = 3) or a monocusp was used (n = 4). The perfusion time was 217–298 min and the aortic clamp time 139–239 min.

**Results:** All patients were extubated within 48 h and discharged home within 10 days. So far no reoperation has been necessary. Left and right ventricular function remained normal (ejection fraction >60%). There was no aortic valve incompetence or residual LVOT obstruction. One patient had mild pulmonary valve incompetence. On patient developed AV block grade III following extensive conus resection and received pacemaker implantation. All other patients showed sinus rhythm.

**Conclusion:** Rotation of the truncus arteriosus is a safe surgical method of anatomical correction of TGA with VSD and LVOT obstruction that can be performed with low morbidity rates. In all children studied, the growth potential of the pulmonary valve was preserved.

### O3-2

#### Bicuspidation of cryopreserved pulmonary homografts for reconstruction of the right ventricular outflow tract

Rega F. (1), Jacobs S. (1), Allaeyns M. (1), Jashari R. (2), Daenen W. (1), Gorenflo M. (3), Heying R. (3), Eyskens B. (3), Gewillig M. (3), Meyns B. (1)

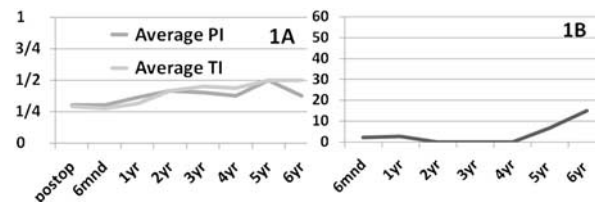
(1) Department of Cardiac Surgery, University Hospital Gasthuisberg, Herestraat 49, 3000 Leuven, Belgium; (2) European Homograft Bank, Brussels, Belgium; (3) Department of Pediatric Cardiology, University Hospital Gasthuisberg, Leuven, Belgium.

**Objectives:** The reconstruction of the right ventricular outflow tract is an essential part of the treatment in congenital heart disease. Cryopreserved homografts became the conduit of choice for this procedure. The supply of small sized pulmonary homografts, however, is limited. We reviewed our experience with bicuspidized cryopreserved pulmonary homografts implanted in the right ventricular outflow tract.

**Methods:** From 1999 through 2009, we reconstructed the pulmonary valve with 12 bicuspidized homografts in 12 patients

(median age 1.6 years; [range 0.04–10.6 yrs]; median weight 8.8 Kg [2.6–25 Kg]. Indications were Tetralogy of Fallot (10), Truncus (1) and Rastelli repair in TGA (1). Median follow-up was 2.2 years [0–6.7 yrs].

**Results:** Median homograft diameter prior to size reduction was 26 mm [19–29 mm] and after bicuspidation 17 mm [12.7–19.3 mm]. One patient died on day 8 after implantation of the homograft due to intractable lung bleeding. No reintervention was needed for graft dysfunction. Early postoperative haemodynamics showed a mean pulmonary and tricuspid valve insufficiency of 1/4 which slightly increased up to <2/4 at 6 years. Pulmonary valve stenosis was absent early postoperative and increased to 15 mmHg at 6 years. Average pulmonary valve and tricuspid valve insufficiency are depicted in Figure 1A. Average pulmonary valve stenosis is depicted in Figure 1B.



**Conclusions:** Bicuspidation of the pulmonary homograft is feasible and follow up to 6 years reflects good outcome as well as excellent haemodynamics. Bicuspidation can be considered in absence of pulmonary homografts of appropriate size for reconstruction of the right ventricular outflow tract.

### O3-3

#### Results of a tissue engineered pulmonary valve in humans

Rickers C. (1), Entenmann A. (1), Fischer G. (1), Scheewe J. (1), Scheid M. (1), Braesen J. (2), Vöges I. (1), Hart C. (1), Kramer H.H. (1)

Dept. Ped. Card., University Hospital Kiel, Germany (1); Dept. Pathology, University Hospital Kiel, Germany (2)

**Introduction:** Tissue-engineered (TEng) valves have recently been developed as an alternative to pulmonary valve replacement with homograft conduits to improve device life-time and function.

**Purpose:** To evaluate the morphological and functional properties of a decellularized porcine xenograft valve conduit (Matrix-P™) and its successor product with additional equine pericardium sheathing (Matrix-P-Plus™; AutoTissue, Germany).

**Methods:** TEng valves had been implanted into the right ventricular outflow tract of 27 pts (age: 15 ± 8.5 yrs; Matrix-P™ n = 10; Matrix-P-Plus™ n = 17) due to pulmonary stenosis or insufficiency. Echocardiography was suspicious of accelerated blood flow in the main pulmonary artery in 13 of those within a median of 5 months (range 2–7) post operation. A comprehensive MRI study was utilized for the assessment of the severity of the stenosis as well as soft tissue characterisation using gradient echo cine, T1 and T2 weighted turbo spin echo and late enhancement sequences.

**Results:** In 10 pts (37%) CMR detected a significant conduit stenosis (Vmax 3.8 ± 1.5 m/s) with mild or moderate insufficiency (RGF 23.4 ± 4.8%) post-surgery. In 3 pts the conduit was considered to be normal. In all pts with a stenosis, T1-weighted images showed significant paravalvular Gadolinium enhancement and wall thickening of the conduit. In 8 pts the “Matrix” valve conduit was replaced by a homograft. Explants were evaluated through histological analysis that showed granulomatous inflammation and increased fibrous tissue content correlating with CMR findings.

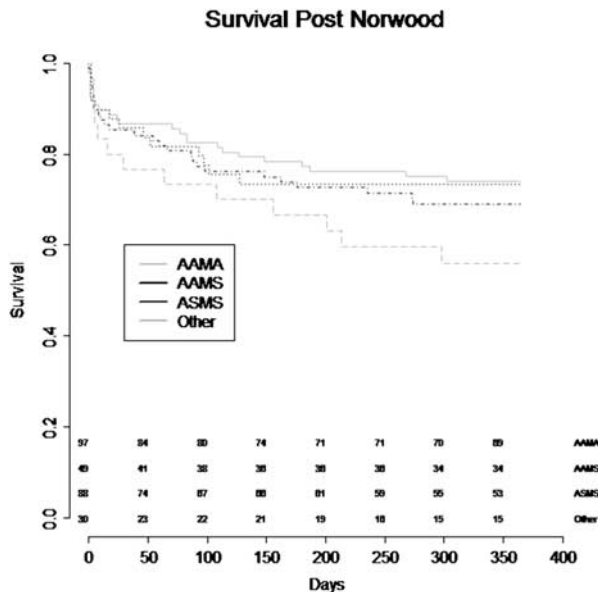
**Conclusions:** As a consequence of the devastating short term results, use of these decellularized pulmonary grafts was terminated at our center after initial use in 27 patients.

### O3-4

#### Early survival in 264 patients following the Sano-modified Norwood procedure: influence of morphologic subtype

Murtuza B. (1), Wall D. (1), Stickley J. (1), Jones T.J. (1), Barron D.J. (1), Brawn W.J. (1)

Department of Paediatric Cardiac Surgery Birmingham Children's Hospital (1)



**Objectives:** One of the most significant technical developments in the Norwood procedure is the Sano modification using a right ventricle-pulmonary artery (RV-PA) conduit rather than a systemic-PA shunt. It has been suggested that the RV-PA conduit might confer a benefit particularly for patients with the aortic stenosis-mitral atresia (AS-MA) morphologic subtype of hypoplastic left heart syndrome (HLHS), though this question remains unanswered. We report here early survival for our large, single-centre experience using the Sano-modified Norwood procedure and the influence of morphologic subtype on survival.

**Methods:** A retrospective analysis was performed of 264 patients who had undergone the Norwood procedure using a RV-PA conduit between May 2002 and December 2009. HLHS morphologic subtypes were classified as: aortic stenosis-mitral stenosis, AS-MS; aortic atresia-mitral stenosis, AA-MS; and AA-MA. There were no patients of the AS-MA subtype. The distinction between valvar atresia and stenosis was made by assessment of flow using colour flow Doppler imaging on 2-dimensional images. Non-classical morphologic variants ("other") included patients such as those with unbalanced complete atrioventricular septal defects. We determined the actuarial survival and the influence of left heart morphology, ascending aorta size and patient weight.

**Results:** In agreement with previous studies, patient weight  $\leq 2.5$  kg was an important predictor of survival, with this group exhibiting lower actuarial survival to 1 year compared with those  $> 2.5$  kg. The smallest ascending aorta size was associated with the AA-MA subtype, though ascending aorta size  $\leq 2.0$  mm was not in itself associated with a lower survival. Further, the AA-MS morphologic subtype was not predictive of a lower 1 year

actuarial survival. In contrast, however, the "other" category of morphology was associated with a significantly lower actuarial survival at 1 year (Figure 1).

**Conclusions:** Use of the RV-PA conduit appears to protect patients from the effects of both ascending aorta size  $\leq 2.0$  mm and AA-MS morphology in terms of actuarial survival to 1 year. These observations may in turn relate to less diastolic runoff and improved coronary perfusion using the RV-PA conduit in these patients.

### O3-5

#### Small Right Ventricle to Pulmonary Artery Conduits in Infancy

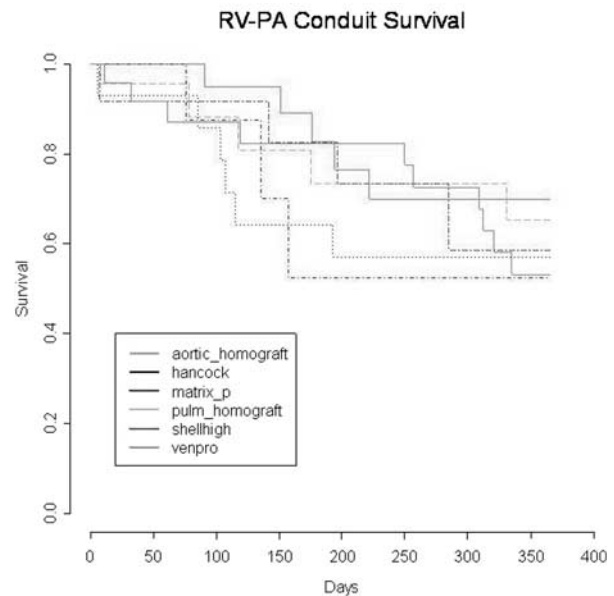
Anderson B.W., Stumper O., Desai T., Stickley J., Jones T., Brawn W., Barron D.

Birmingham Children's Hospital, Birmingham, United Kingdom

**Introduction:** There is debate over the best choice of RV-PA conduit to effect biventricular repair of complex CHD in infancy.

**Aim:** Compare the effective lifespan of the different conduit types in terms of freedom from conduit related catheter or surgical re-intervention.

**Methods:** Retrospective case review of 117 infants over a 14 year period that underwent complete bi-ventricular repair of complex CHD [Truncus (44), IAA/VSD (20) and PAVSD (43)] using an RV-PA conduit [aortic homograft (24), pulmonary homograft (24), Tissuemed (10), Shelhigh (4), Hancock (17), Contegra/Venpro (28), Matrix P (9)]. Conduit survival was defined as the length of time of freedom from conduit related catheter or surgical re-intervention (FfCSRI), death or last point of follow-up. Data was censored for early post-operative deaths, with the analysis conducted for 30-365 days of survival. Indications for intervention were RV pressures  $> 50\%$  systemic and/or impaired RV function.



**Results:** Median age at repair was 24 days (1-344). There were 16 early deaths, not related to conduit obstruction or type. In the survivors, overall conduit lifespan median was 288 days (32-3367). At one year post implantation there was no significant difference in FfCSRI for the different types of conduits, nor was there a significant difference in those implanted at less than 90 days of age.

**Conclusion:** At present, all small RV-PA conduits available for use in the complete repair of complex CHD in infancy are burdened by high stenosis rates, requiring early catheter or surgical re-intervention. The search for the ideal small caliber RV-PA conduit continues.

#### O4-1

##### **Vascular remodeling in adults after coarctation repair – impact of descending aorta stenosis and age at surgery.**

Trojnarcka O. (1), Szczepaniak-Chichel L. (2), Mizia-Stec K. (3), Gabriel M. (4), Bartczak A. (1), Grajek S. (1), Tykarski A. (2), Gasior Z. (3)

Ist Department of Cardiology of University of Medical Sciences, Poznan, Poland (1); Department of Hypertensiology, Angiology and Internal Medicine, University of Medical Sciences, Poznan, Poland (2); Department of Cardiology, Medical University of Silesia, Katowice, Poland (3); Department of General and Vascular Surgery (4)

**Introduction:** Patients after successful repair of coarctation of the aorta (CoA) are at risk of hypertension at rest and associated end-organ damage. The aim of the study was to assess arterial stiffness and function in adults after coarctation repair in relation to descending aorta residual coarctation and patient age at operation.

**Patients and methods:** 85 patients after CoA repair (53 males) aged  $34,6 \pm 10,3$  years; median age at operation  $0,9 \pm 8,2$  years. The control group – 30 individuals (18 males) at mean age  $33,6 \pm 8,2$  years. Flow mediated dilatation (FMD%), nitroglycerine-mediated vasodilatation (NMD), intima-media thickness (IMT) and pulse wave velocity (PWV) were measured by using high-resolution ultrasound in the peripheral arteries. The following central vascular parameters were evaluated: augmentation (AP) and augmentation index (AI).

**Results:** 47 CoAo repaired patients were normotensive, and compared to control, they presented higher values of central parameters AP ( $7,3 \pm 4,6$  vs  $4,4 \pm 3,6$  mmHg;  $p = 0,002$ ) and AI ( $18,6 \pm 10,4$  vs  $13,5 \pm 4,3\%$  mmHg;  $p = 0,03$ ); their peripheral arteries showed increased PWV ( $6,8 \pm 1,2$  vs  $5,4 \pm 0,9$  m/s;  $p = 0,003$ ), while IMT was comparable ( $0,053 \pm 0,01$  vs  $0,051 \pm 0,01$  mm;  $p = 0,06$ ). Patients presented impaired vasodilatation: both FMD% ( $4,8 \pm 2,8$  vs  $8,5 \pm 2,3\%$ ;  $p = 0,00003$ ) and NMD% ( $11,3 \pm 4,6$  vs  $19,8 \pm 7,2\%$ ;  $p = 0,00001$ ). 46/54% patients presented recoarctation (gradient across the descending aorta  $\geq 20$  mmHg). Comparison of recoarctation to non-recoarctation patients revealed that resting systolic and diastolic pressures did not differ between the subgroups ( $143,3 \pm 14,7$  vs  $136,6 \pm 14,9$  mmHg;  $p = 0,06$  and  $81,2 \pm 9,3$  vs  $79,1 \pm 10,1$  mmHg;  $p = 0,99$ ), AP was higher in recoarctation patients ( $10,5 \pm 6,9$  vs  $7,5 \pm 4,1$   $p = 0,02$ ), but AI was comparable ( $21,0 \pm ,8$  vs  $19,3 \pm 8,5\%$   $p = 0,44$ ). Peripheral vascular parameters were similar in all subgroups. There was no significant linear correlation between age at the time of surgery and any of peripheral arterial parameters. Gradient across the ascending aorta correlates positively with AP ( $R = 0,295$ ,  $P = 0,01$ ).

**Conclusions:** Residual stenosis in the descending aorta does not affect the arterial vasodilatation nor stiffness in patients after CoAo repair. Early operation has no impact on peripheral vascular remodeling or central pressure which supports the claim that coarctation of the aorta is a systemic vascular disorder which leads to progressive vascular and end-organ damage despite early correction.

#### O4-2

##### **Male gender, cyanosis, high NYHA score, chromosomal abnormalities and a systemic right ventricle are independent risk factors for cardiac operations of**

##### **congenital heart defects in adulthood: A retrospective study on 530 patients.**

Vögt M. (1), Hörer J. (2), Otto D. (1), Grünewald S. (1), Schreiber Ch. (2), Kaemmerer H. (1), Hess J. (1).

(1) Department of Pediatric Cardiology and Congenital Heart Defects; (2) Department of Cardiovascular Surgery, Deutsches Herzzentrum München, Lazarettstr. 36, D-80636 München.

**Background:** Grown-ups with congenital heart defects are an increasing population requiring cardiac operations. The perioperative risk factors have not yet been identified.

**Objectives:** To clarify criteria for an unfavourable outcome or death we retrospectively analyzed all medical data of a cohort of 530 patients (age >16 years) operated on between 01/2004 and 12/2008. Composite end point was either death or prolonged intensive care of >4 days. 242 patients (45.7%) met these criteria (risk group). 13/530 (2.5%) died within 30 days after the operation.

**Methods and results:** We compared demographic data, cardiac risk factors, non cardiac anomalies, morphological criteria, hemodynamic and operative data of the risk group with the other patients. On univariate analysis gender male, endocarditis, cyanosis, impaired renal function and NYHA class >2 were significant risk factors. Functional single ventricle, systemic right ventricle with biventricular circulation, impaired function and enlargement of the systemic ventricle proved to be morphological risk factors. Out of 16 univariate risk factors seven proved to be significant on multivariate analysis: gender male ( $p = 0,001$ ), cyanosis ( $p > 0,005$ ), NYHA class >2 ( $p = 0,001$ ), chromosomal abnormalities ( $p = 0,004$ ), biventricular circulation with systemic RV ( $p = 0,029$ ), enlarged RV ( $p = 0,014$ ) and operation with extracorporeal circulation ( $p = 0,002$ ).

**Conclusions:** Operative outcome in the GUCH population is influenced by the patients conditions (gender male, chromosomal abnormalities), past history (cyanosis, NYHA class) and also by the underlying morphology (systemic RV). These informations on a large cohort of patients could help counselling more adequately individual adult patients with a congenital heart defect needing a cardiac operation.

#### O4-3

##### **Quality of life of adults after total cavopulmonary anastomosis**

Gouton M. (1), Soufi A. (1), Bozio A. (1), Ninet J. (2), Henaïne R. (2), Bernard Y. (1), Ducreux C. (1), Sassolas F. (1), Di Filippo S. (1) Pediatric Cardiology, Cardiovascular Hospital Louis Pradel, Lyon, France (1); Cardiac Surgery, Cardiovascular Hospital Louis Pradel, Lyon, France (2)

The objective of this study was to assess the quality of life and the impact of medical factors for patients with single-ventricle anatomy, in the long-term after total cavopulmonary anastomosis (TCPA).

**Material and methods:** patients were included if aged > 18years and follow-up after TCPA > 6months. The questionnaire SF-36 was used for physical, pain, mental and social health. Another questionnaire based on a linear scale aimed to established a score of overall quality of life and a weighted composite score. Sixty patients (24 females) aged  $15,4 \pm 8,5$  years at TCPA and  $10,4 \pm 5,9$  years after surgery, responded to the questionnaires.

**Results:** The scores of physical activity and the perceived general health were significantly lower than those of the general population. Scores of mental and social health, and vitality of the 25–34 years age-group were significantly impaired, but were similar to the general population for the other age-groups. The NYHA stage and occupation were correlated with physical



ability. The psychological status was linked to the number of drugs and work. The average score of quality of life allotted by the patient ( $7.02 \pm 1.6$ ) strongly correlated with the weighted composite score ( $r = 0.8$ ). Medical parameters (pulse oxymetry, cyanosis, heart failure, arrhythmia) did not impact on the scores. **Conclusion:** Adults with TCPA perceive impaired health at the physical level. Psychological disturbance is observed in the 24–35 years age-group, with subsequent resilience in older age-groups.

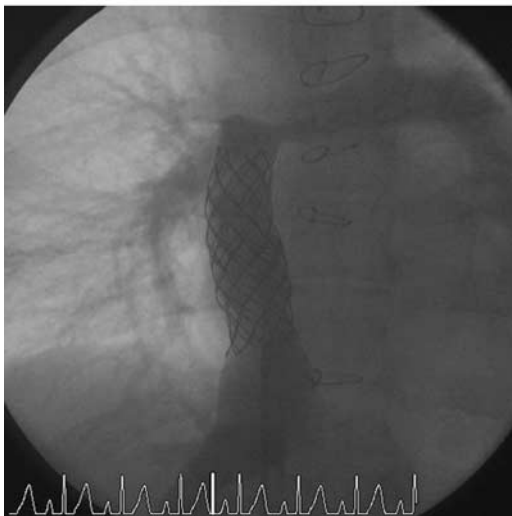
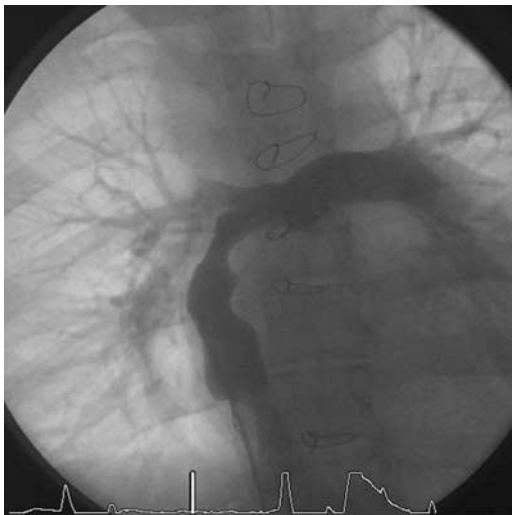
#### O4-4

##### Stenting of the lateral Tunnel in Fontan patients – does size matter in GUCH-Patients?

Haas N.A., Beck C, Laser T.K., Dubowy O., Kececioglu D., Blanz U., Sandica E.

Department for Congenital Heart Disease, Heart and Diabetes Centre North-Rhine Westfalia, Germany

**Introduction:** Total cavopulmonary connection (TCPC) including a lateral tunnel has been the palliation of choice in patients with univentricular physiology for many years. Today, in most centres an extracardiac conduit is placed to complete Fontan hemodynamics; the size of the conduit usually varies from 20 to 24 mm. In many of the adult (GUCH) TCPC-patients the clinical status deteriorates over time and some show significant narrowing of the lateral tunnel. Catheter investigation mostly reveals no or only minimal gradients.



**Methods:** We present our approach in 20 clinically symptomatic patients with TCPC and a morphologic narrowing of the lateral tunnel. There was impaired exercise tolerance in 20, protein losing enteropathy (PLE) in 2, veno-venous collaterals in 5, peripheral edema of the lower extremities in 12 and impaired flow profile in 20. Stenting of the lateral tunnel was performed to increase the size of the tunnel to 24 mm.

**Results:** Stenting was possible in all 20 patients. Clinical improvement was evident after treatment in 17/20 patients, establishment of normal flow in 20/20, improvement of PLE in  $\frac{1}{2}$ , occlusion of collaterals in 5/5, and resolution of edema in 12/12. There were no procedural complications.

**Conclusions:** In GUCH TCPC patients with clinical symptoms, especially those with impaired exercise tolerance and signs of lower body blood flow congestion, careful examination of the morphologic appearance of the lateral tunnels seems important. Augmentation of the narrowed segment to a size of modern extracardiac conduits seems favourable to relieve the clinical symptoms. The invasive blood-pressure gradient is apparently not indicative of treatment indication. In our opinion stenting is the treatment of choice and can be performed safely.

#### O4-5

##### Neurodevelopmental outcome in children and adolescents with functional single ventricle after total cavopulmonary connection (TCPC)

Vahsen N. (1), Bröder A. (2), Toussaint-Götz N. (1), Asfour B. (1), Schneider M. (1)

Deutsches Kinderherzzentrum Sankt Augustin (1); Rheinische Friedrich-Wilhelms-Universität Bonn, Institut für Psychologie (2); Germany

**Introduction:** Total cavopulmonary connection (TCPC) is the well-established surgical standard approach for definite palliation of functionally single ventricle hearts. Although the early and late mortality rates have decreased substantially in the last decades, knowledge about the neurodevelopmental outcome of these patients as well as the predictive value of potential risk factors (e.g. circulatory arrest time) on neurodevelopmental outcome is still inconsistent. Therefore the purposes of the present study were the standardized assessment of the patients' intellectual outcome after the modified Fontan operation and the retrospective analysis of peri- and intraoperative risk factors.

**Methods:** In 104 children aged two to 20 years (mean age  $8.6 \pm 3.9$  years) fluid and crystallized intelligence were either assessed by the Kaufman-Assessment Battery for Children (German version; 2–12.5 years) or the Kaufman Adolescent and Adult Intelligence Test (German version; 12.6–20 years). Patients' results were compared to published norms. Besides the relationship between intellectual outcome and potential risk factors was examined.

**Results:** Patients scored significantly lower on the scales of fluid ( $93.02 \pm 12.03$ ) and crystallized intelligence ( $92.26 \pm 14.44$ ) than the population mean ( $100 \pm 15$ ,  $p < .01$ ). Of all patients, 24% showed an below average performance (i.e.  $IQ < 85$ ). Fluid intelligence correlated significantly negative with the cumulative duration of circulatory arrest ( $r = -.23$ ,  $p = .02$ ). Furthermore children who experienced preoperative (i.e. prior to TCPC) complications causing suboptimal brain perfusion (e.g. low cardiac output, seizures) scored significantly lower on the fluid intelligence scale ( $88.56 \pm 14.18$ ) than children without such complications ( $95.10 \pm 10.63$ ,  $p = .04$ ).

**Conclusions:** Children with a congenital single ventricle physiology after a total cavopulmonary connection are at a high risk of

reduced fluid and crystallized intelligence. One fourth of the patients scored below average which, in the majority of cases, requires a special education. Especially those who experienced longer cumulative circulatory arrest times and/or preoperative complications causing suboptimal brain perfusion were at risk. Early diagnostics and interventions are necessary.

#### O4-6

##### **Recurrence of congenital heart disease in offspring of mothers with congenital heart disease screened prenatally by echocardiography**

Fesslova V. (1), Brankovic J. (1), Piazza L. (1), Villa L. (2), Meli V. (3)

*Policlinico San Donato IRCCS, Dpt. Ped. Cardiology/Center Fetal Cardiology, Milan, Italy (1); 1st Gyn. Obstet. Clinic, University of Milan, Italy (2); Dpt. Materno-Infantile, Policlinico Universitario, Palermo, Italy (3)*

**Background:** Maternal condition of congenital heart disease (CHD) bears a risk of recurrence and therefore these women are nowadays routinely referred for prenatal screening. Pre-conceptual counselling is very delicate in this population and the data relative to the recurrence rates are of extreme use in this field.

**Objective of the study:** to analyze the recurrence of congenital heart disease (CHD) in offspring of mothers affected with CHD.

**Material and methods:** Retrospective-prospective study of 260 pregnancies of 221 mothers with CHD studied by fetal echocardiography in our Centre between January 1995 and December 2008. Thirty four women were followed-up during 2 pregnancies and 5 during 3 pregnancies. Twenty three women, all operated, had cyanotic CHD, in 30 pregnancies, and 198 women had acyanotic CHD, operated in 128, in 230 pregnancies. Twenty seven women in 29 pregnancies had multiple familial risk (2–5 relatives).

**Results:** Eleven probands had CHD (total recurrence rate  $11/260 = 4.2\%$ , 1/30 pregnancies with cyanotic CHD (3.3%), 10/230 pregnancies with acyanotic CHD (4.3%). When mother alone was affected, the recurrence rate was 3.5% (8/231); when mother and another relative were affected the recurrence rate increased to 10.3% (3/29). The recurrence was higher in VSD (3/60 = 5%), ASD 4/71 = 5.6%, ductus arteriosus (1/12 = 8.3) and in AVSD (1/3). Concordant lesions occurred in 4 cases, partially concordant in 2, discordant in 5 cases.

**Conclusions:** Our data confirm a relevant recurrence of CHD in affected mothers. This fact has to be taken in account in prenatal counselling.

#### O4-7

##### **Role of electrophysiological study after surgical pulmonary valve replacement and right ventricular remodelling in GUCH patients with pulmonary regurgitation and aneurysmal RVOT dilatation**

Tavera M.C. (1), Foresti S. (1), Lupo P.P. (1), Ali H. (1), De Ambroggi G. (1), Pittalis M. (1), Giamberti A. (2), Chessa M. (3), Negura D. (3), Micheletti A. (3), Carminati M. (3) and Cappato R. (1).

*Center of Clinical Arrhythmia and Electrophysiology Policlinico San Donato IRCCS, San Donato Milanese, Italy (1); Cardiac Surgery Department Policlinico San Donato IRCCS, San Donato Milanese, Italy (2); Pediatric Cardiology Department, GUCH Unit, Policlinico San Donato IRCCS, San Donato Milanese, Italy (3)*

**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 baseline evaluation included Electrophysiological Study (EPS) with an aggressive protocol for induction of VT. When VT was induced, an electroanatomical map (EAM) of the RV was acquired. During surgery, RF ablation was performed at the critical isthmi of slowing conduction, identified by previous mapping, and aneurismatic tissue was removed. In all patients, EPS was repeated 6 months after surgery in order to evaluate arrhythmic risk.

**Results:** 43 patients were studied from October 2005 to October 2009. 7 of them presented clinical VT. In 9/43 patients VT was reproducibly inducible (5/9 experiencing clinical VT). In these 9 patients and in 1 non inducible patient (clinical VT) EAM and ablation was performed. Post-surgical EPS has been performed in 35/43 patients. VT at 6 months FU has been induced in 1/26 patients with negative baseline EPS and 4/9 patients with positive baseline EPS. In the first patient inducibility was suppressed by drug therapy, in 2 pts ICD has been implanted, while the other 2 pts have already received an ICD pre-operatively. At a median follow up of  $28 \pm 13$  months (6–48) no SCD was recorded. Clinical VT was observed in 2 pts, both of them with post-operative positive EPS and ICD. No patient with negative EPS experienced clinical VT or syncope.

**Conclusions:** Post-operative EPS seems to be predictive of clinical VT, mainly if combined with clinical data. EPS could be useful for management of patients at high risk of VT.

#### O5-1

##### **Relationship of Aortic Pulse Wave Velocity and Baroreceptor Sensitivity to Blood Pressure Control in Patients with Repaired Coarctation of the Aorta.**

Kenny D. (1), Polson J.W. (2), Martin R.P. (1), Cockcroft J.R. (3), Wilson D.G. (4), Caputo M. (1), Paton J.F.R. (2), Wolf A.R. (5)  
*Bristol Congenital Heart Centre, Bristol, UK (1); Department of Physiology, University of Bristol, UK (2); Department of Cardiology, University of Cardiff, UK (3); Department of Paediatric Cardiology, University Hospital Wales, Cardiff, UK (4); Department of Paediatric Anaesthesia, Bristol Royal Hospital for Children, Bristol, UK (5)*

**Objectives:** Increased aortic stiffness may lead to reduced cardiac baroreceptor sensitivity (sBRS). Both variables are independently associated with hypertension, which can develop after successful repair of coarctation of the aorta (CoA). We examined these variables in adolescent CoA patients with and without hypertension to examine the relationship between aortic stiffness, sBRS and hypertension.

**Methods:** We studied 29 adolescent patients following early effective CoA repair and compared data with 20 age-matched controls. CoA patients treated for hypertension, requiring late re-intervention or having residual aortic narrowing were excluded. We measured ambulatory blood pressure (BP), sBRS, heart rate variability, aortic pulse wave velocity (PWV) and cardiac output. **Results:** The CoA group had higher BP on ambulatory monitoring when compared to controls ( $131.2 \pm 3.0$  mmHg vs  $118.6 \pm 2.1$  mmHg,  $p = 0.008$ ). Nine patients (31%) were hypertensive according to standard definitions. These patients had higher aortic PWV than the CoA group without hypertension ( $p = 0.004$ ). There was a significant positive correlation between BP and PWV seen in the CoA group ( $r^2 = 0.5$ ,

$p < 0.01$ ) which was not present in controls ( $r^2 = 0.05$ ). Interestingly the normotensive CoA group had increased sBRS compared to controls ( $p = 0.009$ ). This difference in sBRS was not seen when comparing the hypertensive group to controls. The normotensive CoA group also had a significant reduction in stroke index compared to controls ( $p = 0.02$ ), which was not seen in the hypertensive group ( $p = 0.85$ ).

**Conclusions:** Approximately 30% of patients with early effective CoA repair are hypertensive by adolescence. These patients have higher aortic PWV than normotensive CoA patients. The normotensive CoA group has evidence of increased sBRS relative to controls, which may indicate a compensatory increase in cardiovascular activity, maintaining normal blood pressure via a reduction in cardiac output. Failure of this control mechanism may be the final stage to establishing hypertension in these patients.

	Hypertensive CoA	Normotensive CoA	Controls	P Value
Systolic BP (mmHg)	149.9 ± 2.5	121.9 ± 1.9	118.6 ± 2.1	$P < 0.001†$
PWV ( $\text{ms}^{-1}$ )	6.33 ± 0.34	5.35 ± 0.14	6.15 ± 0.21	$P = 0.004†$
sBRS ( $\text{msmmHg}^{-1}$ )	10.8 ± 1.1	14.0 ± 1.6	9.4 ± 0.6	$P = 0.009*$
SI ( $\text{mlm}^{-2}$ )	72.9 ± 6.0	62.2 ± 3.9	74 ± 3.2	$P = 0.02*$

Data are expressed as mean ± standard error. Statistical significance is defined as ( $P < 0.05$ )

† indicates Hypertensive CoA vs Normotensive CoA

\* indicates Normotensive CoA vs Controls

## O5-2

### Ventricular pressure-volume loops obtained by three dimensional echocardiography and mini pressure wire

Herberg U. (1), Bernhardt C. (1), Breuer T. (2), Doll U. (1), Knies R. (1), Schirrmester J. (1), Vlajnic D. (1), Breuer J. (1)

Paediatric Cardiology, Univ. Bonn (1); Computer Science, University of Applied Sciences, St. Augustin, Germany (2)

**Introduction:** Pressure-volume loops (PVL) provide vital information regarding myocardial function, systolic and diastolic performance as well as cardiac pathophysiology. Until now, the acquirement of PVL using conductance catheters is invasive, cumbersome and expensive. Real-time three-dimensional echocardiography (3DE) allows capture of volume changes of a chamber nearly instantaneously throughout the cardiac cycle. We assessed the feasibility of 3DE combined with pressure data from a miniaturized electromagnetic probe to calculate PVL in children.

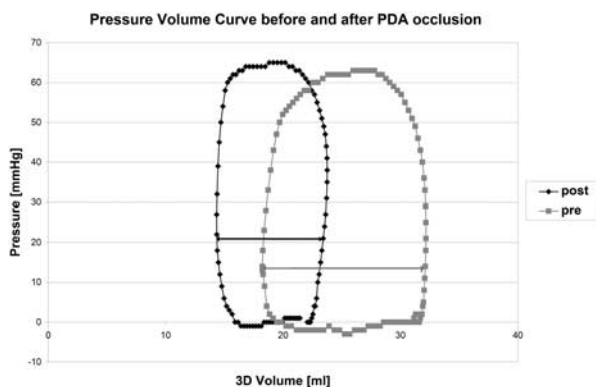


Fig. 1: Pressure volume loops calculated from 3DE volume data before (grey) and after (black) PDA occlusion: reduction of volume loading after occlusion (narrow PVL – black arrow), reduced enddiastolic and endsystolic volumes

**Methods:** In a series of 12 patients (mean age 17 [0–120] months), transthoracic 3DE was performed simultaneously to continuous pressure measurement using a mini pressure wire (RADI, 0.014"; St Jude) during interventional or diagnostic cardiac catheterization. PVL were calculated from contemporaneous 3D volume data and pressure curves. In 2 patients we simultaneously acquired PVL using conductance technology (Leycom).

**Results:** Computation of PVL using non-invasive 3D volume data and pressure curves obtained by mini pressure wire was easy, feasible and reproducible in all patients. In contrast, conductance technology was time consuming and needed additional use of a relatively large-sized conductance catheter and extra calibration. PVL calculated from conductance technology were less reproducible than PVL calculated from 3D echo data.

**Conclusions:** Calculation of pressure-volume loops from 3DE volume curves and simultaneous pressure data obtained by a mini pressure wire is a reliable and reproducible method to assess cardiac function. Due to the small size of the pressure wire used PVL can be acquired even in preterms and neonates.

## O5-3

### Exercise Stress Echocardiography as a Novel, Effective Method to Detect Coronary Allograft Vasculopathy in Paediatric Heart Transplant Recipients: Comparison with Myocardial Perfusion SPECT Imaging

Lunze FI. (1), Gauvreau K. (1), Bergersen L. (1), Geva T. (1), Colan S.D. (1), Blume E.D. (1), Chen M.H. (1)

Harvard Medical School, Children's Hospital Boston, Department of Cardiology (1)

**Background:** Coronary allograft vasculopathy (CAV) is the leading cause of graft loss for children who survive the first year after heart transplant. Exercise stress echocardiography (stress echo) and myocardial perfusion SPECT have emerged as non-invasive options to screen for CAV. We reviewed preliminary safety and efficacy of exercise stress echo in comparison to myocardial perfusion SPECT for the detection of CAV in pediatric cardiac transplant recipients.

**Methods:** All heart transplant patients who underwent routine coronary angiography (angio), stress echo, and also myocardial perfusion SPECT with Tc99m-sestamibi (Stress-MIBI) within 6 weeks of each other from January 2007–August 2009 at our institution were included. Stress echo imaging was performed at rest and within 90 seconds upon completion of exercise, and adverse events were documented. Positive stress echo for ischemia was defined as development or worsening of wall motion abnormalities with exercise. Myocardial perfusion SPECT was performed at rest and after exercise. Positive stress-MIBI for ischemia was defined as development of any reversible perfusion defects. Positive angio was used as the gold standard for diagnosis of CAV. Sensitivity, specificity, positive (PPV) and negative predictive values (NPV) with 95% confidence limits (CI) were calculated using standard definitions.

**Results:** The study population consisted of 23 consecutive cardiac transplant patients, 52%(12) female, mean age  $17.2 \pm 4.5$  yrs, mean time post-transplant  $8.1 \pm 4.5$  yrs. Stress echo and Stress-MIBI were successfully performed without complications in all subjects. Stress echo had 75% sensitivity (95%CI [19%, 99%]) and 100% specificity ([82%, 100%]) to detect ischemia in pediatric cardiac transplant recipients. PPV of stress echo was 100%, while the NPV was 95%. Stress echo was interpreted as normal in 1 patient with distal vessel CAV.

Stress-MIBI showed 0% sensitivity (95%CI [0%, 60%]) and 89% specificity ([67%, 99%]) to detect CAV in our cohort of pediatric

cardiac transplant recipients. PPV of MIBI exercise test was 0% and the NPV was 81%.

Methods		Angiography		Total
		Positive for CAV	Negative for CAV	
Exercise stress echocardiography	Positive for CAV	3	0	3
	Negative for CAV	1	19	20
Total		4	19	23

**Conclusions:** Exercise stress echocardiography is well-tolerated and may be a useful non-invasive screening tool for detection of CAV for pediatric heart transplant recipients. In our small series, stress echo was superior to stress-MIBI as a screening test for ischemia. Further definition of exercise stress echocardiography's performance characteristics and utility in larger clinical series are warranted.

#### O5-4

##### Results of cardiac surgery during neonatal period – focus on morbidity, psychomotor aspects and quality of life

Wolf N. (1), Woerner C. (1), Schulze J. (1), Henjes S. (1), Kostelka M. (2), Janousek J. (1), Daehnert I. (1)  
 Pediatric Cardiology, University of Leipzig, Heart Center, Leipzig, Germany (1); Pediatric Cardiac Surgery, University of Leipzig, Heart Center, Leipzig, Germany (2)

**Introduction:** Based on low mortality after contemporary neonatal cardiac surgery (own data, 1998–2009, N = 630, 30 days mortality = 4.6%) our study focused on morbidity, psychomotor development, social aspects, and quality of life.

**Methods:** All patients (N = 166) after corrective (N = 93) or palliative (N = 73) cardiac surgery during neonatal period operated in 3 calendar years (2000, 2003, 2006) in 1 center were included. Mean age at surgery was 7 days, median Aristotle complexity score 9.0. Postoperative data were retrieved from medical records. Structured questionnaires were separately sent to parents and pediatricians of 120 consecutive neonates (excluding the deceased and patients with syndromes or primary neurological affection).

**Results:** 10 early (6.0%) and 18 late (10.8%) deaths occurred. The median stay on ICU was 12 days (2–226), including median 5 ventilation days (1–150). Secondary chest closure was performed in 59/166 (35.5%), postoperative ECMO was necessary in 6/166 (3.6%) and non-obligatory surgical/catheter re-interventions (those given by staged surgical strategy excluded) in 74/166 (44.6%) of patients. Until now completed questionnaires could be obtained from 75/120 survivors at a median age of 6.1 years. 75% of children had an overall normal development. 60% had normal exercise tolerance, 76% normal gross motor skills, 70% normal fine motor skills, 84% normal speech comprehension, 63% normal active speech, 22% behavioural disorders, and 23% concentration difficulties. 94% have normal visual acuity and 96% normal hearing. 55% of all families reported no, 35% minor, 7% moderate and 1% severe limitations in life because of the heart disease of their children. Changes in the professional career of parents or in the place of family residence due to children's heart disease were reported in 10/75 resp. 3/75 families. 23% get special financial or social aid (attendance allowance or disabled person's pass). 40% have daily cardiac medication.

**Conclusion:** Preliminary analysis of quality of life, psychomotor development and social aspects after neonatal cardiac surgery is

favourable. 90% of the families have no or only minor limitations in life because of the heart disease of their children.

#### O5-5

##### Hypothermia during repair of ventricular septal defects (VSD) in infants prolongs postoperative mechanical ventilation and inotropic support

Fedarava K. (1), Schmitt K. (1), Hübler M. (2), Berger F. (1,3), Miera O. (1), Hetzer R. (2)

Dept. of Congenital Heart Disease/Pediatric Cardiology, Deutsches Herzzentrum Berlin, Germany (1); Clinic for Cardiothoracic Surgery, Deutsches Herzzentrum Berlin, Germany (2); Pediatric Cardiology, Charité-Universitätsmedizin, Berlin, Germany (3)

**Background:** Cardiopulmonary bypass (CPB) triggers the whole body inflammatory response, and it has been suggested that hypothermia may influence these responses. The aim of this prospective study was to compare the inflammatory response and postoperative course in children undergoing CPB for VSD closure, randomized to groups with moderate hypothermia (32°C) or normothermia.

**Methods:** We measured inflammatory markers in dried blood spots of 20 infants with body weight from 5 to 10 kg undergoing open heart surgery who were randomized to surgery with either hypothermia (32°C) or normothermia. Blood was sampled after induction of anesthesia, after CPB, and at 2 hours and 24 hours postoperatively.

**Results:** Hypothermic CPB had no influence on the inflammatory response, fluid balance or blood loss. Unexpectedly hypothermia led to significantly longer mechanical ventilation (27.7 +/- 26.4 h versus 14.0 +/- 7.7 h; p = 0.009) and longer (10.3 +/- 9.75 h versus 4.53 +/- 4.12 h; p = 0.007) as well as higher (27.3 +/- 44.4 versus 5.6 +/- 7.1 mcg/kg; p = 0.003) inotropic support. In three infants postoperative complications were observed: Two patients had a junctional ectopic tachycardia, one a pulmonary hypertension crisis – all occurred in the hypothermia group.

Although temperature treatment does not influence cytokine release, CPB time under hypothermic conditions was directly correlated to increased levels of cytokines. During hypothermic CPB we found a strong correlation between duration and IL-6, IL-10 and IL-8 release, which interestingly could not be demonstrated during CPB under normothermia.

**Conclusions:** Prophylactic moderate hypothermia during corrective cardiac surgery in infants had a negative influence on postoperative course. If cardiac surgery with hypothermic CPB is not avoidable, the time should be kept as short as possible to reduce the inflammatory response and possible adverse clinical effects.

#### O5-6

##### Long- and midterm results of the Growth Stent for the transcatheter treatment of aortic coarctation in infants

Ewert P., Schubert S., Yilmaz S., Peters B., Miera O., Ovroutski S., Nagdyman N., Berger F.

Department of Congenital Heart Diseases, German Heart Institute Berlin, Berlin, Germany

**Introduction:** There is currently no stent available for possible curative transcatheter treatment of aortic coarctation in infants. To overcome this problem we designed a stent, the "growth stent", which was tested to be not restrictive during growth in an animal experiment. We evaluated the feasibility and usefulness of the stent for the treatment of aortic coarctation in infants in a

pilot study since April 2002. We report on the long term follow-up of the first 14 patients.

**Methods and results:** Between April 2002 and January 2008 a total of 18 growth stents were implanted in 17 patients with coarctation or recoarctation. Three patients died independently of the stent due to severe further underlying complex congenital heart diseases (HLHS 2x, unbalanced AVSD with hypoplastic LV 1x). Thus, a subgroup of 15 stents in 14 patients were analysed with a median follow-up of 5.4 years (range 2.0 to 7.9 years). At implantation, the median age was 3.5 months (range 10d to 37m), the bodyweight 5.4 kg (3.5–13.5 kg). Eleven patients suffered from aortic (re-) coarctation, three from stenosis of the aortic anastomosis after a Norwood I procedure. Pressure gradients immediately after stent implantation dropped from 40 mmHg (range 20 to 60 mmHg) to 6 mmHg (range 0 to 15 mmHg). Five patients had one (3pts) or two (2pts) balloondilations three to 28 months (median 12 months) after growth stent implantation. The median pressure gradient dropped from 25 mmHg (range 15 to 30 mmHg) to 15 mmHg (range 5 to 25 mmHg). Nine patients received a large stent 27 months (18–53 months) after Growth Stent implantation. Median body weight was 11.8 kg (9.4 to 15 kg) and implantation was performed through sheaths of 6F to 9F (median 7F). No patient has been operated on the stented coarctation so far.

**Conclusions:** The growth stent is suitable for the treatment of aortic coarctation in infants and can be overstented later on – whenever necessary – with a larger stent without causing restriction. Mid- and long term results up to a follow-up of nearly eight years are excellent without the need for surgery.

#### O5-7

##### **Prognosis of non-compaction cardiomyopathy in the pediatric population**

*Veyrier M. (1), Teyssier G. (3), Henaine R. (2), Sassolas F. (1), Ducreux C. (1), Gouton M. (1), Ninet J. (2), Bozio A. (1), Di Filippo S. (1)*

*Pediatric Cardiology, Cardiovascular Hospital Louis Pradel, Lyon, France (1); Cardiac Surgery, Cardiovascular Hospital Louis Pradel, Lyon, France (2); Pediatric Intensive Care, Hopital Nord, Saint-Etienne, France (3)*

The aim of this study was to determine factors of outcome in children diagnosed with non-compaction of the myocardium.

**Material and methods:** Patients less than 18 years of age with diagnosis of non-compaction of the myocardium (according to echocardiographic criteria) were reviewed for clinical, ECG and echocardiographic data and outcomes. Cases with structural congenital heart disease were excluded.

**Results:** From 1996 to 2009, 31 patients (17 males), aged 0 to 207 months at diagnosis (median 7.5 months), were followed-up for 0.5 to 99 months (median 12.5 months). Seventeen presented with heart failure (55%), 14 (45%) with syncope or chest pain or no symptom. Familial recurrence was 48%. ECG was abnormal in all cases, QT > 440 ms in 22.5%. N/C index was  $1.6 \pm 3.5$  (median 2.3) and >2 in 54.8% (17 cases). LV apex was involved in 96.7%, with more than 3 locations in 29% and RV involvement in 13%. Shortening fraction was  $24.8 \pm 11.6\%$  (median 24%). Six patients died (19.3%) at median age 7.7 months (3.6 to 43) and median follow-up 2.7 months (0.5 to 12.7). Five underwent heart transplantation (16%) at median age 12 months (6 to 72) and median follow-up 2.8 months (1.8 to 27.8). At least one episode of heart failure occurred in 17 (57%), arrhythmic or thromboembolic events in respectively 4 (13%) and 3 (9.6%). Freedom from death or transplantation was 70% at 6 months and 60% at 1 and 5

years follow-up. Age at diagnosis < 1 year, low shortening fraction, N/C > 2, three locations or more, heart failure and thromboembolic events were factors for poor prognosis.

**Conclusion:** Non-compaction cardiomyopathy in children carries on high rate of mortality and morbidity. Heart loss occurs more frequently within 3 months after diagnosis in infants with severely impaired systolic function and/or thromboembolic events.

#### O6-1

##### **Impact of antenatal diagnosis on outcomes of neonates with coarctation of the aorta**

*Di Filippo S. (1), Abid D. (1), Henaine R. (2), Sassolas F. (1), Ducreux C. (1), Ninet J. (2), Bozio A. (1)*

*Pediatric Cardiology, Cardiovascular Hospital Louis Pradel, Lyon, France (1); Cardiac Surgery, Cardiovascular Hospital Louis Pradel, Lyon, France (2)*

The objective of the study was to review coarctation of the aorta (COAO) in neonates and assess the influence of antenatal diagnosis on prognosis.

**Material and methods:** all patients diagnosed with COAO who underwent surgery before 2 months of life, from 2004 to 2008, were included.

**Results:** Among 106 neonates (63 males) admitted for diagnosis of COAO at median age 5.5 days, 29 (27.3%) had antenatal diagnosis (24 ventricular asymmetry, 5 aortic arch hypoplasia). The other 81 patients presented with heart failure (23 mild, 15 moderate, 19 cardiogenic shock) or no symptom (18 absence of femoral pulse, 6 murmur). PGE was required in 65 (61.3%), mechanical ventilation in 56 (52.8%: 77% of patients with PGE). Ventricular septal defect was associated in 50 (47%), aortic valve bicuspidia in 53 (50%); mitral valve was normal in 100 (94.3%). Antenatal diagnosis was associated with more severe anatomy (LV diameter: 15.7 vs 18.2 mm,  $p = 0.004$ ; ascending aorta: 7.2 vs 8.2 mm,  $p = 0.01$ ), less heart failure ( $p < 0.0001$ ), less mechanical ventilation ( $p = 0.008$ ), higher LV shortening fraction ( $p = 0.03$ ) and lower pulmonary pressure ( $p = 0.03$ ). Surgery (Crafoord: 82, Crafoord + banding: 14, Crafoord + VSD patch: 9) was performed at median age 12 days (2 to 63 days: earlier in antenatal diagnosis cases: 11.5 vs 21.5 days,  $p = 0.04$ ), median weight 3 kg (1.6 to 5.3), median time from diagnosis 3 days (0 to 59). Postoperative complication occurred in 20 cases (19%), more frequently in case of VSD closure (62.5%). Median CICU stay was 8 days (1 to 80), hospital stay 11 days (1 to 80), longer in antenatal diagnosis cases: 11.5 vs 21.4 days,  $p = 0.02$ . Aortic restenosis occurred in 6 cases (6%), 45 to 340 days postoperative (median 79 days), and was associated with postoperative complication. Freedom from restenosis at 1 month, 2 months and 1 year was respectively 94% and 90%. One patient died before surgery, 3 within 2 months after surgery. Antenatal diagnosis did not impact on postoperative course, nor survival, nor restenosis.

**Conclusion:** Antenatal diagnosis can detect COAO with most severe anatomy (27.3% of all), allows to anticipate heart failure and LV impairment and leads to earlier surgery, but does not impact on long-term outcome.

#### O6-2

##### **Impact of Prenatal Diagnosis on Outcome of Pulmonary Atresia with Intact Ventricular Septum**

*Tuo G. (1)(2), Serafino M. (1), Volpe P. (3), Zannini L. (1), Petrucci L. (4), Calevo M.G. (5), Marasini M. (1).*

*Department of Pediatric Cardiology and Cardiac Surgery, G. Gaslini Institute, Genoa, Italy (1); Department of Internal Medicine, Cardiology Unit, University of Genoa, Genoa, Italy (2); Fetal Medical Unit,*

Di Venere and Sarcone Hospital, Bari, Italy (3); Department of Intensive Care Unit, G. Gaslini Institute, Genova (4); Epidemiology and Biostatistics Service, Scientific Directorate, G. Gaslini Institute, Genoa, Italy (5)

**Objectives:** To assess the impact of foetal echocardiography on the management of pregnancy and of newborns affected by Pulmonary Atresia and Intact Ventricular Septum (PAIVS) and to compare the outcomes among infants with and without prenatal diagnosis.

**Methods:** All patients with PAIVS diagnosed prenatally (Group A) and postnatally (Group B) from January 1994 to May 2009 were reviewed. Radiofrequency (RF) perforation of the pulmonary valve (PV) was the first procedure in all cases without monopartite right ventricle (RV) and/or RV dependent coronary circulation (RVDCC). For foetuses with a favourable RV morphology the delivery by caesarean section was scheduled at 36 weeks of gestational age (GE) to anticipated the PV perforation, hoping to break the intracavity overgrowth and to maximise the chance of an eventual biventricular-repair.

	Group A (22)	Group B (27)	p value
Gender (%)			*
Male	12 (55)	20 (70)	
Female	10 (45)	7(30)	
GA at birth (week) mean $\pm$ SD	36.5 $\pm$ 2	39.7 $\pm$ 0.7	<0.0001
Weight at birth (g) mean $\pm$ SD	2540 $\pm$ 454	3310 $\pm$ 608	<0.0001
"Partite status" of the RV (%)			0.03
Monopartite	5(23)	2 (7)	
Bipartite	11 (50)	8 (30)	
Tripartite	6(27)	17 (63)	
Tricuspid Z value mean $\pm$ SD	-2.90 $\pm$ 2.2	-1.31 $\pm$ 1.8	0.01
Pulmonary Z value mean $\pm$ SD	-2.52 $\pm$ 1.4	-1.63 $\pm$ 1.5	0.05
RV-CA fistulas	5(23)	2(7)	*
RV-dependent coronary circulation	3(14)	0	*
PGE1 infusion duration (days) mean $\pm$ SD	6.5 $\pm$ 3.6	7.2 $\pm$ 3.6	
mBT shunt as initial palliation (%)	5(23)	2(7)	
RF perforation at birth (%)	17(77)	25 (93)	
Age at RF perforation (hours) mean $\pm$ SD	3.41 $\pm$ 3.6	46.64 $\pm$ 18.1	<0.0001
Needing of mBT shunt +/- RVOT after RF perforation (%)	6 (35)	12 (48)	
Follow up (years) mean $\pm$ SD	2.8 $\pm$ 3.9	5.6 $\pm$ 4.2	0.02
Late result (%)			
Biventricular repair	13 (59)	22 (81)	
One and half ventricular repair	1 (5)	3(11)	
Univentricular repair	3(14)	2(7)	
Postnatal Death			0.03*
Early death (%)	3(14)	0	
Late death (%)	2(9)	0	

\*Fisher exact test

**Results:** Fifty patients were retrospectively identified. No false positive or negative prediction of PAIVS occurred in our series during the same period. Among Group A there was one intrauterine death at 32 weeks of GE and no termination of pregnancy; five patients had a unipartite ventricle of whom 3 died early whereas the others had a univentricular repair; two infants of group B showed a unipartite ventricle and both had a univentricular repair. Z score for tricuspid valve and PV were significantly lower in Group A. RF perforation was successfully performed in 17 and 25 patients of groups A and B respectively and at the correspondent follow up, among group A, 3 (18%) premature infants died early and one died later during a surgical RVOT enlargement, 13 (76%) patients achieved biventricular repair without any other operation in 8 of them (62%) and 1 (6%) had a univentricular repair; anyone of Group B died, 22 patients (88%) had a biventricular repair without any other procedure in 16 of them (73%), and 3 (12%) underwent bidirectional Glenn procedure (Table).

**Conclusions:** Prenatal echocardiography can accurately diagnose PAIVS and usually detects the cases with the worst morphological features. In spite of this, no significant differences were found among patients with and without prenatal diagnosis who underwent RF perforation as for mortality and achievement of biventricular repair.

### O6-3

#### 3D reconstruction of the first and early second trimester human fetal heart

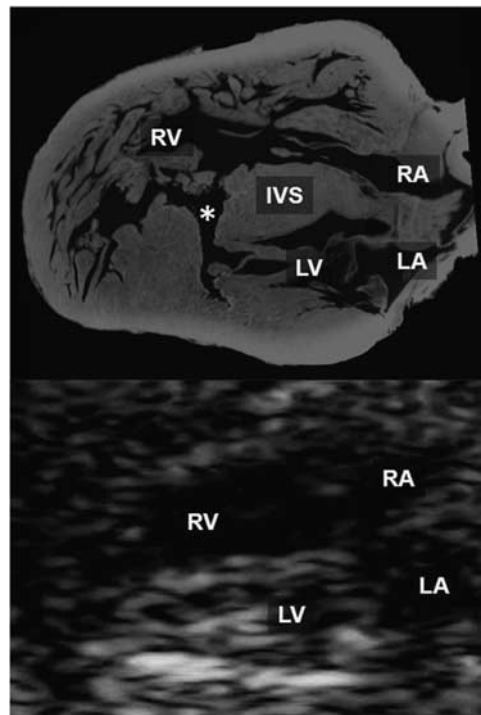
Gardiner H. (1), Matsui H. (1), Mohun T. (2), Ho S.Y. (3)  
 Division of Cancer, Faculty of Medicine, Imperial College at Queen Charlotte's and Chelsea Hospital, London (1); Division of Developmental Biology, MRC National Institute of Medical Research, London (2); Cardiac Morphology Unit, Royal Brompton Hospital and National Heart and Lung Institute, Imperial College London, UK (3)

**Introduction:** High Resolution Episcopic Microscopy (HREM) produces high quality three dimensional images from small samples in a variety of species, including the human where we described the normal first trimester heart shows large atrial appendages with relatively small atrial chambers, prominent coronary arteries and spiral arrangement of the ventricles. (1,2).

**Aims:** To investigate HREM three dimensional reconstruction of the first and early second trimester human fetal heart at postmortem and compare with antenatal ultrasound findings in Hypoplastic Left Heart (HLH).

**Setting:** Tertiary fetal medicine, developmental biology and morphology departments.

**Design:** Observational comparative study.



**Methods:** At postmortem the fetal heart is photographed, embedded in resin and then processed by automatic sectioning with serial photography of each surface of the sectioned block allowing 3D reconstruction of highly aligned digital images modelled with an isotropic resolution of 3 microns. Volume rendering allows examination of surface features, such as coronary arteries and multi-planar virtual sectioning provides detailed morphological information.

**Results:** In the 15 week HLH heart the course of the left anterior descending coronary artery delineates the left ventricle (LV). Multi-planar virtual sectioning revealed a small muscular ventricular septal defect\* not visualised on echo. (Figure) Histological staining confirmed the defect was not an artefact. The aortic valve was unifoliate and unicommissural (unicuspid) and mild dysplasia of the posterior leaflet of the mitral valve was confirmed on histological staining.

**Conclusions:** For clinical audit and training, HREM allows detailed morphological assessment of the early human fetal heart.

#### References.

1. Weninger WJ, Geyer SH, Mohun TJ, Rasskin-Gutman D, Matsui T, Ribeiro I, Costa Lda F, Izpisua-Belmonte JC, Muller GB. High-resolution episcopic microscopy: a rapid technique for high detailed 3D analysis of gene activity in the context of tissue architecture and morphology. *Anat Embryol (Berl)*. 2006;211(3):213–221.
2. Matsui H, Mohun TJ and Gardiner HM. Three dimensional reconstruction imaging of the human foetal heart in the first trimester. *European Heart Journal* 2009; doi: 10.1093/eurheartj/ehp511.

#### O6-4

##### **Prolongation of the Atrio-Ventricular Conduction in Fetuses Exposed to Maternal Anti-Ro/SSA Antibodies does not Predict Progressive Heart Block**

Jaeggi E. (1), Silverman E.D. (2), Golding F. (1), Weber R. (1)  
Divisions of Cardiology and Rheumatology (1); The Hospital for Sick Children, Toronto, Canada (2)

**Background:** It has been suggested that anti-Ro antibody-associated congenital complete atrio-ventricular block (CAVB) may be preventable if detected and treated early when low-grade heart block is present. Consequently, dexamethasone has been advocated and used to treat prolonged fetal AV conduction (AVC) >2 z-scores, consistent with a diagnosis of 1st degree AV block (Friedman et al, *Circulation* 2008; Rein et al, *Circulation* 2009). We examined the prevalence and outcome of AV prolongation without steroid treatment.

**Methods:** Between 7/2003 and 12/2008, 150 fetuses of 129 anti-Ro antibody-positive women were enrolled in this prospective, single center study. Antibody-levels were confirmed by ELISA. Our protocol included weekly evaluation of the fetal AVC by echocardiography between 19 (17–23) and 24 (23–35) gestational weeks. AV times were compared to institutional reference data (Nii et al, *Heart* 2006) and postnatal electrocardiogram.

**Results:** Of 137 cases with persistently normal AVC throughout the observation period, one was subsequently diagnosed with CAVB at 28 weeks, after the serial evaluation had ended. Of 11 untreated fetuses with AV time measurement between 2 and 3 z-scores, none had postnatal AV block. Of 2 untreated cases with persistent AV prolongation between 3 and 6 z-scores, only one had 1st degree block at birth which has not progressed postnatally. No other cardiac complications were detected. Thus, fetal AV prolongation between 2 and 6 z-scores had a low positive predictive value of 8% in detecting 1st degree block after birth and a 0% post-test probability of CAVB.

**Conclusions:** Our findings question the rationale of a management strategy that relies on the identification and treatment of AV prolongation to prevent CAVB. When it occurred, CAVB was not predicted and untreated 1st degree block did not progress.

#### O6-5

##### **Impact of prenatal diagnosis on outcome of univentricular heart disease**

Stiasny B., Fasnacht M., Prêtre R., Bauersfeld U., Valsangiacomo Buechel E.R.  
University Children's Hospital Zurich, Switzerland

**Background:** Congenital heart disease (CHD) with single ventricle (SV) physiology is a rare and complex condition. The systemic- or the pulmonary circulation is frequently duct-dependent. Prenatal diagnosis may help to optimise postnatal management, by immediate administration of prostaglandin. However treatment and prognosis may remain challenging for professionals and families. We sought to assess the impact of prenatal diagnosis of SV CHD on the neonatal survival and on the early postoperative outcome of these children.

**Methods:** The medical records of 117 consecutive patients with SV diagnosed either prenatally or postnatally were retrospectively analysed regarding termination of pregnancy, prenatal or postnatal death, treatment, early postoperative morbidity and mortality. The influence of prenatal diagnosis on survival and postoperative morbidity was assessed.

**Results:** Of 65 (55%) prenatally diagnosed cases, 15 (13%) pregnancies had been interrupted and one intrauterine death occurred. Additional 52 (45%) SV were diagnosed postnatally, giving a total of 101 liveborns. Treatment, consisting of cardiac surgery (n = 49) or catheter-guided intervention (n = 19) was initiated in 57 (56%), comfort care adopted in 26 (6%) neonates and two children did not require neonatal intervention. Outcome parameters of the prenatal- and postnatal diagnosed groups are shown in Table 1.

	prenatal	postnatal
Prostaglandin at admission	79%	56%
Intubation at admission	6%**	42%**
pH at admission	7.29 ± 0.09	7.31 ± 0.12
Lactate at admission	3.09 ± 1.84	6.48 ± 8.19
Catecholamine preop	7%**	32%**
CPR preoperatively	0	13%
Intubation time (d)	5.46 ± 5.44	7.58 ± 7.30
Catecholamine postop	85%**	100%**
CPR postoperatively	15%	24%
ICU stay (d)	12.36 ± 7.80	16.00 ± 10.47
Hospitalisation time (d)	37.69 ± 31.65	44.48 ± 41.19
Overall mortality	55%	40%
Early mortality	11%	23%
Drugs on day 30	2.23 ± 1.28	2.13 ± 1.43

\*\*p < 0.05

**Conclusions:** Univentricular CHD remains a lesion with high neonatal mortality and morbidity. Prenatal diagnosis may lead to better preoperative patient's conditions but does not improve postoperative outcome.

#### O6-6

##### **Cardiac function between 11 and 35 weeks' gestation in trisomy 21 fetuses**

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

**Background:** An increased nuchal translucency (NT), a recognised marker for trisomy 21, has been associated with cardiac defects (CHD) and possible cardiac dysfunction.

**Aim:** Determination of cardiac function throughout gestation in fetuses with trisomy 21 (T21).

**Methods:** Echocardiography was performed on 48 trisomy 21 fetuses, 191 normal controls, (88 with a normal NT and 103 with a NT  $\geq$  95th percentile), between 11 and 35 weeks' gestation.

**Measurements performed:** E- and A-wave peak velocity, E/A ratio, E/TVI ratio over the atrioventricular valves, myocardial performance index, acceleration time (AT) and peak velocity (PVel) over the semilunar valves, stroke volume and cardiac output and ductus venosus pulsatility index for veins at 11–14 weeks'.

**Results:** 11–13.9 weeks' gestation: Aortic and pulmonary PVel and right ventricular (RV) A-wave velocity were significantly lower and RV E/A velocity ratio significantly higher in the T21 fetuses. The TV E-velocity was significantly lower in the T21 fetuses without CHD compared to the controls with an increased NT. 14 and 21.9 weeks' gestation: RV and left ventricular (LV) E- and A-wave velocities were significantly reduced in T21 fetuses. The aortic PVel and LV E/TVI were significantly lower in T21 fetuses without CHD compared to the controls, while the Aortic AT was also significantly lower compared to the controls with an increased NT. 22 and 35 weeks' gestation: RV and LV A-wave velocity, RV E/TVI and aortic PVel were significantly reduced and the pulmonary AT significantly increased in the T21 fetuses. **Conclusions:** Cardiac function is altered in T21 fetuses, even in the absence of CHD and irrespective of NT thickness. Our findings suggest a maturational delay in both systolic and diastolic function with loading effects, predominately preload between 11–13.9 weeks and increased LV afterload after 14 weeks' gestation.

#### O6-7

##### **Prenatal brain pathology in congenital heart disease – does oxygen saturation of cerebral blood influence its occurrence?**

McLzoch E. (1), Brugger P. (4), Hanslik A. (1), Ulm B. (3), Salzer-Muhar U. (1), Prayer D. (3)

(1) Clinical Division of Pediatric Cardiology; (2) Clinical Division of General Pediatrics and Neonatology, Department of Pediatrics and Adolescent Medicine, Medical University of Vienna, Austria; (3) Clinical Department for Neuroradiology, University Clinic of Radiology, Medical University of Vienna, Austria; (4) Clinical Department of Obstetrics and Fetomaternal Medicine, University Clinic of Obstetrics and Gynaecology, Medical University of Vienna, Austria

**Aim:** To identify the influence of lower than normal oxygen saturation of cerebral blood on fetal brain pathology in fetuses with CHD.

**Methods:** 67 pregnant women underwent 1 or 2 fetal MR-examinations between 20 and 38 gestational weeks. MR was done on a 1.5 T superconducting system. The type of cardiac malformation was defined by fetal echo. Fetuses with a defined chromosomal abnormality or suspected dysmorphic malformation were excluded.

**Study population:** In total, 51 fetuses were included into the study. 20 fetuses had a right heart pathology (group 1), 9 had a left heart pathology (group 2) and 22 fetuses had either a complex pathology or a shunt lesion (group3). A lower than normal oxygen saturation in the carotid arteries and cerebral blood due to the altered hemodynamics was hypothesized in a subgroup of 30/51 fetuses (59%).

**Results:** 32 fetuses had a brain pathology and 19 had a regular brain development at the time of investigation. The association between CHD and fetal brain pathology itself was significant ( $p = 0.01$ ). Fetuses in group 3 had significantly more abnormal MRT findings than fetuses in group 1 or 2 while this was not true for those fetuses supposed to have lower than normal cerebral blood oxygen saturation.

**Conclusion:** Fetal MRI of the brain in CHD is a new method to investigate early onset signs of changes in fetal brain development. Brain injury in fetuses and newborns with congenital heart disease is probably multifactorial and not only influenced by oxygen saturation of cerebral blood.

#### O7-1

##### **Epicardial ablation of ventricular tachycardia in arrhythmogenic right ventricular cardiomyopathy in children**

Pokushalov E., Romanov A., Artemenko S., Shugayev, Turov A., Shirokova N.

State Research Institute of Circulation Pathology, Novosibirsk, Russian Federation

**Introduction:** Efficacy of endocardial ventricular tachycardia (VT) ablation in arrhythmogenic right ventricular cardiomyopathy in children may be limited. The aim of this study was to evaluate the safety and efficacy of epicardial VT ablation in children with right ventricular cardiomyopathy.

**Methods:** Sixteen patients (aged 13 years; range, 11 to 16 years) underwent endocardial and epicardial mapping and epicardial VT ablation after failed endocardial VT ablation were included.

**Results:** Nineteen VTs were localised on the epicardium lateral wall with the use of activation, entrainment, or pace mapping. Two VTs demonstrated a reentry circuit and remaining 17 VTs showed a focal endocardial activation pattern. Low voltage areas (LVA) were observed on all these maps. LVA localised close to the tricuspid annulus on the lateral free wall and/or the inferior wall of the right ventricle. Epicardial VTs were localised opposite ineffective endocardial ablation sites in 4 patients (25%) and/or adjacent ineffective endocardial ablation sites in 11 patients (87.5%). The basal right ventricular thickness assessed by electroanatomic map was  $>7$  mm in 7 of 16 patients. During  $16 \pm 9$  months, 14 of the 16 patients (87.5%) had no VT and 2 patients had VT recurrence with the same morphology.

**Conclusions:** It is reasonable to suggest that epicardial catheter ablation using transpericardial approach may be an effective alternative to endocardial catheter ablation of ventricular tachycardia in children with an epicardial arrhythmogenic substrate.

#### O7-2

##### **Arrhythmia in patients older than 40 years undergoing transcatheter closure of atrial septal defect**

Wang, JK, Wu, MH, Lin, FY, Lin, JL, Tsai, SK  
National Taiwan University Hospital, Taipei, Taiwan

**Background:** We investigate the incidence & outcomes of arrhythmia before and after transcatheter closure of atrial septal defect (ASD) in patients older than 40 years.

**Patients & Methods:** During an 8.8-year period, 179 patients (median 53 years, 43 males) above 40 years of ages underwent catheter closure of ASD with Amplatzer septal occluders. The presence of significant arrhythmia was assessed using 12-lead ECG or Holter monitor. Patients were follow-up at 1 month, 3 months, 6 months and yearly after the procedure.

**Results:** Significant arrhythmia prior to intervention was documented in 31 patients (17%): 4 atrial flutter, 13 paroxysmal



atrial fibrillation (AF), 9 persistent AF and 5 supraventricular tachycardia. One patient with paroxysmal AF underwent ablation before the procedure. Patients with atrial flutter or AF (total  $n = 26$ ) were significantly older ( $63 \pm 10$  vs.  $53 \pm 10$  years,  $P = 0.048$ ) than those without atrial arrhythmias. Catheter closure with Amplatzer septal occluder was successful in 177 but failed in 2. One out of the 2 failure cases had persistent AF before the procedure and developed migration of the device. Surgery with a concomitant maze procedure was performed in this patient. New onset arrhythmia which occurred within 2 months after the procedure was documented in 22 patients (12%): AF in 18, sick sinus syndrome in 1 and supraventricular tachycardia in 3. Of the 31 patients with arrhythmia prior to intervention, 16 patients were in sinus rhythm after a mean follow-up period of  $3.8 \pm 2.1$  years. In the remaining 15 patients, supraventricular tachycardia was documented in 3, atrial flutter in 1, persistent AF in 9 & paroxysmal AF in 2. Of the 22 patients with new onset arrhythmia, 1 had persistent AF and others were in sinus rhythm at 12 months follow-up. One patient who had bigeminy developed AF 1.5-year later.

**Conclusions:** Arrhythmia was common among unrepaired ASD patients above 40 years of ages. Paroxysmal AF may subside after catheter closure but persistent AF persisted in most. New-onset arrhythmia is usually self-limited. Catheter closure provided protection from developing atrial arrhythmia in patients without history of arrhythmia.

### O7-3

#### Non-fluoroscopic Navigation for Accessory Pathway Ablation in Pediatric and Young Adult Congenital Patients: Impact of Accessory Pathway Location

Papagiannis J. (1), Avramidis D. (1), Kazantzidis S. (1), Kovra S. (1), Alexopoulos C. (2), Kirvassilis G. (2)

Pediatric Cardiology Mitera Children's Hospital (1); Pediatric Cardiac Anesthesia Mitera Children's Hospital (2)

**Introduction:** Because of the concern of long-term effects of ionizing radiation in growing organisms, reduction of fluoroscopy is highly desirable in pediatric patients undergoing catheter ablation. The purpose of this study was to assess the impact of the use of a non-fluoroscopic navigation system based on accessory pathway (AP) location.

**Methods:** We compared the fluoroscopic exposures of two groups of pediatric patients who underwent consecutive catheter ablation procedures for APs. Group A consisted of 70 pts in whom fluoroscopy alone was used and group B consisted of 121 pts in whom a non-fluoroscopic system (NavX™ St Jude Medical) was used to minimize fluoroscopic exposure. One way ANOVA was used to compare fluoroscopy times between groups A and B and also between subgroups defined by AP location (right, left or septal). A  $p$  value of 0.05 was considered significant. Access to the left atrium was achieved by transseptal approach in all but 2 pts in whom a retrograde approach was used.

**Results:** There were 70 pts in group A (0.3 to 33 yrs, mean  $11.7 \pm 4.3$ ) and 121 pts in group B (5 to 35 yrs, mean  $11.1 \pm 3.5$ ). Congenital heart disease was present in 16 pts in group A and 11 pts in group B. Fluoroscopy time was 5.2–135 ( $32.2 \pm 28.4$ ) min in group A vs 0.4–54.3 ( $8.98 \pm 7.8$ ) min in group B ( $P < 0.001$ ). When assessment of fluoroscopic exposures was performed based on AP location, significant reduction was observed regardless of AP location (see table).

**Conclusions:** The use of a non-fluoroscopic system in all pediatric catheter ablation procedures resulted in significant reduction of fluoroscopy exposure regardless of accessory pathway location.

AP location	Group A	Group B	P-value
Right	$n = 7$	$n = 19$	0.024
	$21.62 \pm 18.3$ 4.69–38.56 min	$9.26 \pm 8.15$ 5.33–13.19 min	
Left	$n = 29$	$n = 42$	<0.001
	$29.57 \pm 16.85$ 23.16–35.98 min	$8.75 \pm 5.86$ 6.92–10.57 min	
Septal	$n = 34$	$n = 60$	<0.001
	$36.74 \pm 36.66$ 23.95–49.53 min	$9.06 \pm 8.94$ 6.74–11.37 min	

### O7-4

#### Acute and Long-Term Outcome after Catheter Ablation of Atrial Tachycardia in post-Fontan Patients

Hessling G., Wu J., Pflaumer A., Hoppmann P., Estner H., Jilek C., Deisenhofer I., Hess J.

Deutsches Herzzentrum München, Munich, Germany

**Objectives:** Data about the long-term outcome of atrial tachycardia (AT) ablation in post-Fontan patients (pts) are scarce. We studied our acute and long-term ablation results in these patients and investigated whether it is necessary to ablate all induced AT.

**Methods:** The study included 50 ablation procedures in 36 pts (9 to 46 years, mean age  $25.3 \pm 8.9$ , 13 females). Pts underwent one ( $n = 23$ ), two ( $n = 12$ ) or three ( $n = 1$ ) procedures. Ablation was performed during ongoing AT ("tachycardia group",  $n = 27$ ), or after induced tachycardia ("sinus group",  $n = 23$ ). Acute ablation success was defined as termination of all spontaneous (tachycardia group) or induced (sinus group) AT. Follow-up was available 1, 3, 6, 12 and 24 months after ablation and thereafter yearly.

**Results:** In 27 procedures of the tachycardia group, 46 AT (intraatrial reentry tachycardia [IART]  $n = 40$  and focal atrial tachycardia [FAT]  $n = 6$ ) were mapped and ablated (1.7 AT per procedure). These included 27 forms of ongoing clinical AT and 19 forms of AT arising after termination of the clinical AT. Acute success was reached in 26/27 (96%) procedures. After 7/27 procedures (26%) AT recurred after a median of 15 months follow-up. The mean fluoroscopy time was  $24.9 \pm 17.1$  min and mean procedure duration was  $301.7 \pm 111.2$  min.

In 23 procedures of the sinus group, 39 forms of induced AT were mapped and ablated (IART  $n = 28$  and FAT  $n = 11$ ; 1.7 AT per procedure). Acute success was reached in 19/23 studies (82.6%). After 8/23 procedures (35%) AT recurrence occurred after a median of 34 months. The mean fluoroscopy time was  $26.3 \pm 11.6$  min and mean procedure duration was  $322.7 \pm 119.9$  min.

Looking at AT recurrence during long-time follow-up, ablation of all induced tachycardias was not more advantageous than only ablating ongoing/spontaneous AT ( $P = 0.50$ ).

**Conclusions:** Catheter ablation of AT in post-Fontan patients has a high acute success rate but tachycardia recurrence is quite frequent. Ablation of all induced tachycardias was not more advantageous than only ablating ongoing/spontaneous AT with a trend towards shorter procedure and fluoroscopy time in the latter group with fewer recurrences.

### O7-5

#### LV Pacing Preserves Ventricular Synchrony and Function in Children with AV Block and Structurally Normal Heart as Compared to RV Pacing Sites: A Multi-Center Study

Janousek J. (1), van Geldorp I. E. (2), Prinzen F.W. (3), Vanagt W. (3), Delhaas T. (2), Elders J. (4), Frühl A. (5), Kubus P. (1), Tomaske

M. (6), Ganame J. (7), Nagel B. (8), Papagiannis J. (9), Wren C. (10), Nürnberg J.-H. (11), Gebauer R.A. (12); for the Working Group on Cardiac Dysrhythmias and Electrophysiology of the AEPC Kardiocentrum and Cardiovascular Research Center, University Hospital Motol, Prague, Czech Republic (1); Departments of Pediatric Cardiology and Physiology, Maastricht University Medical Center, Maastricht, Netherlands (2); Department of Physiology, Maastricht University Medical Center, Maastricht, Netherlands (3); Canisius Wilhelmina Hospital, Dept of cardiology, Nijmegen, Netherlands (4); Department of Pediatrics, Oslo University Hospital Rikshospitalet, Oslo, Norway (5); Department of Pediatric Cardiology, University Children's Hospital, Zurich, Switzerland (6); University Hospitals Leuven, Leuven, Belgium (7); Department of Pediatrics, Division of Pediatric Cardiology, Medical University Graz, Graz, Austria (8); Division of Pediatric Cardiology, Mitera Children's Hospital, Maroussi, Greece (9); Pediatric Cardiology, The Newcastle upon Tyne Hospitals – NHS Trust, Newcastle, United Kingdom (10); Abt. für Angeborene Herzfehler/Kinderkardiologie, Klinikum Links der Weser, Bremen, Germany (11); Department of Pediatric Cardiology, University of Leipzig, Heart Center, Leipzig, Germany (12)

**Objectives:** Chronic right ventricular (RV) pacing is often associated with left ventricular (LV) dysfunction. We investigated which pacing site carries least adverse effects on LV synchrony and function.

**Methods:** Cross-sectional data were collected from 10 centres on 73 children (<18 years) with complete AV block and structurally normal heart undergoing chronic permanent pacing (>1 year) from the RV free wall (RVFW, N = 26), RV apex (RVA, N = 34) and LV apex or mid-lateral wall (LV, N = 13) for a median of 5.3 (IQR 3.1–8.5) years. Echocardiographic analysis was performed in a core lab. Differences between pacing sites were evaluated by one-way ANOVA on ranks followed by pair-wise multiple comparison procedures using the Dunn's method. Multivariable analysis was performed by multiple logistic regression.

**Results:** In comparison of the 3 pacing sites LV pacing was associated with the highest biplane LVEF, shortest inter-ventricular mechanical delay (IVMD), shortest septal to posterior wall motion delay (SPWMD) and shortest septal to lateral mechanical delay as measured from circumferential strain (SLMD) (Table, median and IQR). Pacing from the RVFW (but not the presence of maternal anti Ro/La antibodies, pre-implantation LV size and function, age at implantation, duration of pacing, pacing mode and QRS duration) was the only significant multivariable predictor (OR 14.4, CI 2.4–86.3, P = 0.004) of decreased LVEF (<50%).

**Conclusions:** LV pacing carries best long-term preservation of LV mechanical synchrony and systolic function in children with complete AV block and structurally normal heart. RVFW pacing is a significant determinant of decreased LV function. (JJ and PK supported by the research project of University Hospital Motol MZOFNM2005).

Pacing site:	RVFW(1)	RVA (2)	LV (3)	P 1vs2	P 2vs3	P 1vs3
LVEF %	50 (43–56)	55 (51–59)	60 (56–64)	NS	NS	<0.05
IVMD ms	50 (35–61)	39 (20–59)	6 (–8–17)	NS	<0.05	<0.05
SPWMD ms	62 (15–80)	40 (0–75)	0 (–39–0)	NS	<0.05	<0.05
SLMD ms	123 (78–143)	69 (45–110)	–18 (–99–18)	NS	<0.05	<0.05

## O7-6

### Massive septal hypertrophy as a risk factor for malignant cardiac arrhythmias in children with hypertrophic cardiomyopathy

Boon G.B. (1), Bökenkamp R. (1), Blank A.C. (3), Frohn-Mulder I.M.E. (4), Delhaas T. (5), Wolterbeek R. (2), Kuipers I.M. (6), Rammeloo L. (7), Reimer A. (8), Blom N.A. (1,6)

Pediatric Cardiology (1) and Medical Statistics (2) LUMC, Leiden; Pediatric Cardiology UMC, Utrecht (3); Erasmus MC Rotterdam (4); MUMC/AZM, Maastricht (5); AMC, Amsterdam (6); VUMC, Amsterdam (7); UMC St. Radboud, Nijmegen (8); The Netherlands

Hypertrophic cardiomyopathy (HCM) is a common cause of sudden cardiac death (SCD) in children. Non-sustained ventricular tachycardia, syncope, abnormal blood pressure response to exercise, family history of sudden death, and massive left ventricular (LV) hypertrophy > 30 mm are recognised risk factors for SCD in adults with HCM. The predictive value of the adult risk factors for SCD in children is not known.

**Aim:** Investigation of septal thickness and its progression with time as a risk factor for (VT/VF) and SCD in children with HCM.

**Methods:** Multicenter retrospective study in 7 centers in the Netherlands between June 1978 and June 2008. Data from children (0–18 years) with a maximal left ventricular wall thickness of  $\geq 2$  SD above the population mean on echocardiography were included. Measurements were standardized for weight. Statistical analysis was performed by Kaplan-Meier curves and Cox regression.

**Results:** 91 patients were identified with a mean age of diagnosis of 5.9 (+/-5.2) years and mean follow-up of 7.3 (+/-5.2) years. Mean z-score for diastolic interventricular septal thickness (IVSD) at diagnosis was +4.68 SD. Mean increase in z-score was 0.047 SD/year. Sixteen patients suffered from VT/VF at a mean age of 13.0 +/- 4.1 years and 7 of them eventually died. The z-scores for IVSD at diagnosis and at the last contact were significantly associated with VT/VF and SCD, with a Hazard Ratio (HR) of 1.74/SD and 1.73/SD respectively. Massive septal hypertrophy z-score >+7 SD at diagnosis and z-score >+6.5 SD at the last contact had a HR of 8.2 and 9.6 respectively and were therefore suggested as cut-off values.

**Conclusion:** The highest incidence of malignant arrhythmias in children with HCM is between 8–18 years. Septal thickness is a risk factor for VT/VF and SCD in children. Therefore, screening should begin at an early age and prophylactic ICD-implantation might be considered in a subset of patients with massive septal hypertrophy above a z-score of +6.5 SD.

## O7-7

Withdrawn

## O8-1

### Experimental development of a polydioxanone electrospun tissue-engineered valved patch as a new tool for repair of the right ventricular outflow tract in congenital heart diseases.

Kalfa D. (1), Bel A. (2), Chen-Tournoux A. (1), Della Martina A. (3), Rochereau P. (1), Coz C. (1), Bellamy V. (1), Bensalah M. (4), Mousseaux E. (4), Bruneval P. (5), Larghero J. (6), Menasché Ph. (1,2)  
INSERM U633, Laboratoire de Recherches Biochirurgicales, Paris, France (1); Assistance Publique-Hôpitaux de Paris APHP, Hôpital Européen Georges Pompidou HEGP, Department of Cardiovascular Surgery; University Paris Descartes, Paris, France (2); Bioring Inc., Lonay, Switzerland (3); APHP, HEGP, Department of Radiology; University Paris Descartes, Paris, France (4); APHP, Hôpital Saint-Louis, Laboratory of Cell Therapy; University Paris Diderot, Paris, France (5); APHP, HEGP, Department of Pathology; University Paris Descartes; INSERM U 970, Paris, France (6)

**Background:** Many frequent congenital heart diseases require a surgical reconstruction of the RVOT using a valved patch. Currently used devices in clinical practice (such as glutaraldehyde-treated pericardium, cryopreserved homografts, synthetic Gore-Tex® or Dacron® patches) lack regeneration potential and lead to calcifications, stenosis and reoperations with a high morbidity and mortality.

**Objective:** We investigated the function, histological changes and potential of growth and tissue regeneration of polydioxanone (PDO) electrospun bioabsorbable valved patches seeded with mesenchymal stem cells (MSCs) in the RVOT of growing lambs. **Methods:** Autologous blood-derived MSCs were labeled with quantum dots and seeded on PDO electrospun valved patches. Those were implanted into the RVOT of 6 growing lambs (group I) followed up until 8 months. Unseeded PDO valved patches (group II, n = 2) and standard-of-care autologous pericardial patches fitted with a polytetrafluoroethylene (PTFE) monocusp (group III, n = 2) were used as controls. Results were assessed by echocardiography, magnetic resonance imaging (MRI), histology, immunohistochemistry and biochemical assays.

**Results:** Tissue-engineered RVOT were neither stenotic nor aneurismal and displayed a growth potential, with less fibrosis, less calcifications and no thrombus compared with control groups. The PDO scaffold was completely degraded and replaced by a viable, three-layered, endothelialized tissue and an extracellular matrix with elastic fibers similar to that of native tissue. Detection of quantum dots at 1 month suggested that at least some of the cells were derived from the grafted cells.

**Conclusion:** A polydioxanone electrospun tissue-engineered valved transannular patch seems to be a promising device in restoring a living RVOT and could ultimately lead to applications in the treatment of congenital RVOT diseases.

## O8-2

### Development of major aorto-pulmonary collateral arteries in VEGF120 isoform mouse embryos with Tetralogy of Fallot

Rammeloo L.A.J. (1), Wisse L.J. (2), DeRuiter M.C. (2), Gittenberger-de Groot A.C. (2)

Department of Pediatric Cardiology, VU University Medical Center, Amsterdam, The Netherlands (1); Department of Anatomy and Embryology, Leiden University Medical Center, Leiden, The Netherlands (2)

**Introduction:** Comparable to the development of tetralogy of Fallot (TOF) in humans, Vegf120/120 knock-out mouse embryos develop TOF with various degrees of pulmonary stenosis. To investigate the development of the pulmonary stenosis in relation to time and the possible development of major aorto-pulmonary collateral arteries (MAPCA) we studied a Vegf120/120 knock out mouse model at different embryological developmental stages.

**Methods:** To study the development of the right ventricular outflow tract, pulmonary arteries, ductus arteriosus and formation of MAPCA, both Vegf-wildtype as well as Vegf 120 material from day 10.5 until day 19.5 of the day of development was studied. Material including the whole heart and aortic arch was used for analysis.

**Results:** Of the 49 eligible Vegf120 embryos, 36 embryos had ventral displacement of the OFT and a subaortic ventricular septal defect and various degrees of pulmonary stenosis. A time related development in severity of pulmonary stenosis was observed. Absent ductus arteriosus (aDA) was seen in 14. In 3 pulmonary atresia (PA) was found and these embryos had

developed MAPCA and lacked a DA. PA with MAPCA were only seen from day 16.5. In all, the MAPCA arose from both subclavian arteries, running posterior in the thoracic cavity, along with the phrenic nerve. At the side of the hilus the MAPCA connected the pulmonary arteries. No cardiac defects were observed in the wild type embryos (N = 54).

**Conclusions:** A time related development of pulmonary atresia can lead, in case of absent ductus arteriosus, to the development of major aorto-pulmonary collateral arteries as an alternative route for pulmonary perfusion in the Vegf120 knock-out mouse model.

## O8-3

### Right ventricular adaptation to pressure and volume load in mice

Bartelds B. (1), Borgdorff M.A. (1), Smit-van Oosten A. (1), Takens J. (1), Nederhoff M. (2), van Gilst W.H. (3),

de Windt L.J. (4), Berger R.M. (1)

Center for Congenital Heart Disease, University Medical Center Groningen, Groningen, The Netherlands (1); Department of Medical Physiology, University of Utrecht, Utrecht, The Netherlands (2); Cardiovascular Research Centre, GUIDE, Groningen, The Netherlands (3); Department of Medical Physiology, University of Maastricht, The Netherlands (4)

**Introduction:** Right ventricular (RV) adaptation and dysfunction are major determinants of long-term morbidity and mortality in congenital heart disease. However, its molecular mechanisms are yet unknown. To study these mechanisms, molecular characteristics of abnormally loaded RV's have to be coupled to clinically relevant functional characteristics. Therefore we characterized mouse models of pressure versus volume overload of the RV, using clinical relevant outcomes.

**Methods:** Mice were subjected to pulmonary artery banding (PAB) via a left lateral thoracotomy, aorto-caval shunt (Shunt) via a laparotomy, or sham-operated (Control). Four weeks after surgery, mice were functionally evaluated with either cardiac MRI or voluntary cage wheel exercise, after which RV's were harvested.

	Control	PAB	Shunt
Body Weight (g)	23.6 ± 2.6	24.8 ± 1.4	25.3 ± 2.1
Exercise time			
pre-post surgery (min)	-100 ± 64	-374 ± 255*	-150 ± 174
RV weight/body weight	0.86 ± 0.16	1.61 ± 0.25*	1.45 ± 0.27*
MRI derived volumes			
Enddiastolic (µl)	30 ± 5	39 ± 6§	76 ± 22*†
Endsystolic (µl)	10 ± 3	14 ± 3§	33 ± 13*†
Stroke Volume (µl)	20 ± 3	24 ± 5	43 ± 10*†
Output (ml/min)	8.1 ± 1.0	8.9 ± 0.7	20.0 ± 6.2*†
Ejection Fraction (%)	68 ± 4	62 ± 6	58 ± 9*
Gene expression			
beta/alpha MHC	1.0 ± 0.5	14.0 ± 7.7*†	0.5 ± 0.1
MCIP	1.0 ± 0.3	7.6 ± 2.6*†	2.6 ± 0.5

N = 4-6 per group, \*p < 0.05 vs Control, †p < 0.05 vs Shunt, §p < 0.1 vs Control (ANOVA+LSD)

This study was supported by the Netherlands Heart Foundation (NHS2006T038)

**Results:** PAB-mice had reduced exercise capacity (Table) but normal RV output at slightly higher RV volumes at rest (Table), indicating a harbinger of decompensated RV hypertrophy, before failure. In contrast, Shunt-mice had normal exercise capacity and higher RV output at higher RV volumes at rest, indicating a pattern of compensated RV hypertrophy. PAB-mice and Shunt-mice had similar increases in RV hypertrophy as assessed by RV/body weight (Table), but different molecular activation patterns.

Beta/alpha-MHC expression ratio increased in PAB-mice, but not in Shunt-mice. Modulatory calcineurin interacting protein (MCIP), the Calcineurin inhibiting protein, was increased in PAB-mice, but only moderately in Shunt-mice (Table), indicating different molecular patterns of RV adaptation.

*Conclusions:* Pressure or volume load of the RV in the mouse induced similar degrees of hypertrophy, however with different functional and molecular adaptation. These models allow to study the RV specific molecular, morphological and functional response patterns to pressure and volume load.

#### O8-4

##### **Functional role of TASK-1 in the heart—studies in TASK-1 deficient mice**

Donner B.C. (1), Schullenberg M. (1), Geduldig N. (1), Decking U. (2), Zacharowski K. (3), Wisden W. (4), Schmidt K.G. (1)  
(1) University Children's Hospital Düsseldorf, Department of Paediatric Cardiology; (2) Institute for Heart Physiology, Düsseldorf; (3) Clinic for Anaesthesiology, University Frankfurt; (4) Molecular Neuroscience, Imperial College London

*Introduction:* TASK-1, a potassium background channel (K2P channel) is mainly expressed in the heart and the brain. Pharmacological inhibition of TASK-1 in isolated cardiomyocytes results in prolongation of the action potential duration and early afterdepolarizations pointing to a functional role in repolarization. *Methods:* By cardiac phenotyping the TASK-1 deficient mouse (TASK-1  $-/-$ ) we used techniques from molecular/cellular biology (RT-PCR, immunoblot), measured monophasic action potential duration (MAPs) in the Langendorff perfused heart and applied in vivo cardiac diagnostics (echo-cardiography, surface and telemetric ECG for 24h, heart rate variability, defined physical stress (treadmill and swimming)). By transient ligation of the left anterior descending artery ischemia/reperfusion was induced and the infarct size, incidence of premature ventricular beats and heart rate turbulence was studied.

*Results:* Besides TASK-1, TWIK-2 and TREK-1 were the most predominantly expressed K2P channels in the heart. No compensatory up-regulation in TASK-1  $-/-$  mice was seen for eight K2P channels examined. There were no structural or functional abnormalities found by echocardiography. Electrophysiological studies recording monophasic action potentials revealed a significant prolongation of action potential duration at spontaneous rhythm and at atrial pacing at physiological rates. In accordance with these findings, in surface ECGs of TASK-1  $-/-$  mice a significant prolongation of the corrected QT interval was detected (TASK-1  $+/+$ :  $22.76 \pm 0.54$ ,  $n = 7$ , TASK-1  $-/-$ :  $26.33 \pm 0.89$ ,  $n = 11$ ). Telemetric ECG analyses during normal daily activity and during physical exercise like swimming and treadmill challenge did not reveal a higher incidence of arrhythmias in TASK-1  $-/-$  mice compared with their TASK-1  $+/+$  littermates. Studying heart rate variability by using time and frequency domain measurements TASK-1 showed a significant reduced overall heart rate variability and an increase in sympathetic tone. Ischemia/reperfusion injury resulted in a comparable infarct size in TASK-1  $-/-$  mice compared to TASK-1  $+/+$ . Heart rate turbulence was not altered significantly in TASK-1  $-/-$  animals.

*Conclusion:* These results suggest that deficiency of TASK-1 seems to play a functional role in the repolarization of the cardiac action potential and contributes to the maintenance of heart rate variability.

Supported by Grants from the German Society of Pediatric Cardiology and the University Düsseldorf.

#### O8-5

##### **Spontaneous differentiation of embryonic stem cells leads to functionally diversity of cardiac myocytes**

Hannes T. (1), Halbach M. (2), Wolff M. (3), Pfannkuche K. (3), Doss M.X. (3), Sachinidis A. (3), Hescheler J. (3), Brockmeier K. (1), Pillekamp F. (4), Khalil M. (1)

Department of Pediatric Cardiology, University of Cologne, Germany (1); Department III of Internal Medicine, University of Cologne, Germany (2); Institute for Neurophysiology, University of Cologne, Germany (3); Department of Pediatric Cardiology, University of Duesseldorf, Germany (4)

*Introduction:* Pluripotent stem cell-derived cardiomyocytes are thought to be an outstanding *in vitro* model for the early heart development and in drug testing and hold great potential for cellular cardiomyoplasty. They have been shown to resemble different subtypes of cardiomyocytes in regard of their electrophysiological properties. So far, it is not clear to which extent different embryonic stem cell lines are comparable in their quality of differentiation into cardiomyocytes.

*Methods:* Two different transgenic stem cell lines (CGR8/AMPIGX-7, Doss 2007) and D3/aPIG44, Kolossov 2005) were grown on feeder cells in the presence of leukemia inhibiting factor and differentiated by the hanging drop method. In order to obtain alpha-myosin heavy chain positive cardiomyocytes at a high degree of purity, puromycin was added. After 14 days of differentiation, transmembrane potentials were recorded with microelectrodes at 37°C.

*Results:* Without purification, action potential frequency in D3/aPIG44 ( $n = 45$ ) was markedly higher than in CGR8/AMPIGX-7 ( $n = 20$ ) ( $7.7 \pm 0.1$  Hz vs.  $1.7 \pm 0.2$  Hz,  $p < 0.001$ ). The action potential of D3/aPIG44 was markedly shorter (APD20:  $7 \pm 0.1$  Hz vs.  $36 \pm 5$  Hz, APD50:  $14.6 \pm 0.2$  Hz vs.  $80.6 \pm 10.2$  Hz, APD90:  $29 \pm 0.4$  Hz vs.  $188.9 \pm 13.2$  Hz, all  $p < 0.001$ ). Comparison of purified ( $n = 28$ ) and non-purified ( $n = 45$ ) D3/aPIG44 showed a slightly smaller frequency of purified ESC-CM clusters ( $7.2 \pm 0.1$  Hz vs.  $7.7 \pm 0.1$  Hz,  $p < 0.01$ ) and an increase in APD20 ( $7 \pm 0.2$  Hz vs.  $8.4 \pm 0.4$  Hz,  $p < 0.05$ ) and in APD90 ( $20 \pm 0.4$  Hz vs.  $29.6 \pm 0.4$  Hz,  $p < 0.05$ ). In contrast, purified CGR8/AMPIGX-7 ( $n = 20$ ) had a strong decrease of APDs with purification compared to unpurified CGR8/AMPIGX-7 (APD20:  $26.3 \pm 3.1$  Hz vs.  $36 \pm 5$  Hz, APD50:  $50.1 \pm 6.1$  Hz vs.  $80.6 \pm 10.2$  Hz, APD90:  $79.8 \pm 7.3$  Hz vs.  $188.98 \pm 13.2$  Hz, all  $p < 0.05$ ). The frequency did not significantly change.

*Conclusions:* The spontaneous differentiation of D3/aPIG44 and CGR8/AMPIGX-7 leads to electrophysiologically distinct cardiomyocyte phenotypes in regard of beating frequency, APD and effect of purification under control of the alpha-myosin heavy chain-promoter. CGR8/AMPIGX-7 strongly resemble the ventricular phenotype, while D3/aPIG44 share many properties with sino-atrial cells. This questions the comparability and validity of the spontaneous differentiation of embryonic stem cells as a model for cardiac development and drug testing and underlines the necessity of unraveling factors to control cardiac subtype differentiation.

#### O8-6

##### **Mutations in TRPM4 cause autosomal dominant isolated cardiac conduction disease**

Bouvaquet P. (1, 2, 3), Lui H. (1,2), El Zein L. (1,2), Bozio A. (3), Mégarbané A. (4), Blaysat G. (5), Vilain E. (6)

EA 4171, Université de Lyon, F-69008 Lyon, France (1); Laboratoire Cardiogénétique, Hôpitaux de Lyon, Lyon, France (2); Service Cardiologie Pédiatrique, Hôpitaux de Lyon, Lyon, France; Unité de

*Génétique Médicale, Université Saint Joseph, Beyrouth, Liban (4); Service de Cardiologie Pédiatrique, CHU Grenoble, F-38043, Grenoble, France (5); Service de Cardiologie, Hôpital Necker-Enfants Malades, Paris, France (6)*

**Introduction:** Isolated cardiac conduction block is a relatively common condition in the young and the elderly populations. Genetic predisposing factors have long been suspected because of numerous familial case reports. Deciphering genetic predisposing factors of conduction blocks may give a hint at stratifying conduction block carriers in a more efficient way.

**Methods and Results:** One Lebanese family (L1) and 2 French families (F1 and F2) with autosomal dominant isolated cardiac conduction blocks were used for linkage analysis. In these 3 families, affected individuals had right bundle branch block, left anterior or posterior hemiblock, AV block either alone or in combination but no left bundle branch blocks. A maximum combined multipoint lod score of 10.5 was obtained on a genomic interval including more than 300 genes. After screening 12 genes of this interval for mutation, we found a heterozygous missense mutation of the TRPM4 gene in each family: p.Arg164Trp (F1), p.Ala432Thr (L1) and p.Gly844Asp (F2). These missense mutations change a conserved residue and were not observed in more than 300 chromosomal controls. This gene encodes the TRPM4 channel, a calcium-activated non-selective cation (CAN) channel of the transient receptor potential melastatin (TRPM) ion channel family. Furthermore, we showed by immunohistochemistry that wild-type TRPM4 channel signal level is higher in bovine atrial cardiomyocytes than in common ventricular cells but is highest in Purkinje fibers. Small bundles of highly TRPM4 positive cells were found in the subendocardium and in rare intramural bundles.

**Conclusion:** The TRPM4 gene is a causative gene in isolated cardiac conduction disease and TRPM4 channel is highly expressed in cardiac Purkinje fibers.

## O8-7

### Ductal Stenting: Histopathology and Immunohistochemistry in 24 specimen

*M. Sigler (1), R. Foth (1), T. Quentin (1), M. Schneider (2), T. Kriebel (1), T. Paul (1), I. Michel-Behnke (3)*

*Department of Paediatric Cardiology and Intensive Care Medicine, Goettingen University, Germany (1); German Paediatric Heart Center, St. Augustin, Germany (2); Paediatric Cardiology, University of Vienna, Austria (3)*

**Introduction:** Safety and efficacy of stenting of the arterial duct is well described. In contrast, there is only little information on tissue reactions to these implants. We analysed a series of 24 human explanted ductus stents (bare metal).

**Methods:** Explanted stent specimen were processed using a uniform protocol after surgical removal from pediatric patients with congenital heart disease. Age at implantation was 2 to 28 days; time interval between implantation and explantation was 5 days to 14 months. Seven stents had been redilated prior to explantation. The specimen were fixed in formalin and embedded in methylmethacrylate. We performed standard stainings as well as immunohistochemical analysis of tissue reactions.

**Results:** On gross examination, mild to moderate proliferation of pseudointima was seen within the lumen of all ductus stents. Superficial cells stained positive for von Willebrand factor (vWF) thus demonstrating complete endothelialisation. Neo-tissue formed within the stents contained smooth muscular cells and connective tissue as demonstrated by immunohistochemistry. Very few inflammatory cells were detected in local relation the stent struts.

**Conclusion:** Histopathological work-up of 24 ductus stents revealed complete endothelialisation and only very little inflammatory reactions. A varying degree of tissue proliferation was seen within the lumen of the stents with a pattern of cells and connective tissue closely resembling intimal proliferation as described for stented coronary artery segments.

## O9-1

### Fetal assessment of borderline small left ventricles: Can we predict those with a need for a postnatal intervention?

*Weber R. (1), Ayala-Arnez R. (2), Atiyah M. (2), Manlhiot C. (2), McCrindle B.W. (2), Hickey E.J. (2), Jaeggi E.T. (2), Nield L.E. (2) University Childrens Hospital, Zurich, Switzerland (1); The Hospital for Sick Children, Toronto, Canada (2)*

**Background:** Many forms of congenital heart disease (CHD) can be associated with borderline small left ventricles (bLV). Fetal predictors of a need for an intervention are scant. We sought to prospectively determine fetal echocardiographic predictors of fetal and neonatal outcome.

**Methods:** Fetuses were included who had any form of CHD with normally related arteries and a left ventricle (LV) that was <2 standard deviations below normal for length or diameter and had forward flow across the mitral valve (MV) and aortic valve (AoV). Factors associated with a need for early (<30 days) intervention and no intervention were sought from fetal and postnatal echocardiograms using univariate and multivariate logistic regression models.

**Results:** From 2005–2008, 46 fetuses with bLV had a fetal diagnosis (+fetal coarctation/+transverse arch hypoplasia): atrioventricular septal defect 7(+2/+0), double outlet right ventricle (RV) 2(+0/+0), Shone's complex 19(+9/+4), RV/LV disproportion 18(+13/+11; 4 both, 2 none). Mean gestational age was 25+6 weeks at 1st fetal echo. There were 7 pregnancy terminations, 3 fetal demises/stillbirth and 4 compassionate care. N = 32 livebirths either had a biventricular repair (n = 20, n = 2 dead), univentricular palliation (n = 2, both alive) or no intervention (n = 8, 25%). Overall survival of livebirths at 6 months of age was 79%. Factors associated with an early need for intervention on 1st fetal echo were: obstructed or retrograde arch flow (p = 0.08, odds ratio (OR) 3.3), Shone's diagnosis (p = 0.02, OR 4.9), coarctation (p = 0.05 OR 11.4), presence of LV outflow obstruction (p = 0.05, OR 12.5). Factors associated with no intervention were a larger, though still hypoplastic, mitral valve z-score (mean -3.8 vs. -4.2 intervention group, p = 0.04, OR 2.8) and lower transverse arch to left common carotid artery ratio (mean 1.32 vs. 1.89 intervention group, p = 0.05, OR 0.2).

**Discussion:** Although fetuses with bLV form a heterogeneous group of lesions, postnatal need for early intervention can be predicted by fetal echocardiography. A relatively high proportion of neonates will require no intervention and have a benign outcome.

## O9-2

### In-utero intervention for fetal critical aortic stenosis: indications, complications, results and outcome in 23 consecutive patients

*Tulzer G., Arzt W., Veit I., Wertaschnigg D., Gitter R., Mair R. Children's Heart Centre Linz, Austria*

Stenotic aortic valves can be treated during prenatal life by balloon valvuloplasty with the goal of preventing intrauterine death (IUD), reversing heart failure or avoiding univentricular postnatal circulation. The purpose of this study is to review indications, complications, results and outcome in 23 consecutive fetuses with critical aortic stenosis. From 12/2001 to 12/2009 24

aortic balloon valvuloplasties were attempted in 23 fetuses. Median GA: 27,6 weeks (21,6–33,1). All fetuses had retrograde aortic arch flow, left to right foramen ovale shunt and all but 1 severe EFE. Mitral regurgitation was present in 15, mitral stenosis in 9 fetuses. Four patients presented with hydrops. Technical success was achieved in 16 (69,6%) patients (with 1 IUD, 4 still in-utero). From 11 successfully treated live born patients, 8 (72,7%) achieved a biventricular circulation. postnatally. Out of 7 fetuses with technical failure there were 2 IUD, 1 still in-utero and all 4 liveborn patients went on to univentricular palliation. Reasons for failure were unfavourable fetal position and smaller LVs. Bradycardia necessitating epinephrine administration was observed in 9/24 procedures, LV-thrombus (resolved spontaneously) occurred in 5, pericardial effusion > 3 mm in 3, aortic regurgitation in 11 cases. All fetuses with hydrops had successful procedures; time to resolution of hydrops was 18–34 days. Biventricular circulation was achieved in 3 patients, 1 still in-utero. Up to now 15 patients were live born at a median GA of 36+6 (32+6–39+0), follow-up is median 13,2 months (1,8–59). All of the 11 biventricular patients had an aortic balloon dilation in the 1st week of life, 4 patients needed a Ross-Konno procedure between day 7–21d, 1 patient had a coarctation repair and died at 3 months of sepsis.

In selected fetuses with critical aortic stenosis successful in-utero valvuloplasty may prevent a univentricular circulation after birth in more than 70%. Technical success was dependent on patient selection, fetal position and a dedicated team. Complications were frequent but could be managed in the majority of patients. To achieve a biventricular circulation after birth, aortic valve interventions were necessary in all patients with early aortic valve replacement in half of them.

### O9-3

#### **Patient selection for neonatal aortic balloon dilation. Who is suited for biventricular circulation?**

Georgiev S. (1), Eicken A. (2), Balling G. (2), Hess J. (2)  
Department of Pediatric Cardiology, National Heart Hospital – Sofia, Bulgaria (1); Department of Paediatric Cardiology and Congenital Heart Disease, German Heart Center Munich, Germany (2)

**Introduction:** Neonatal valvular aortic stenosis (AoS) represents a spectrum of different degrees of hypoplasia and malformation of all left heart structures. The decision for a biventricular (balloon dilatation or surgical valvotomy) or univentricular pathway (Norwood operation) needs to be done within the first days of life. There are numerous attempts to create algorithms for prediction of successful biventricular circulation in neonatal AoS.

**Methods:** Firstly, an analysis regarding the mortality and morbidity of all patients who underwent balloon aortic valvuloplasty during a ten year period in a single tertiary institution was made. Secondly, the echocardiograms of all patients with patent aortic and mitral valves treated by balloon aortic valvuloplasty or the Norwood operation between Jan. 2006 and Sept. 2008 were reviewed. The Rhodes - and Colan scores, and the univentricular repair survival advantage tool (UVR-SA) scores were obtained. The results were compared to the actual correction type and outcome.

**Results:** During the last 10 years 54 newborns with AoS were treated by initial balloon dilatation. Median follow up was 2.8 years (range 28 days to 10.11 years). Early mortality was 2% and overall mortality – 13%. Up to now, thirty (56%) patients needed no further interventions. The risk scores were applied retrospectively to 28 patients. Nineteen of them were treated by an initial AoVP and nine by an initial Norwood operation. In three a secondary Norwood operation was done. Out of the 24 patients

predicted to be univentricular candidates by the Rhodes score, 12 (50%) live now with biventricular circulation. The Colan score predicted 19 univentricular repairs and 7 (37%) of these patients now live with biventricular circulation. The UVR-SA tool resulted in 12 univentricular and 16 biventricular decisions. Two (17%) of the supposed univentricular patients are now alive with a biventricular circulation and 2 (12%) of the supposed biventricular patients have undergone a Norwood operation.

**Conclusion:** Neonatal AoS can successfully be treated by initial balloon valvuloplasty after proper patient selection. Many patients require additional interventions later in life. Retrospective analysis of the application of the current risk scores did not reliably predict outcome. Treatment decisions are based on local experience and expertise.

### O9-4

#### **Percutaneous pulmonary valve implantation – Initial two center experience**

Eicken A. (1), Ewert P. (2), Hager A. (1), Peters B. (2), Fratz S. (1), Kuehne T. (2), Hess J. (1), Berger F. (2)

Klinik für Kinderkardiologie und angeboren Herzfehler, Deutsches Herzzentrum München, Technische Universität München, Germany (1); Deutsches Herzzentrum Berlin, Abteilung für angeborene Herzfehler/Kinderkardiologie, Germany (2)

**Objective:** Dysfunction of valved conduits in the right ventricular outflow tract (RVOT) limits durability and enforces repeated surgical interventions. We report on our combined initial two center experience with percutaneous pulmonary valve implantation (PPVI).

**Methods and Results:** 80 pts with RVOT conduit dysfunction and two pts with a stenosed RA-RV homograft after a modified Fontan operation (median weight 63 kg, range 28–99 kg, median age 22.9 yrs, range 9.1–69.6 yrs, diagnoses: TOF/PA 42, TAC 13, TGA 8, other 19) were scheduled for PPVI since December 2006. PPVI was successfully performed in all patients (pre stenting 77/82 pts). The median systolic RVOT gradient decreased from 36 mmHg, range 5–91 mmHg to 14 mmHg, range 0–73 mmHg ( $p < 0.001$ ) and the ratio RVP/AoP decreased from 61%, range 30–132% to 37%, range 20–80% ( $p < 0.0001$ ). The median enddiastolic RV-volume index (MRI) decreased from 106 ml/m<sup>2</sup>, range 56–227 ml/m<sup>2</sup> to 89 ml/m<sup>2</sup> ( $p < 0.0001$ ). Pulmonary regurgitation was significantly reduced in all pts. One patient died due to coronary artery compression. There was one transient and one permanent AV-block. During follow-up (median 189 days, range 1–763 days) one Melody valve had to be removed surgically 6 months after implantation due to bacterial endocarditis. In 7/82 pts a repeated dilatation of the valve was done due to a significant residual systolic pressure gradient, which resulted in a valve-in-valve procedure in one.

**Conclusions:** This study shows that PPVI is safe and it effectively improves the hemodynamics in a selected patient collective. The rate of complications at short term follow-up is low. However, the intervention is technically challenging. Longer clinical follow-up is needed.

### O9-5

#### **Stenting of aortic coarctation: Acute, intermediate, and long-term results of a prospective multi-institutional registry – CONGENITAL CARDIOVASCULAR INTERVENTIONAL STUDY CONSORTIUM (CCISC)**

Holzer R.J. (1), Qureshi S. (2), Vincent J. (3), Ghasemi A. (4), Sievert H. (5), Alday L.E. (6), Peirone A. (7), Gruenstein D.H. (8),

Forbes T. (9), Weber H.S. (10), Zeller T. (11), Cheatham J.P. (1); Slack M. (12), Jarvois A.J. (13), Hoyer M.H. (14), Rome J. (15) Nationwide Children's Hospital, Columbus, USA (1); Evelina Children's Hospital, London, United Kingdom (2); Morgan Stanley Children's Hospital, New York, USA (3); Beheshti University of medical Science, Tehran, Iran (4); CardioVascular Center, Frankfurt, Germany (5); Senatorio Allende, Cordoba, Argentina (6); Private Hospital of Cordoba, Argentina (7); University of Minnesota Children's Hospital, Minneapolis, USA (8); Children's Hospital of Michigan, Detroit, USA (9); Hershey Medical Center, USA (10); Children's Medical Center, Dallas, USA (11); Children's National Medical Center, Washington, USA (12), Hope Children's Hospital, Oak Lawn, USA (13); Indiana School of Medicine, Indianapolis, USA (14), Children's Hospital of Philadelphia, USA (15);

**Introduction:** Since the 1980s, stent implantation has evolved as an important therapeutic strategy for coarctation. However, the available data is frequently flawed by short follow up, lack of adequate follow-up imaging, and retrospective nature of data collection.

**Methods:** Data was prospectively collected using a multi-center registry (CCISC). Between 2000–09, 302 patients from 34 centers with a median weight of 58 kg (11–156 kg) and a median age of 15 years (2–63 years) underwent stent implantation for coarctation. 44% of eligible patients completed intermediate follow-up (3–18 months) with integrated imaging (cath, CT, MRI), while 21% completed long-term follow-up (>18–60 months). Procedural success was defined as UL/LL systolic gradient of less than 20 mmHg, lack of significant reobstruction, and freedom from unplanned reintervention.

**Results:** Acute procedural success was 96%. Cumulative intermediate success was 86% and cumulative long-term procedural success was 77%. There was no difference in acute, intermediate and long-term success, when comparing immediate invasive post-stent gradients of <10 mmHg, <15 mmHg, and <20 mmHg. 4% of patients required unplanned reinterventions, while 9% had planned/staged reinterventions. Aortic wall complications were seen in 3% patients (dissection n = 7, aneurysm n = 3), 2 of which were seen acutely, 6 at intermediate follow up and 2 at long-term follow-up. Other adverse events (n = 15) occurred mainly acutely and included technical complications such as stent malposition (n = 9). At long-term follow up 23% of patient continued to have a systolic blood pressure above the 95th centile, 9% had an upper-to lower limb blood pressure gradient in excess of 20 mmHg, and 32% were taking antihypertensive medication.

**Conclusion:** This study documented acute, intermediate and long-term outcome data comparable or superior to other surgical or interventional series. However, even with successful initial stent therapy, coarctation continues to require long-term follow up and has associated long-term morbidity, relating to aortic wall complications, systemic hypertension, recurrent obstruction as well as need for reintervention. Operators should aim for an immediate invasive gradient reduction to <20 mmHg.

#### O9-6

##### **Stenting the arterial duct in neonates and infants with congenital heart disease and duct-dependent pulmonary blood flow: a multicentre experience of 65 patients.**

Udink ten Cate F.E.A. (1), Hamza H. (2), Agha H. (2), Kohli V. (3), Emmel M. (1), Qureshi S.A. (4), Sreeram N. (1), Rosenthal E. (4) Departments of Pediatric Cardiology of the Heart Center Cologne, University Hospital Cologne, Cologne, Germany (1); Cairo University Children's Hospital, Cairo, Egypt (2); Pediatric Cardiology and

Congenital Cardiac Surgery Unit, Indraprastha Apollo Hospital, New Delhi, India (3); Department of Congenital Heart Disease, Evelina Children's Hospital, London, United Kingdom (4)

**Objectives:** Ductal stenting in neonates and infants with duct-dependent pulmonary blood flow potentially avoids the need for a surgical systemic-to-pulmonary shunt. Our purpose was to describe a 18-year multicenter experience with neonates and infants undergoing ductal stenting for duct-dependent pulmonary circulation.

**Methods:** Between 1991 and 2009, 65 neonates and infants (39 male, 60%) underwent cardiac catheterization for ductal stenting in 4 participating centres. Median age and weight was 3.2 kg (2.4–6.0 kg) and 9 days (2–249 days). Diagnoses included pulmonary atresia with intact ventricular septum (PA-IVS), n = 33 (51%), critical pulmonary stenosis (cPS) n = 10 (15.5%), PA with ventricular septum defect (PA-VSD) n = 5 (7.5%), tetralogy of fallot (TOF) n = 3 (4.5%), and single ventricle physiology n = 14 (21.5%).

**Results:** Ductal stenting was successful in 52/65 (80%) patients. The stent diameter ranged from 3.0–6.0 mm with varying lengths. Primary vascular access was obtained using the femoral (n = 57, 88%) or axillary artery (n = 8, 12%). The ductal morphology was horizontal and tubular (n = 38, 58%), tortuous (n = 22, 34%), and vertical (n = 5, 8%). Stenting failed in 11/22 patients with a tortuous duct, and in 2/38 (5.2%) patients with tubular ductal morphology. Incomplete stenting occurred in 3 patients. There were 12 procedure related complications: femoral artery obstruction (n = 7), necrotising enterocolitis (n = 2), right pulmonary artery stenosis (n = 2), pseudoaneurysm (n = 1). Redilation of the stent was required in 4 patients. The duration of hospital stay ranged from 2 to 97 days. No procedure related deaths occurred.

**Conclusion:** Percutaneous ductal stenting in the current era has become a feasible and safe therapy in neonates and infants with ductal-dependent pulmonary blood flow. Successful stent implantation is related to ductal morphology. Atypical ducts may warrant alternative vascular approaches.

#### O9-7

##### **Mid-term follow-up of treatment of middle aortic syndrome in children**

Brzezińska-Rajszyz G., Zubrzycka M. (1), Rewers B. (1), Książek J. (1), Antoniewicz J. (2), Tirska-Kmieć A. (3), Kościeszka A. (4), Ostoja-Chyżyńska A. (5)

The Children's Memorial Health Institute, Warsaw, Poland Heart Catheterization Laboratory(1); Department of Nephrology, Kidney Transplantation and Hypertension(2); Department of Cardiology (3); Computed Tomography (4); Department of Surgery and Transplantology (5)

**Background:** Middle aortic syndrome (MAS) is an uncommon cause of arterial hypertension in children and young adults. Interventional therapy of MAS has been reported in the literature.

**Aims:** The aim of the study was to analyse the results of complex interventional and surgical therapy in patients with MAS performed in single center.

**Material/Methods:** Twenty children (aged 3–17 yrs, mean 11.2 yrs), with severe arterial hypertension (AH) resistant to multidrug therapy and with a diagnosis of MAS underwent complex interventional and surgical treatment. There were no inflammatory signs or symptoms suggesting Takayasu disease in the group. 20pts had narrowing of thoracic and/or abdominal

aorta (length 2–9 cm, minimum diameter 1.5–5 mm). Aortic narrowing was isolated in 10pts (groupA) and coexisted with stenosis of abdominal branches in 10pts (groupB). In groupA 17 and in groupB 10 stents were implanted in the aorta. Additional interventional (balloon angioplasties of renal, mesenteric and celiac arteries, stent implantation of truncus coeliacus) and surgical procedures (renal artery autotransplantation) were performed in groupB.

**Results:** In groupA, mean systolic gradient in aorta decreased from  $45 \pm 6.3$  mmHg before to  $15.2 \pm 7.4$  mmHg after stent implantation. During mean  $7.81 \pm 5.04$  yrs (range 0.6–14 yrs) follow-up additional procedures included elective stent redilation (3pts), second stent implantation (small in-stent aneurysm)(2pts), balloon angioplasty (neointimal hyperplasia)(4pts). Two pts did not need antihypertensive treatment, whilst all the others have better control of AH with lower doses of medications.

In groupB as a result of complex primary therapy AH was better controlled with lower doses of medications. During a mean follow-up of  $5.72 \pm 2.64$  yrs, additional procedures – balloon angioplasty for stent redilation (3pts), neointimal hyperplasia (3pts), additional stent implantation to aorta due to narrowing progression (1pt) and small aneurysm formation above the stent (1pt), balloon renal artery angioplasty (1pt), redilation of stent in truncus coeliacus (1pt) were performed. AH was better controlled on the lower doses of medications.

**Conclusions:**

1. MAS can be treated successfully with stent implantation.
2. Due to complexity of the disease, combined interventional and surgical treatment may be necessary resulting in better control of arterial hypertension.
3. Careful follow-up of patients with MAS allows for recognition of indications for additional interventions.

#### O10-1

##### **Subcutaneous treprostinil in pediatric pulmonary arterial hypertension.**

*Lévy M., Bajolle E., Szezepanski I., Bonnet D.  
Centre de Référence Malformations Cardiaques Congénitales Complexes – M3C, AP-HP, Université Paris Descartes, Paris, France*

**Background:** Therapeutic options at time of worsening remain unclear in children who do worse but remain in functional class III while being on combined oral therapy with endothelin receptor antagonists and PDE-5 inhibitors.

**Objective:** To evaluate the use of a subcutaneous prostanoid as an add-on therapy in pediatric PAH.

**Patients and methods:** Nine children with PAH (range 1.5–17 y, median 5 y) received subcutaneous treprostinil at time of worsening. Five had idiopathic PAH, one being familial. Four patients had PAH associated with but not caused by congenital heart defect. All had severe PAH and were non responsive to inhaled nitric oxide. They were all on combined oral therapy with bosentan and sildenafil. Indication to add treprostinil was clinical deterioration with right ventricular failure in 8 patients and switch from IV epoprostenol in one. Treprostinil was initiated at a fixed dose of 1.25 ng/kg/min and then daily increased by 1.25 ng/kg/min to reach an average dose of 20 ng/kg/min.

**Results:** Local tolerance was good with tailored management of site injection. One child died of acute right ventricular failure after one month. Another 17 years old girl died 3 weeks after a Potts' shunt. In the remaining 7 pts, No serious adverse events was noted during the median 6 months follow-up. All patients were clinically improved with significant increase in 6MWT

distance in the 3 oldest children and change from WHO functional class III to class II. Echocardiography showed an improved right ventricular function, increased pulmonary flow with increased acceleration time.

**Conclusion:** Subcutaneous treprostinil can be a valuable alternative in pediatric PAH patients already receiving combined oral therapy. It may help to avoid complications of central lines and to improve quality of life.

#### O10-2

##### **Heart Transplantation in GUCH Population**

*Vignati G., Ribera E., Bruschi G., Gagliardone M.P., Montorsi E., Aresta F., Pains R., Marianeschi S.M., Corato A., Frigerio M., Iorio F.S., Martinelli L., Santoro F.*

*CardioThoracic Department – Niguarda Hospital – Milan (Italy)*

**Introduction:** Heart transplantation (HTx) is a recognised treatment for end stage heart failure (HF) of any cause, including congenital heart disease (CHD). CHD patients HTx-ed to date is small. However, advances in paediatric surgery has increased the demand for HTx for grown-up with congenital heart disease (GUCH) patients.

**Methods:** From October 1985 to December 2009, 870 HTx were performed in our Centre; 29 (3.6%) were performed for GUCH patients. Mean age at HTx was  $31.7 \pm 13.8$  yy; 13 patients (45%) had been previously-operated once, five (17%), twice and three pts more than three-time. Eight pts (27,5%) were single ventricle anatomy previously palliated; just one patient underwent Fontan procedure. The underlying diagnoses of other patients were repaired or uncorrected defects with late ventricular dysfunction. The indication for HTx was based on deterioration of quality of life and advances HF. Severe pulmonary hypertension was a contraindication for HTx.

**Results:** In three cases (10,3%) femoral artery cannulation was needed. Fifteen pts (51,7%) required additional surgery at HTx. Two patients (6,8%) died early after HTx for hepatic failure and multy organ failure (Fontan patient). Median ventilation time was 20 hours, median ICU stay was 6 days. Mean follow-up (FU) time of discharged patients was  $73 \pm 55$  months. Three patients (11.%) died at a mean time of seven years after HTx for coronary allograft vasculopathy, late graft failure and rejection. One, five and ten years survival was 93,2%, 93,2% and 82,8% respectively.

**Conclusions:** The total number of HTx in GUCH pts is still small. CHD is identified as one of the strongest risk factor for 1 year-mortality after HTx in adults. In contrast, in those pts who survive 3 years, CHD has a marked 10 years survival advantage independently of age. In our experience, successful HTx in GUCH patients can be perform with good short and long-term results, comparable with those of non-CHD patients. Previous palliation or pulmonary artery reconstruction do not affect outcome after HTx. Patients with Fontan circulation still remains controversial.

#### O10-3

##### **Transplant coronary artery disease is diagnosed later and is more severe in pediatric heart recipients transplanted during infancy**

*Tissot C., Pietra B.A., Ivy D.D., Campbell D.N., Mitchell M.B., Miyamoto S.D.*

*The Children's Hospital, Denver, Colorado, USA*

**Introduction:** Transplant coronary artery disease (TCAD) represents the largest cause of late graft loss and the most frequent indication for retransplantation in pediatric recipients. Intravascular

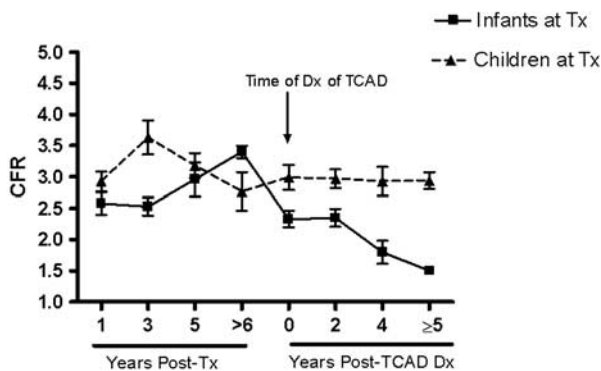


ultrasound (IVUS) and coronary flow reserve (CFR) allow early detection and follow-up of TCAD.

**Methods:** We retrospectively reviewed pediatric heart recipients with TCAD. TCAD was diagnosed by angiography or IVUS. Patients transplanted at <6 months of age (infants) were compared to those transplanted at >6 months (children). Stable TCAD was defined as no change in severity after serial evaluation and progressive TCAD as worsening disease or death as a result of TCAD.

**Results:** Of 310 transplanted children, 52 (17%) were diagnosed with TCAD. Of those, 20 (38%) were infants and 32 (62%) were children at the time of transplantation. Age at TCAD diagnosis ( $8.3 \pm 3.4$  vs  $14 \pm 5.8$ ,  $p < 0.001$ ) and the number of prior rejection episodes ( $3.3 \pm 1.7$  vs  $5.8 \pm 3.6$ ,  $p < 0.01$ ) were significantly lower in the infant group. Nevertheless, infants with TCAD had significantly more episodes of rejection compared to those without TCAD ( $0.83 \pm 1.14$  vs  $3.35 \pm 1.73$ ,  $p < 0.0001$ ). More patients in the children group had stable TCAD (76% vs 55%,  $p < 0.01$ ), of those 9 (28%) were diagnosed by IVUS, compared to none in the infant group. Compared to children, infants had a longer time from transplantation to diagnosis of TCAD ( $8.0 \pm 3.4$  vs  $5.4 \pm 3$  years,  $p < 0.01$ ) but their angiographic score was higher at diagnosis ( $3 \pm 1$  vs  $1.7 \pm 1.3$ ,  $p < 0.01$ ). Coronary flow reserve in the infant group was significantly lower ( $2.3 \pm 0.5$  vs  $3 \pm 0.3$ ,  $p < 0.05$ ) at the time of TCAD diagnosis and showed a progressive decline over time compared to the children group (CFR 2 years post-TCAD diagnosis  $2.1 \pm 0.6$  vs  $2.9 \pm 0.7$ ,  $p < 0.01$ ). There was a trend towards shorter time from diagnosis of TCAD to death or retransplantation in the infant group ( $0.9 \pm 0.9$  vs  $2.8 \pm 3.4$  years,  $p = 0.06$ ).

**Conclusion:** TCAD in infant recipients is diagnosed later, appears worse by angiography and is associated with a lower CFR at the time of diagnosis which tends to decline over time. Despite fewer rejections in infant recipients with TCAD, there is a trend towards shorter time to death or retransplantation, reflecting a more aggressive nature of TCAD.



#### O10-4

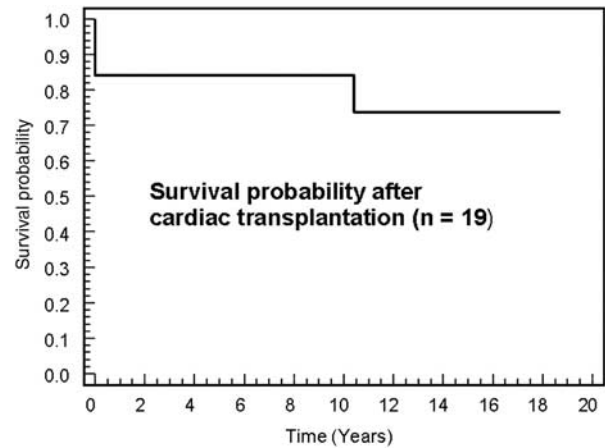
##### Survival and follow-up after cardiac transplantation for congenital heart diseases.

Guillaume M.P., Timmermans T., Van Caeneghem O., Poncelet A., Gurné O., Carbonez K., Sluysmans T.  
Cliniques Universitaires Saint-Luc, UCL, Brussels, Belgium

**Introduction:** The number of patients with congenital heart disorder requiring a cardiac transplant is limited. We report our experience concerning survival and complications of congenital patients after cardiac transplantation in our hospital.

**Method:** From 1985 to January 2010, 19 patients (9 children and 10 adults) were transplanted (7 single ventricle, 5 left heart valvular

disease (4 aortic and 1 mitral), 2 D-TGA after Senning or Mustard, 1 congenitally corrected transposed great arteries, 2 Fallot, 1 truncus, 1 complete AV canal. Median age at cardiac transplant was 20.8 years (min 7 years, max 64 year) after a medium delay on the waiting list of 135 days (min 7 days – max 3 years).



**Results:** Four patients died respectively after 1, 2, 6 days and 10 years post transplant. The rate of survival was 84% at 10 years and 74% at 20 years of follow-up. 1985–2010 – Survival Probability of Congenital. Patients after cardiac transplantation (n = 19)

Complications were acute rejection (n = 5) with 3 deaths at day 1, day 6 and 10 years post transplant, chronic renal insufficiency (14/15) with necessity of dialysis in 3 patients, infections (14/15; septicemia n = 4, organ infection n = 6, local infection = 7), hypertension (11/15), arrhythmia (2/15, pacemaker 1/15), cerebral vascular accident (2/15), hyperlipidemia (14/15), dysthyroidia (5/15), diabetes (1/15), hyperuricemia (1/15), osteoporosis (3/15).

**Conclusion:** Survival and quality of life after cardiac transplant for congenital heart disease are globally good. Complications are nevertheless frequent, particularly renal insufficiency.

#### O10-5

##### More than ten years after heart transplantation in children

Di Filippo S. (1), Ducreux C. (1), Henaine R. (2), Boissonnat P. (3), Sebbag L. (3), Roussoulières A. (3), Gouton M. (1), Ninet J. (2), Sassolas F. (1), Bozio A. (1)

Pediatric Cardiology, Cardiovascular Hospital Louis Pradel, Lyon, France (1); Cardiac Surgery, Cardiovascular Hospital Louis Pradel, Lyon, France (2); Department of Heart Transplantation, Cardiovascular Hospital Louis Pradel, Lyon, France (3)

Mid-term outcomes of pediatric transplanted patients are reported to be favourable, but very long-term graft and patient survival is still a matter of debate. The aim of this study was to assess status and long-term outcome of children who completed at least 10 years follow-up after heart transplantation (HTx).

**Material and methods:** The records of patients <18 years old at HTx, who survived at least 10 years posttransplant, were reviewed for clinical, biological, echocardiographic and angiographic data.

**Results:** Since 1987, 91 patients less than 18 years of age, received a HTx. Among them, 27 (14 males) survived at least 10 years after transplant; the others either died (22) or had less than 10-year follow-up (42). Indication for HTx was congenital heart disease in 10 and cardiomyopathy in 17. Age at HTx was  $10.3 \pm 6.3$  years (2 days to 17.8 years) : 8 were aged <5 years, 5 were 5 to 10 years old and 14 were 10 to 18 years old. Follow-up was  $16.2 \pm 4.1$  years (10 to 25.3). Four patients died at 11, 12 and 14 years posttransplant,

none from coronary artery disease. One underwent second transplantation at 16.6 years after first transplant. Graft coronary artery disease occurred in 7 cases (26%): severe in only 3 cases (11%), moderate in 1 and mild in 3. Renal function was normal in 16 cases (59.2%), severely impaired in 5 patients (2 on peritoneal dialysis). Four patients had moderate and 2 mild renal dysfunction. Chronic renal failure was more frequent in patients aged <5 years at HTx (62.5% vs 0%,  $p = 0.001$ ). Coronary artery disease occurred in 50% of patients aged <5 years at HTx, 16% of others ( $p = 0.06$ ), whereas length of follow-up was similar between both groups. One patient experienced posttransplant lymphoproliferative disease at 14 years of follow-up.

**Conclusion:** More than 10 year- follow-up results of pediatric heart transplantation are favourable. Patients less than 5 years of age at the time of transplantation are more likely to develop graft coronary artery disease and renal impairment, and should be the most intensively monitored.

#### O10-6

##### Specific vasodilator therapy in children with severe pulmonary hypertension

Alehan D., Şahin M., Yıldırım I., Karagöz T., Özer S., Özkutlu S. Hacettepe University, Department of Pediatrics, Section of Pediatric Cardiology, Ankara, Turkey

**Introduction:** Pulmonary hypertension is a fatal disease with few therapy options and limited experience in children. Children with severe pulmonary hypertension receiving specific vasodilator treatment are presented.

**Methods:** 47 patients with severe pulmonary hypertension received specific vasodilator therapy in addition to conventional therapy between September 2003–December 2009. A detailed history, physical examination with electrocardiogram and echocardiographic evaluation, heart catheterization, vasoreactivity test and six minute walking test were performed before initiation of therapy; these tests, except heart catheterization, were repeated every 3 months during follow-up. Complete blood count, liver function tests, brain natriuretic peptide level and measurement of oxygen saturation were done and recorded during each visit. Patients were divided into two groups: those with idiopathic pulmonary hypertension (IPAH) (n:10) and those with associated pulmonary hypertension (APAH) (n:37) due to left-to-right shunted congenital heart defects.

**Results:** Initial therapy was bosentan in 34 patients and inhaled iloprost in 12 patients, sildenafil and iloprost in 1 patient. One patient developed severe increase in liver function tests which necessitated discontinuation of bosentan therapy, it was exchanged with iloprost. A second specific vasodilator therapy was added in 15 patients. Of the fifteen patients receiving combined therapy with two drugs, a third drug was added in four patients. Median follow-up time was  $20 \pm 16.5$  months (4–74 mo.). Mean value for six minute walking test increased from  $403 \pm 83$  m to  $482 \pm 62$  m ( $p:0.01$ ). Although clinical symptoms improved in most of the patients, pulmonary artery pressures remained unchanged in ten patients in whom a second catheterization was performed. Seven patients died during follow-up due to progressive clinical deterioration.

**Conclusion:** Pulmonary arterial hypertension is a progressive fatal disease with limited therapeutical options. We believe specific vasodilator therapy causes improvement in quality of life, supported by improvement of symptoms and effort capacity documented with increases in six minute walking test. However, in patients that a second catheterization was performed, despite the subjective clinical improvement patients described, a significant decrease in mean

pulmonary artery pressures could not be shown. Additional prospective, multicenter studies are needed to determine effect of specific vasodilator therapy on life expectancy.

#### O10-7

##### Neurocognitive deficits in children requiring heart transplantation in infancy

Logeswaran T. (1), Behnke-Hall (1), Bauer J. (1), Akintürk H. (1), Schranz D. (1), Hahn A. (2)

Pediatric heart centre, Giessen, Germany (1); Neuropediatric centre, Giessen, Germany (2)

**Introduction:** Heart transplantation (HTx) has become the treatment of choice for infants with end stage heart disease, but data about their neurodevelopmental outcome are rare. Our aims were to assess the neurological and cognitive development of children who underwent HTx during the first two years of life, and investigate which factors contribute to neurocognitive deficits.

**Methods:** Between 1988 and 1999, 42 children underwent HTx in our Pediatric Heart Center due to congenital heart disease (n = 38) or cardiomyopathy (n = 4). 38 Patients (two refused, two were lost during follow-up) were re-examined at least 10 years thereafter. Mean age at re-examination was 13.6 yrs (range 11–23 yrs). Re-evaluation included a detailed neurological examination and assessment of the intelligence coefficient (IQ) by means of the Culture Fair Test (CFT20-R). The following potential risk factors were extracted from the records: waiting time from registration till transplantation, duration of cardiopulmonary bypass (CPB), deep hypothermic arrest (DHCA), additional surgical procedures with extracorporeal circulation, cardiac decompensation and rejection episodes.

**Results:** Mean IQ was  $86 \pm 13$  (range 63–111). 18 patients (47%) had IQs within the normal range ( $\geq 85$ ), 16 (42%) within the range of learning disability (84–70), 3 were mildly mentally retarded (IQ < 70) and 1 was severe mentally retarded (IQ < 50). 4 children (11%) had distinct neurological deficits (i.e. hemiparesis or tetraparesis). Cardiac decompensation was significantly more frequent ( $p = 0.03$ ) in patients with an IQ < 85 (9 out of 20 = 45%) compared to the patients with an IQ  $\geq 85$  (3 out of 18 = 17%). The other variables were not significantly different between the two groups. 27 out of 38 patients had DHCA. Patients with distinct neurological deficits (4) had longer DHCA times ( $55 \pm 12$  vs.  $35 \pm 11$  min.) compared to those with normal neurological status (23); this difference was significant ( $p = 0.01$ ). However, validity of the data was restricted due to inhomogeneous sample size.

**Conclusion:** Neurocognitive deficits are frequent in patients requiring HTx in infancy. Our data suggest that cardiac decompensation is an important factor contributing to cognitive deficits and longer DHCA may cause neurological impairment. Prevention of cardiac decompensation may improve the neurodevelopmental outcome in these patients.

#### 11-1

##### Doctors don't meet information needs of children and teenagers with CHD

Lesch W. (1), Specht K. (1), Lux A. (2), Frey M. (1), Bauer U. (1) German Heart Institute Berlin, Competence Network for Congenital Heart Disease, Berlin, Germany (1) Otto-von-Guericke-Universität Magdeburg, Magdeburg, Germany (2)

**Introduction:** Patients with congenital heart defects (CHD) require lifelong medical care. Lack of cardiological follow-up examinations

and risky behaviour in the transition phase may cause severe complications (Somerville 1997, Mackie 2009) and have a negative influence on the medical outcome. Good knowledge about the heart disease seems to be an important factor for compliance. Therefore we asked children, teenagers and young adults with CHD regarding knowledge and information needs about their CHD.

**Methods:** Descriptive, cross-sectional study. We asked 1.372 patients registered in the German National Registry for CHD to complete an internet-based questionnaire. 52,3% responded (174 children aged between 10–13, 147 teenagers aged between 14 and 17 and 147 adults aged between 18 and 30: 275).

**Results:** Only 33,9% of the children are able to define their CHD (teenagers: 44,9%). The most important source of information is the family (78,7% of the children and 63,9% of the teenagers ask their parents regarding their CHD), followed by the doctor (55,2% of the children and 63,9% of the teenagers ask their doctor). 13,2%/39,5% (children/teenagers) of patients use the Internet as an information source. Doctors are often not able to meet the information needs of the patients (table A). 59,8% of the children and 35,4% of the teenagers state that the doctor only talks to their parents instead of talking to them. Only 40,8% of the children and 41,5% of the teenagers state that they fully understand the explanations of their doctor. This leads to a gap of knowledge about their chronic condition and to many open questions in this patient group. Main topics of interest depend on the age of the patient and are: physical capacity; leisure, sports and travel; employment and career.

**Conclusions:** Young patients with CHD have major gaps in their knowledge about their condition. Doctors do not take the chance to sensitise this patient group to their condition. This lack of knowledge may have harmful consequences. Target group specific information channels are needed, which complement the doctor.

Table A: Comparison of topics of interest and topics discussed with the doctor

Medical topics	Children aged between 10 and 13			Children aged between 14 and 17		
	Information need	Discussed with doctor	Statistical significance	Information need	Discussed with doctor	Statistical significance
Options of treatment	18,4	17,8		31,3	22,4	,041 <sup>a</sup>
Follow-up care		16,1			40,8	
Symptoms	13,8	3,4		18,4	11,6	
Health problems	19,5	12,1	,029 <sup>a</sup>	31,3	20,4	,005 <sup>a</sup>
Medication	10,9	9,2		13,6	18,4	
Physical capacity	48,3	36,8	,006 <sup>a</sup>	66,7	62,6	
Research	10,3	3,4	,008 <sup>a</sup>	10,2	9,5	
Health politics	0,6	0,6		0	0	
No topic	27	37,4	,004 <sup>a</sup>	10,9	17	
Living with a CHD						
Education	24,1	16,7		25,9	24,5	
Employment and career	36,2	8	,000 <sup>a</sup>	36,7	15	,000 <sup>*</sup>
Leisure, sports and travel	58,6	44,3	,001 <sup>a</sup>	52,4	52,4	
Depressions	2,9	0		5,4	0,7	,039 <sup>o</sup>
Anxieties	11,5	2,9	,001 <sup>a</sup>	13,6	1,4	,000 <sup>a</sup>
Friends and Family	10,9	3,4	,004 <sup>a</sup>	8,2	1,4	,006 <sup>*</sup>
Love and sexuality	8	0,6	,000 <sup>a</sup>	13,6	1,4	,000 <sup>a</sup>
Pregnancy, family planning	13,2	1,7	,000 <sup>a</sup>	25,2	4,8	,000 <sup>*</sup>
Smoking, alcohol, drugs	13,8	1,7	,000 <sup>a</sup>	22,8	9,5	,000 <sup>*</sup>
Tattoos, piercing	12,1	2,9	,001 <sup>*</sup>	11,6	2,7	,001 <sup>a</sup>
Beauty	14,4	4	,000 <sup>a</sup>	12,9	2,7	,001 <sup>a</sup>
Legal issues	1,7	0		3,4	0,7	
Insurances	3,4	0,6		10,9	0,7	,000 <sup>a</sup>
No topic	17,8	39,7	,000 <sup>a</sup>	12,9	25,9	,001 <sup>a</sup>

<sup>a</sup>binominal distribution

## PW1-1

### Cardiac Magnetic Resonance Imaging (CMRI) for non-invasive assessment of cardiovascular disease in the follow-up of Kawasaki disease

Kuipers I.M. (1), Tacke C.E.A. (1), Groenink M. (2,3), Spijkerboer A.M. (2), Kuijpers T.W. (1)

Emma Children's Hospital, Amsterdam Medical Center (AMC), University of Amsterdam, Amsterdam, The Netherlands (1); Dept of Radiology, AMC, University of Amsterdam, Amsterdam, The Netherlands (2); Dept of Cardiology, AMC, University of Amsterdam, Amsterdam, The Netherlands (3)

**Background:** Kawasaki disease (KD) is an acute vasculitis in children. KD is the most important cause of acquired coronary artery disease in childhood and young adults. The current guidelines of the American Heart Association (AHA) recommend long-term follow-up of KD patients with serial echocardiography, and nuclear stress-tests and invasive coronary angiography (CAG) in selected patients. Cardiac magnetic resonance imaging (CMRI) is increasingly used as a non-invasive and radiation-free imaging method. We evaluated the feasibility of CMRI during the follow-up of KD patients.

**Methods:** In this single-center study KD patients were recruited during follow-up as outpatients for CMRI with adenosine-stress testing. Until CMRI assessment, patients had been followed by serial echocardiography. CMRI findings were compared with these longitudinal data.

**Results:** Forty patients (73% male, median age 14.7 years) underwent CMRI. Median time interval between disease onset and CMRI was 11.0 years. Coronary artery aneurysms (CAAs) were identified in 12 patients (30.0%) and were unexpected in 7 of these 12 patients because of normal echocardiography findings. Reversible ischemia was observed in 4 (10.0%) and an area of infarction in 3 patients (7.5%). CMRI led to conventional or CT-scanning angiography in 6 patients, resulting in a change in treatment in 7 (17.5%), including coronary artery bypass grafting in 2 patients (5.0%).

**Conclusion:** CMRI is a reliable and sensitive imaging tool for CAA and cardiac function evaluation during follow-up of KD patients. CMRI may be used as an alternative non-invasive and radiation-free imaging method. We recommend incorporation of CMRI performance in addition to echocardiography in the AHA guidelines for long-term follow-up of KD patients.

## PW1-2

### Cardiac synchrony and function with chronic single-site left versus right ventricular epicardial pacing in paediatric patients after patch closure of ventricular septal defects

Tomasek M. (1), Janousek J. (2), Kubus P. (2), Gebauer R.A. (3), Bauersfeld U. (1)

Division of Paediatric Cardiology, University Childrens Hospital Zurich, Switzerland (1); Kardiocentrum and Cardiovascular Research Center, University Hospital Motol Prague, Czech Republic (2); Department of Paediatric Cardiology, Heart Center, University of Leipzig, Germany (3)

**Introduction:** Numerous anatomical pathologies or post-surgical injuries of the conduction system may cause ventricular dyssynchrony in paediatric patients. We evaluated the impact of chronic single-site left (LVP) versus right ventricular pacing (RVP) on ventricular synchrony and function in patients with patch closure for a ventricular septal defect (VSD).

**Patients and Methods:** A total of 23 paediatric patients with large perimembranous VSD patch closure ( $\geq 5$ mm) were enrolled.

Surgical complete heart block was the indication for single-site epicardial right (RVP, n = 9; pacing duration:  $9.6 \pm 5.4$  years) or left ventricular free wall pacing (LVP, n = 14, pacing duration:  $5.7 \pm 3.5$  years). Patients with <1 year and <90% ventricular pacing were excluded. Conventional echocardiography and myocardial 2D strain analysis in the short axis view (circumferential) and 4-chamber view (longitudinal) were obtained.

**Results:** IVMD was significantly prolonged for RVP. Moreover, RVP was associated with significant LV dyssynchrony as evidenced by a prolonged SPWMD and LV intraventricular mechanical delay (Septal-to-lateral delay, SD-time to peak systolic 2D strain). Early systolic bulging of the VSD patch towards the right ventricle led to a prolonged SPWMD for both pacing sites. Global systolic LV function tended to be higher for LVP than RVP (see Table).

**Conclusion:** Conventional and 2D strain echocardiographic measurements indicate preserved LV synchrony and function in paediatric patients after VSD patch closure with LVP but not RVP. However, large VSD patches are acting like a functional aneurysm with paradoxical motion on echocardiography in both groups.

Pacing site	LVP	RVP	P-value
QRS duration (ms)	158 ± 29	164 ± 15	NS
Interventricular mechanical delay, IVMD (ms)	16 ± 14	51 ± 29	0.005
Septal-to-posterior wall motion delay, SPWMD (ms)	125 ± 87	328 ± 40	<0.0001
Septal-to-lateral delay, circumferential (ms)	35 ± 28	115 ± 54	0.0002
Septal-to-lateral delay, longitudinal (ms)	39 ± 31	110 ± 81	0.006
SD-time to peak systolic 2D strain, circumferential (ms)	33 ± 15	76 ± 41	0.002
SD-time to peak systolic 2D strain, longitudinal (ms)	29 ± 10	54 ± 28	0.005
LV ejection fraction (%)	56 ± 8	50 ± 7	NS
LV myocardial performance index	0.37 ± 0.1	0.6 ± 0.1	<0.0001

NS = not significant; SD = Standard deviation.

### PW1-3

#### Echocardiographic Evaluation of Left Ventricular Function over time and Risk Factors for Coronary Artery Lesion Formation in Incomplete Kawasaki Patients

Rhie S.K., Kang S.J.

Dept. of Pediatrics, CHA University, Kyunggi-do, South Korea

Clinical characteristics and risk factors for coronary artery disease in incomplete Kawasaki disease (KD) patients are less studied than in typical KD patients. In addition, the diagnosis of incomplete KD is easily missed, thus leading to delayed treatment and subsequent coronary artery lesions (CAL). In the acute stage of KD, evidence of ventricular dilatation can be found, especially in patients with CAL. Therefore we studied the changes of left ventricular dimensions (LVVID) of CAL(+) patients over time as well as clinical risk factors predisposing to CAL in incomplete KD. We reviewed the medical records of 177 (male 112, female 65, age :  $28.23 \pm 22.63$  months, <12 months : 31.6%, >60 months 10.2%) incomplete KD patients who were treated at CHA Hospital from January 2004 to May 2009. Of these patients, 66 developed CAL (37.3%) and 2 patients had relapsed. LVVID was measured in the acute stage in 24 CAL(+) patients. After 2 months, mean percentile of LVEDD (LVEDD/normal value by BSA) changed from  $1.044 \pm 0.068$  to  $1.027 \pm 0.107$  (mean ± standard deviation), mean percentile of LVESD changed from  $1.095 \pm 0.119$  to  $1.066 \pm 0.192$ , and fractional shortening changed from  $33.82 \pm 4.412$  to  $34.94 \pm 6.21$ . CAL(+) group had a longer duration of total fever than

CAL (-) group ( $6.35 \pm 2.439$  vs.  $5.49 \pm 1.590$ , p-value:0.012). The CAL(+) group had fewer diagnostic symptoms than CAL(-) group ( $3.03 \pm 0.701$  vs.  $3.32 \pm 0.620$ , p-value:0.004). Fewer CAL (+) patient had skin rash compared to CAL(-) patients (p-value:0.025). CAL(+) group showed more males (79%) than CAL(-) patients (54%) (p-value:0.001). BCG erythema didn't show correlation to CAL (p-value:0.864) by chi-square. Among the CAL(-) patients, 30.0% had BCG erythema and 56% had skin rash. Multivariable logistic regression analysis showed that the factors which were significantly associated with the development of CAL were the total number of symptoms (OR:0.255, CI:0.080–0.814, p-value:0.021) and sex (OR:0.287, CI:0.133–0.618, p-value:0.001). Conclusively, LVESD and LVEDD was mild increased in CAL(+) patients but resolved in all patients within 6 to 8 months. Moreover fewer symptoms, absence of rash, longer duration of total fever and male are associated with CAD in incomplete KD. Therefore these patients should be monitored for CAL, especially if unexplained fever continues and immediate management should be initiated.

### PW1-4

#### Reduced left ventricular twist and global strain in patients after tetralogy of Fallot correction is related to right ventricular dysfunction: evidence for detrimental role of apical ventricular-ventricular interaction on left ventricular function

van der Hulst A.E. (1), Delgado V. (2), Holman E. R. (2), Kroft L.J.M. (3), de Roos A. (3), Hazekamp M.G. (3), Blom N.A. (1), Bax J.J. (2), Roest A.A.W. (1)

(1) Dept of Pediatrics, Division of Pediatric Cardiology,

Leiden University Medical Center, Leiden, The Netherlands;

(2) Dept of Cardiology, Leiden University Medical Center, Leiden,

The Netherlands; (3) Dept of Radiology, Leiden University Medical Center, Leiden, The Netherlands

**Objectives:** In patients with corrected tetralogy of Fallot (cToF), left ventricular (LV) dysfunction is closely related to right ventricular (RV) dysfunction, indicating adverse ventricular-ventricular interactions. However, the mechanism that links RV dysfunction to LV dysfunction remains unclear. The current study evaluates the presence of adverse ventricular-ventricular interactions in cToF patients and healthy controls using 2-dimensional (2D) speckle tracking analysis and cardiac magnetic resonance imaging (CMR).

**Methods and results:** Thirty-two cToF patients and 19 controls were studied. With CMR, biventricular ejection fractions were assessed. Using 2D speckle tracking, global and regional RV and LV strain and LV twist were assessed. In cToF patients, a reduced RV ejection fraction, but preserved LV ejection fraction was observed. In addition, global RV strain, global LV strain and LV twist were decreased in cToF patients. cToF patients showed regional LV dysfunction with reduced apical strain ( $-15.9 \pm 2.8\%$  vs.  $-19.8 \pm 2.9\%$ ,  $p < 0.001$ ) and apical rotation ( $5.6 \pm 2.4^\circ$  vs.  $10.0 \pm 2.0^\circ$ ,  $p < 0.001$ ). Global RV strain correlated with global LV strain ( $r = 0.66$ ,  $p < 0.001$ ) and LV twist ( $r = -0.72$ ,  $p < 0.001$ ). Furthermore, close relations were observed between apical RV strain and apical LV strain ( $r = 0.62$ ,  $p < 0.001$ ) and apical LV rotation ( $r = -0.67$ ,  $p < 0.001$ ).

**Conclusion:** In cToF patients and controls, reduced RV strain was strongly related to reduced LV strain and LV twist. Furthermore, apical RV strain correlated with apical LV strain and apical LV rotation, indicating adverse apical ventricular-ventricular interactions.

## PW1-5

### Flow-sensitive four-dimensional (4D) magnetic resonance imaging for fast, multiple and accurate blood flow quantification in patients with congenital heart disease

Nordmeyer S., Khasheei A., Riesenkampff E., Schmitt B., Messroghli D., Berger E, Kuehne T  
German Heart Institute Berlin, Germany

**Introduction:** Flow-sensitive four-dimensional velocity-encoded magnetic resonance imaging (4D VEC MRI) is a novel technique that allows visualizing and quantifying flow in “one-shot” in multiple vessels simultaneously. However, this technique has not yet been validated for quantifying (i) different flow dynamics such as slow continuous venous and fast pulsatile arterial at the same time and (ii) abnormal flow conditions. In this study, we sought to validate the quantitative use of 4D VEC MRI for venous and arterial blood flow in healthy volunteers and for abnormal flow conditions in different congenital heart diseases.

**Methods:** Stroke volumes (SV) in arterial (aorta, main, right and left pulmonary artery) and venous vessels (superior vena cava and right lower pulmonary vein) were compared between standard two-dimensional (2D) and 4D VEC MRI scans (gated and non-gated) in healthy volunteers (n = 7), using a 3T Phillips MRI system. Comparisons of SV and regurgitant fractions (RF) were made between 2D and non-gated 4D VEC MRI methods in patients with congenital heart disease (tetralogy of Fallot with pulmonary insufficiency, n = 5; bicuspid aortic valve with aortic insufficiency or stenosis, n = 5). Agreement between pulmonary and aortic flow volumes was also compared.

**Results:** Bland-Altman testing showed excellent agreement of SV and RF acquired by 2D and 4D VEC MRI (gated and non-gated) in all studied vessels in healthy volunteers and in patients with pulmonary or aortic regurgitation. In volunteers and patients there was agreement between pulmonary and aortic flow. However, patients with bicuspid aortic valves had altered blood flow patterns with vortex formation (turbulences) in close proximity to the aortic valve. These areas of turbulence could be clearly visualized using 4D but not when using 2D VEC MRI. There was good agreement for pulmonary and aortic flow measurements distal to the turbulences, but disagreement for measurements obtained in the area of vortex formation.

**Conclusions:** 4D VEC MRI allows for accurate “one-shot” blood flow quantification in multiple vessels and thus can reduce scan time in patients that require multiple flow acquisitions. Furthermore, and importantly this technique enables visualizing turbulent flow profiles and thus can guide to select the optimum plane for obtaining accurate flow data.

## PW1-6

### Dobutamine Stress-Magnetic Resonance Imaging in repaired Tetralogy of Fallot: Agreement of flow- versus volumetric-derived parameters of cardiac function

Valverde I. (1,2), Parish V. (1), Razavi R. (1,2), Greil G. (1,2), Head C. (2), Qureshi S. (2), Rosenthal E. (2), Schaeffter T. (1), Beerbaum P (1,2).

Imaging Sciences. King's College London. UK. (1); Evelina Children Hospital. Paediatric Cardiology. St. Thomas' & Guy's Foundation Trust. London. UK.

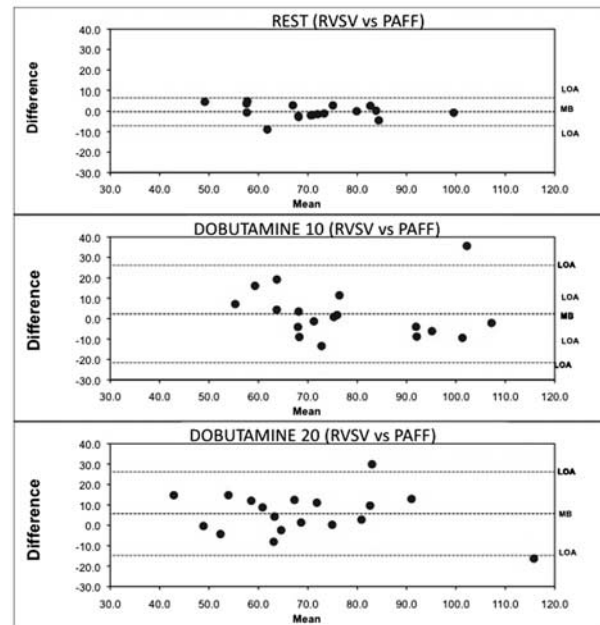
**Introduction:** Dobutamine stress magnetic resonance imaging (DS-MR) is currently being investigated for its utility to determine cardiovascular function in repaired tetralogy of Fallot (r-TOF) with chronic pulmonary regurgitation (PR). Little is known about the comparability of parameters of cardiovascular function

derived by MR flow versus MR volumetry at increasing dobutamine stress levels.

**Methods:** 18 patients with r-TOF and severe chronic PR (age  $34 \pm 12.7$  years, RV-EDV  $126.1 \pm 26.3$  ml/m<sup>2</sup>, PA-RF[flow]  $44 \pm 15\%$ ) underwent cardiac MR at baseline and subsequently at 10 and 20 mcg/kg/min dobutamine. Flow rate in the great arteries (phase-contrast MRI) and functional parameters from axial ventricular volumetry (cine-SSFP-MRI) were measured and analogous parameters calculated for comparison between both modalities (Table 1).

	Rest	Dobutamine 10 $\mu$ g/kg/min	Dobutamine 20 $\mu$ g/kg/min
Heart rate (bpm)	66 $\pm$ 9	86 $\pm$ 14	115 $\pm$ 24
Blood pressure (mmHg)	121 $\pm$ 14 / 71 $\pm$ 6	140 $\pm$ 21 / 75 $\pm$ 4	155 $\pm$ 23 / 77 $\pm$ 7
<b>PC-Flow analysis</b>			
• AO-FF (ml/m <sup>2</sup> )	43 $\pm$ 5	49 $\pm$ 8	36 $\pm$ 15
• PA-FF (ml/m <sup>2</sup> )	71 $\pm$ 13	77 $\pm$ 18	66 $\pm$ 19
• PA-BF (ml/m <sup>2</sup> )	32 $\pm$ 14	35 $\pm$ 18	28 $\pm$ 15
• PA-RF (%)	44 $\pm$ 15	42 $\pm$ 17	40 $\pm$ 15
<b>Volumetric analysis</b>			
• LV-SV (ml/m <sup>2</sup> )	42 $\pm$ 6	47 $\pm$ 9	41 $\pm$ 11
• RV-SV (ml/m <sup>2</sup> )	71 $\pm$ 12	79 $\pm$ 16	72 $\pm$ 17
• PA-BF (ml/m <sup>2</sup> )	28 $\pm$ 14	34 $\pm$ 20	32 $\pm$ 20
• PA-RF (%)	38 $\pm$ 14	40 $\pm$ 18	42 $\pm$ 21

Table 1



**Results:** Under resting conditions, we observed excellent agreement (Bland-Altman) and strong correlation (Pearson coefficients = 0.88–0.96,  $p < 0.05$ ) for volumetric LV stroke volume (LV-SV) versus aortic forward flow (Ao-FF), RV-SV versus pulmonary artery forward flow (PA-FF) as shown in Fig. 1, and absolute pulmonary regurgitant volume by volumetrics (RV-SV minus LV-SV) versus backward flow. However, only fair agreement and correlation was present at dobutamine 10  $\mu$ g/kg/min (Pearson coefficients 0.41/0.41,  $p < 0.05$ ) which did not further deteriorate at 20  $\mu$ g/kg/min. In contrast, for pulmonary regurgitant fraction (PA-RF) the agreement between volumetrics (% of [RV-SV minus LV-SV]/RV-SV), and flow (% of backward

flow/forward flow) was poor at rest and equally at DS-MR. Image quality (4-level score) decreased slightly progressive from rest to DS-MR for both modalities (Friedman's Test,  $p > 0.05$ ) but interobserver variance was consistently satisfactory under stress with all interobserver intra-class correlation coefficients  $> 0.90$  ( $p < 0.001$ ).

**Conclusions:** In repaired TOF with chronic severe PR, agreement between MR flow and volumetry was excellent under resting conditions, except for pulmonary regurgitant fraction, but limited at DS-MR. Interobserver variance did not increase during DS-MR although image quality became moderately reduced. Hence, measures of RV and LV stroke volume, absolute pulmonary regurgitant volume and –importantly– pulmonary regurgitant fraction should not be used interchangeably between both modalities when performing DS-MR in repaired TOF.

**PW1-7**

**Interpretation of left ventricular diastolic dysfunction in children: Time for a new approach?**

Friedberg M.K., Dragulescu A, Mertens L. Hospital for Sick Children, Toronto, Canada

**Background:** Evaluation of left ventricular(LV) diastolic function (DF) in children is difficult and based on adult definitions describing progression from delayed relaxation to restrictive physiology. However, it is unknown whether this classification applies to children. Normal pediatric echo DF values are available but their use in classifying DF has not been shown. Our objectives were to test classification of DF in children with cardiomyopathy (CM) using both adult and pediatric criteria.

**Methods:** 3 investigators independently classified DF as normal, delayed relaxation, restrictive physiology or indeterminate in 4 groups: controls; dilated(DCM), hypertrophic(HCM) and restrictive(RCM) cardiomyopathy, using adult criteria and pediatric reference data (abnormal if  $</> \pm 2$  SD for age). Parameters analyzed were: isovolumic relaxation time, pulmonary venous(PV) S/D, PV A reversal(AR), deceleration time, E/A, AR–A duration, e' and left atrial volume. Inconsistency between echo parameters and lack of agreement between investigators were used to test DF classification. We further analyzed utility of individual parameters to identify diastolic dysfunction.

Criteria	Normal controls (n = 50)		DCM (n = 50)		HCM (n = 50)		RCM (n = 25)	
	Adult	Peds	Adult	Peds	Adult	Peds	Adult	Peds
Age (median, range)	10.3y (0.1–18.3)		8.2y (0.1–17.7)		11y (0.3–18)		12.3y (0.6–18)	
Indeterminate due to inconsistent data	18%	34%	36%	46%	35%	42%	11%	39%
Agreement between all 3 observers	67%	93%	55%	51%	39%	49%	85%	55%
Agreement on classification of DF as abnormal	17%	0	44%	18%	25%	13%	85%	43%
Percentage of abnormal data for individual criteria								
Isovolumic relaxation time	41%	0	62%	49%	46%	26%	89%	33%
PVS/D	18%	4%	30%	24%	24%	23%	52%	44%
PV A reversal	17%	15%	16%	20%	32%	40%	78%	78%
Ar–A duration	2%	-	20%	-	28%	-	78%	-
Deceleration time	29%	11%	83%*	51%*	51%	37%	96%	85%
E/A	43%	0	58%*	11%*	47%	10%	78%	48%
e'	0	0	62%	51%	58%	52%	69%*	63%†
E/mean e'	11%	0	64%	42%	57%	50%	63%*	44%†
LA volume	11%	-	48%	-	56%	-	96%	-

\*A blended mitral inflow precluded analysis in 12% of cases with DCM  
 †Only 60% of echocardiograms in this group had tissue Doppler data

**Results:** 273 echos in 175 children(0–18 yrs) were studied(table). Inconsistencies between echo parameters within patients were relatively common and agreement between observers was poor.

Individual DF parameters were often not informative. For example, PV Doppler was abnormal in only 15–30% of DCM and HCMpts, but was abnormal in a similar % of controls. Delayed relaxation was never diagnosed in DCM and RCM and was rarely diagnosed in HCM patients (7% using adult criteria and 14% by pediatric data). Using pediatric values, most DF parameters were normal in the majority of DCM or HCM pts.

**Conclusion:** Definition of diastolic dysfunction in children using either adult criteria or pediatric reference data is problematic. Pediatric reference data successfully define normals, but due to the large range of normal values, diastolic dysfunction is identified in only a small proportion of cardiomyopathy patients. Discrepancies between criteria within individual pts are common, adversely affecting interpretation and inter-observer agreement. These results suggest that new criteria need to be validated for DF analysis in children.

**PW1-8**

**Higher Dose Dobutamine Stress MR In Tetralogy Of Fallot: Right Ventricular End Systolic Volume (RV-ESV) Is A Reliable Parameter Of Systolic RV Function With Low Observer Variance**

Parish V. (1), Valverde I. (1), Head C. (2), Rosenthal E. (2), Qureshi S. (2), Greil G. (1), Schaeffter T. (1), Razavi R. (1), Beerbaum P. (1) Imaging Sciences, King's College, London, UK (1); Guy's and St Thomas' NHS Trust, London, UK (2)

**Introduction:** In repaired Tetralogy of Fallot (r-TOF) timing of pulmonary valve replacement continues to be debated. Recent work has highlighted the response of right ventricular endsystolic volume (RV-ESV) during dobutamine stress MR (DS-MR) as a potential discriminative marker of early RV dysfunction. However, there is concern that reduced image quality due to motion artefacts at higher doses of Dobutamine may translate into impaired reproducibility and observer performance limiting diagnostic utility, for which endsystolic volumes in particular are known to be at risk. We sought to investigate changes in image quality and observer variance between rest and higher-dose DS-MR.

**Methods:** As part of a prospective trial we identified 18 adult patients (age  $32 \pm 15$ ) with r-TOF, RV dilatation and severe PR who had completed axial ventricular MR volumetry at baseline and during dobutamine infusion of 10 mcg/kg/min and 20 mcg/kg/min (DS-MR). Two independent observers analyzed data at baseline and stress (interobserver); one observer repeated this after 6months (intraobserver). Four independent observers assessed image quality at baseline and stress (4-level modified McConnell-Score, 1 = non-diagnostic to 4 = excellent).

**Results:** Observer variances were expressed as coefficient of variance and mean differences (Tables 1&2). Interobserver comparison showed good agreement for left ventricle (LV) and RV enddiastolic volume measurement equally at rest and both stress levels. At stress the coefficient of variance increased for LV-ESV with each level suggesting less agreement between observers for this measure. In contrast RV-ESV was much more comparable with a coefficient of variance below 10 at each condition. Excellent intraobserver agreement could be seen for all volumetric parameters. Image quality analysis showed that with higher concentrations of dobutamine there is a non-significant reduction in image quality for both ventricles (mean LV-scores 3.0, 2.8, 2.7; RV-scores 3.56, 3.17 and 3.11, at baseline, 10mcg and 20mcg dobutamine respectively. All studies were diagnostic (score  $> 2$ ).

**Conclusions:** Except for LV-ESV, there was good interobserver agreement with little changes between rest and increasing stress levels. Intraobserver agreement was excellent throughout. There-

fore, RV-ESV and its response to stress can be considered as a reliable, robust and reproducible parameter of RV systolic function, particularly when analyzed by a single experienced observer.

Table 1: Inter-observer variability

	Coefficient of variance % (p value (diff between 2 observers t-test))			
	LV-EDV	LV-ESV	RV-EDV	RV-ESV
Baseline	3.5 (p = 0.35)	2.7 (p = 0.31)	2.9 (p = 0.55)	5.0 (p = 0.06)
Dobutamine 10	1.6 (p = 0.46)	12.8 (p = 0.59)	4.2 (p = 0.64)	7.3 (p = 0.01)
Dobutamine 20	4.6 (p = 0.15)	17.9 (p = 0.34)	3.7 (p = 1.11)	8.6 (p = 0.07)
	Mean difference [Range difference min; max]			
	LV-EDV	LV-ESV	RV-EDV	RV-ESV
Baseline	0.99 [-6.5; 11.3]	1.48 [-13.5; 14.4]	-0.92 [-12.1; 10.5]	2.33 [-6.2; 9.8]
Dobutamine 10	1.39 [-8.9; 21.6]	0.66 [-6.9; 7.8]	1.01 [-18.7; 15.4]	3.67 [-4.9; 16.5]
Dobutamine 20	-1.67 [-10.5; 6.3]	1.24 [-7.2; 12.2]	-2.97 [-18.6; 9.3]	2.9 [-9.9; 15.5]

Table 2: Intra-observer variability

	Coefficient of variance % (p value (diff between 2 data sets t-test))			
	LV-EDV	LV-ESV	RV-EDV	RV-ESV
Baseline	2.7 (p = 0.95)	5.8 (p = 0.65)	1.7 (p = 0.93)	1.7 (p = 0.59)
Dobutamine 10	1.7 (p = 0.83)	6.2 (p = 0.48)	2.4 (p = 0.91)	2.3 (p = 0.52)
Dobutamine 20	1.8 (p = 0.29)	6.4 (p = 0.43)	2.1 (p = 0.19)	3.8 (p = 0.99)
	Mean difference [Range difference min; max]			
	LV-EDV	LV-ESV	RV-EDV	RV-ESV
Baseline	-0.08 [-6.8; 6.5]	-0.49 [-7.0; 5.5]	0.11 [-5.8; 8.1]	-0.31 [-3.1; 2.7]
Dobutamine 10	0.15 [-3.4; 3]	-0.63 [-5.3; 2.6]	0.20 [-3.9; 14.78]	0.43 [-4.6; 3.5]
Dobutamine 20	0.68 [-2.2; 4.1]	-0.56 [-3.6; 2.36]	2.05 [-6.78; 11.8]	-0.01 [-4.6; 6.7]

### PW1-9

#### New insights in patients with aortic coarctation – visualization of flow-sensitive data from 4-dimensional MRI

Fischer M. (1), Unterhinninghofen R. (2), Ley-Zaporozhan J. (3), Rengier F. (4,5), Eichhorn J. (1), Ley S. (3)  
 University Children's Hospital, Department of Paediatric Cardiology, Heidelberg (1); Institute of Anthropomatics, University of Karlsruhe (2); University Children's Hospital, Department of Paediatric Radiology, Heidelberg (3); University Hospital, Department of Diagnostic Radiology, Heidelberg (4); German Cancer Research Center, Department of Radiology, Heidelberg (5)

**Introduction:** Cardiovascular flow measurements using phase contrast MRI are well established techniques in paediatric cardiology, normally acquired 2-dimensionally. Modern sequence protocols allow the acquisition of velocity vector fields now in all three spatial directions with high temporal resolution

(4-dimensional MR). This information can be visualized using streamlines, particle traces or vector maps to enhance previously unknown aspects of flow like turbulences, helical flow profiles, and local velocity inhomogeneities. The aim was to characterize different flow aspects in patients with aortic coarctation (CoA). **Methods:** Using a 1.5T MRI (Magnetom Avanto®, Siemens) a stack of phase-contrast FLASH 2D sequences with tridirectional flow acquisition was used to cover the whole aorta (spatial resolution  $1.6 \times 1.6 \times 2.1$  mm, free breathing). In an initial study, flow analysis was performed in 13 patients ( $f/m = 6/7$ , median age 17.1 [3.8–33.1] years) with CoA (native:  $n = 2$ , recoarctation after surgery/balloon angioplasty:  $n = 11$ ) using in-house developed software (Mediframe®).

**Results:** In all patients detailed flow pattern in the aorta could be visualized. Beside the 3D visualization of stenotic jets and quantification of flow velocity for each aortic voxel, individual helical flow pattern were found in 6 patients. The mean peak velocity of blood flow distal to the side of aortic coarctation was 278 [190–400] cm/s).

**Conclusions:** In patients with aortic coarctation new methods of flow visualization help understanding complex local flow pattern. Even more these visualizations improve to understand pathomorphological changes of the aortic wall before and after surgical or interventional repair treatment.



### PW1-10

#### Cardiac-committed human embryonic stem cells as a therapeutic agent in congenital right ventricular dysfunction?

Lambert V. (1), Capderou A. (2), Leuret E. (1), Rücker-Martin C. (2), Gouadon E. (2), Raymond N. (1), Dinanian S. (3), Stos B. (1), Pucéat M. (4), Serraf A. (1), Renaud J.F. (2)  
 Centre Chirurgical Marie-Lannelongue (1); Le Plessis Robinson, France; INSERM U-999 (2); Le Plessis Robinson, France; Centre Hospitalier Universitaire Antoine Bécère (3); Clamart, France; INSERM Avenir Program, stem cell and cardiogenesis (4); Evry, France

**Introduction:** Advances in cardiac surgery have significantly improved short-term prognoses of patients with congenital heart diseases but, at long-term, right ventricular (RV) failure may occur, leading to morbidity and mortality. Since conventional therapy gives poor results, cell therapy may be a therapeutic

option for cardiac repair. The ability of embryonic stem cells (ESC) differentiation towards a cardiomyogenic phenotype makes them attractive candidates. Our aim was to evaluate, in a large animal model, the effects of cardiac-committed human ESC transplantation on the RV function.

**Methods:** A model of RV dysfunction was created in 6 piglets using a surgical procedure mimicking RV tract sequelae of repaired tetralogy of Fallot (pulmonary regurgitation and pulmonary branch stenosis) leading, after 4 months, to a chronic combined (volumetric and barometric) RV overload. Three pigs received vehicle and 3 others received HUES-24 derived human cardiac progenitor cells injected at multiple sites into the free wall of RV myocardium. All pigs were immunosuppressed using tacrolimus. Myocardial function was measured 3 months after cell transplantation by conductance catheter technique using maximal elastance ( $E_{max}$ ) slope and ventricular energetics (stroke work, pressure-volume area). The risk of ventricular arrhythmia was evaluated by programmed ventricular stimulation at the end of the follow-up. Structural remodelling was assessed by histological examination.

**Findings:** All pigs survived and no complication (inflammation, teratoma, infection) occurred related to either myocardial injections or immunosuppression. No ventricular arrhythmia was induced. In all treated pigs, myocardial contractility was improved as assessed by an increasing  $E_{max}$  slope relative to baseline (mean values:  $0.61 \pm 0.02$  vs  $0.40 \pm 0.01$ ,  $p < 0.001$ ) and to controls at the follow-up end ( $0.32 \pm 0.02$ ,  $p < 0.05$ ). Whereas stroke work was similar in both groups, pressure-volume area tended to increase in the treated group ( $6813 \pm 1773$  vs  $4171 \pm 804$  mmHg.mL in controls) suggesting that the total energy output was augmented. In treated pigs, myocardial fibrosis appeared peritrabecular whereas, in controls, it was found in peritrabecular and perimyocyte areas.

**Conclusion:** Cell therapy by multiple trans-epicardial injections of ESC derived human cardiac progenitor cells in failed RV secondary to chronic overload is feasible, improves RV myocardial contractility and allows a better adaptation to chronic overload.

#### PW1-11

##### Is early tissue Doppler diastolic velocity ( $E'$ ) useful to diagnose left ventricular diastolic dysfunction in children with cardiomyopathy?

Friedberg M.K., Dragulescu A., Mertens L.  
Hospital for Sick Children, Toronto, Canada.

**Background:** Early diastolic tissue Doppler (TDI) velocities ( $e'$ ) may be useful to diagnose diastolic dysfunction (DD) when 'traditional' diastolic parameters are discrepant or in the presence of mitral pseudonormalization. Recent ASE guidelines place  $E'$  as a determining criterion to diagnose DD: a low  $E'$  determines diastolic dysfunction and an elevated  $E/E'$  ratio determines elevated filling pressures. However, it is not known whether this is applies to children. The objective of this study was to examine whether  $E'$  and  $E/E'$  are abnormal in children with cardiomyopathy (CM).

**Methods:**  $E'$  was measured from digitally stored pulsed TDI at the lateral mitral annulus and basal septum in controls; dilated (DCM), hypertrophic (HCM) and restrictive (RCM) CM patients.  $E/E'$  ratio was calculated using a mean of  $E'$  at these 2 locations. Using normal pediatric reference data, we defined  $E'$  as either normal or abnormal (abnormal if  $</> \pm 2SD$  for age). We further assessed whether  $E'$  and  $E/E'$  were consistent with other echocardiographic diastolic parameters (isovolumic relaxation time, pulmonary venous S/D, PV A reversal, deceleration time, E/A ratio) in individual

patients. The % of abnormal TDI data in each group and agreement with conventional criteria for DD were analyzed.

**Results:** 262 echoes in 164 children were studied (figure). 4 patients had blended TDI waves at the lateral and septal walls precluding analysis. All children in the control group had normal values. Percentages for each CM group are shown in the table. Approximately half of all CM patients had normal septal  $E'$  values. The proportion of normal values was even higher for the mitral lateral annulus  $E'$  and for  $E/\text{mean } E'$  ratio. The  $E'$  was discrepant from conventional criteria in 60–70% in all CM groups.

	Normal controls (n = 50)	DCM (n = 5D)	HCM (11 = 50)	RCM (n = 14)
Age (mean $\pm$ SD)	9.7 $\pm$ 5.1	7.8 $\pm$ 5.4	10.4 $\pm$ 4.5	9.8 $\pm$ 5.5
Lateral mitral $E'$				
Lateral mitral $E'$ (mean $\pm$ SD)	18.2 $\pm$ 3.9	11.6 $\pm$ 4.7	12.5 $\pm$ 4.6	10.4 $\pm$ 4.8
% of abnormal values	0	25%	30%	56%
Disagreement with conventional criteria	7%	62%	60%	69%
Septal $E'$				
Septal $E'$ (mean $\pm$ SD)	13.9 $\pm$ 2.5	7.9 $\pm$ 2.7	8.3 $\pm$ 3.3	7.8 $\pm$ 3.4
% of abnormal values	0	53%	54%	56%
Disagreement with conventional criteria	7%	66%	60%	69%
$E/\text{mean } E'$ ratio				
$E/\text{mean } E'$ ratio (mean $\pm$ SD)	6.7 $\pm$ 1.7	11.4 $\pm$ 4.5	11 $\pm$ 5	11.5 $\pm$ 6
% of abnormal values	0	42%	46%	44%
Disagreement with conventional criteria	7%	54%	62%	63%

**Conclusions:** Pediatric reference data for tissue velocities successfully define normals but have large ranges. In half of the patients with CM  $E'$  is within normal range and is often inconsistent with other echo parameters for DD. This suggests that  $E'$  is less useful for definition of DD in children with CM than in adult population and that recent ASE guidelines are not applicable to the pediatric CM population.

#### PW1-12

##### Quantification of regional left and right ventricular longitudinal function by 2D speckle tracking imaging in children after atrioventricular node slow pathway ablation

Sileikiene R. (1), Vaskelyte J. (2), Baksiene D. (1), Mizariene V. (2), Ziuteliene A. (1), Jurkevicius R. (3)

Department of Children Diseases, Kaunas Medical University Hospital, Kaunas, Lithuania (1); Institute of Cardiology, Kaunas, Lithuania (2); Kaunas University of Medicine, Lithuania (3)

**Introduction:** Radiofrequency ablation (RFA) of slow pathway in the case of tachycardia is the method of therapy. But the regional impairment of systolic and diastolic function of left and right ventricles (LV, RV) may occur. 2D speckle tracking imaging (2D STI) technique was used for quantification of regional myocardial function.

**Purpose:** To assess regional LV and RV longitudinal function by 2D STI in children at late follow-up after slow pathway ablation and in healthy children.

**Methods:** The study group consisted of 21 subject: 11 children (group 1), mean age 16,  $7 \pm 2$ , 7 years, who underwent RFA 5,  $5 \pm 2$ , 5 years ago and 10 healthy children (group 2), mean age 16,  $1 \pm 1$ , 7 years. Conventional echocardiographic measurements did not differ between the groups. Measurements were performed at basal, mid and apical segments of the LV inferior, anterior, lateral wall, the ventricular septum and the RV lateral wall using 2D STI for the assessment of strain ( $\epsilon$ ) and strain rate (SR).

**Results:** LV and RV longitudinal deformation values were homogenous for all LV segments and there were no significant



differences between the subjects of both groups: LV peak  $\varepsilon$   $-21,55\% \pm 3,62\%$  in group 1 vs.  $-20,77\% \pm 3,81\%$  in group 2; peak systolic  $\varepsilon$   $-17,48\% \pm 2,04\%$  vs.  $-18,17\% \pm 3,27\%$ ; peak systolic SR  $-1,23 \pm 0,46$  s<sup>-1</sup> vs.  $-1,32 \pm 0,525$  s<sup>-1</sup>, early diastolic SR  $1,71 \pm 0,36$  vs.  $1,86 \pm 0,25$  s<sup>-1</sup>, late diastolic SR  $0,95 \pm 0,43$  vs.  $1,04 \pm 0,29$  s<sup>-1</sup>. RV peak  $\varepsilon$   $-20,63\% \pm 5,43\%$  vs.  $-20,66\% \pm 5,43\%$ , peak systolic  $\varepsilon$   $-17,89\% \pm 5,75\%$  vs.  $-19,54\% \pm 5,38\%$ , peak systolic SR  $-1,33 \pm 0,60$  vs.  $-1,25 \pm 0,65$  s<sup>-1</sup>, early diastolic SR  $1,81 \pm 0,41$  vs.  $1,67 \pm 0,39$  s<sup>-1</sup>, late diastolic SR  $1,03 \pm 0,44$  vs.  $0,93 \pm 0,29$  s<sup>-1</sup>.

**Conclusions:** This study results suggest that data collected from children after RFA of slow pathway did not differ from data of healthy children, measured by 2D STI. There was no impairment of regional longitudinal function in children late after RFA.

#### PW2-1

##### Long-term left ventricular adaptation in children and adolescent athletes

Barkeviciene A. (1), Baksiene D. (2), Raskauskiene N. (1), Vainoras A. (2), Brozaitiene J. (1)

Institute of Psychophysiology and rehabilitation of Kaunas Medical University, Palanga, Lithuania (1); Kaunas Medical University, Kaunas, Lithuania (2).

**Objectives:** Left ventricular adaptation to long-term exercise training in athletic children and adolescents is influenced by several factors including age, anthropometric data, sporting discipline, training duration and volume, systolic blood pressure at rest and at peak exercise. The aim of this study was to identify the factors influencing left ventricular morphometric parameters in child and adolescent athletes.

**Methods:** The study procedure was approved by regional bioethics committee. One hundred sixty seven male athletes aged 14.8 (SD1.6, range 12–17 years) participating in basketball, rowing and cycling and 168 healthy sedentary controls matched for age, sex and body surface area (BSA) were studied echocardiographically to assess LV internal diastolic diameter (LVIDd), interventricular septal thickness (IVSTd) and LV posterior wall thickness (LVPWTd). Left ventricular mass (LVM) was calculated. The participants completed cycle ergometer test to assess systolic blood pressure (SBP) at rest and at peak exercise. Multivariate regression analysis was used to identify independent determinants of LVIDd, IVSTd, LVPWTd and LVM for both the athlete and control subjects.

**Results:** All absolute echocardiographic parameters (LVIDd, IVSTd, LVPWTd and LVM) and parameters indexed to body surface area and height of athletes were greater than analogous parameters in controls, except for LVIDd indexed to height. In univariate analysis, LVM of athletes correlated with age, height, body weight, BSA, training volume, training duration, SBP at rest and at peak exercise. LVIDd was associated with training volume and duration. IVSTd and LVPWTd correlated with training volume. LVM of controls had correlations with age, height, BSA and SBP at peak exercise. In multivariate analysis, only age and body weight were independent predictors of LVM of athletes ( $R^2 = 63.6\%$ ). Body weight, training volume and sporting discipline were independent determinants of LVIDd ( $R^2 = 53.5\%$ ). Age and training volume predicted IVSTd ( $R^2 = 50.6\%$ ), BSA and age were predictors of LVPWTd ( $R^2 = 41.3\%$ ). All echocardiographic parameters of controls were dependent on age and BSA ( $R^2 = 59.8\%$ ).

**Conclusion:** Training volume was independent predictor of interventricular septal thickness and LV internal diastolic diameter, sporting discipline predicted LV internal diastolic

diameter, LV mass and LV posterior wall thickness of athletes were predicted by age and anthropometric parameters.

#### PW2-2

##### Brain Natriuretic Peptide response in functionally univentricular heart: from first days of life to the follow-up of total cavo-pulmonary connection.

Cantinotti M., Margaryan R., Arcieri L., Maizza A., Storti S., Prontera C., Luisi S., Bernabei M., Crocetti M., Assanta N., Murzi B.

Pediatric Cardiac Surgery Unit, G Monasterio Foundation and Institute of Clinical Physiology-National Research Council, Heart Hospital, Massa, Italy

**Introduction:** To evaluate brain natriuretic peptide (BNP) behaviour and its possible implication on the evaluation of functionally univentricular heart (FUH) during different stages of Univentricular Physiology Palliation (UPP).

**Methods:** Jul 2005–Mar 2009: BNP was measured in 179 patients with FUH and 350 healthy control subjects.

We identified 4 group of patients.

1. 48 un-palliated FUH. Mean age 0,37 months (range 1 day–2,6 yrs).
2. 37 had palliated FUH (Norwood stage 1, pulmonary artery banding, MBT shunt). Mean age 15,4 months (range 2,4 months–13,1 yrs).
3. 52 Bidirectional Glenn (BDG). Mean age 5 years (range 0,7–24,7 yrs).
4. 42 TCPC. Mean age 10,7 years (range 5,8–25,3 yrs).

Within each of the three main group of palliated patients we identified 2 further subgroup: complicated and uncomplicated. Complicated were considered those with at least one of the following (impaired ventricular systolic function; at least moderate valvular regurgitation; systemic obstruction; venous obstruction; pulmonary arteries stenosis; rhythm disturbance; neurologic disorders, others).

**Results:** In all the groups BNP values were significantly higher than in controls.

BNP values were significantly higher in un-palliated FUH than in all other subgroups ( $P < 0.001$ , by Scheffé test after one way ANOVA). Through all the stages of UPP a significant trend to decrease of BNP values was noted ( $P < 0.0001$ , Spearman rank correlation  $-0.429$ ). In all the groups patients with right ventricular main chamber anatomy had higher BNP values than both left and balanced FUH ( $P < 0.001$ , Scheffé test). BNP values positively correlated with the degree valvular regurgitation, NYHA class ( $P < 0,001$ ) and amount of medications ( $P < 0,01$ ) and negatively with systolic function and O<sub>2</sub> saturation ( $P < 0,001$ ). In all the groups complicated pts had BNP significant higher than others ( $P < 0,001$ ). In Glenn and TCPC uncomplicated cases BNP values were not significantly different than normal subjects.

**Conclusions:** FUH shows higher levels of BNP than normal subjects do. A progressive fall of BNP values has been observed during the different stages of univentricular physiology palliation. Right ventricular main chamber anatomy had higher BNP values. In Glenn and TCPC BNP values of uncomplicated cases were similar to normal subjects.

#### PW2-3

Withdrawn

**PW2-4****BMPR2 mutation in Children with Pulmonary Arterial Hypertension associate with Congenital Heart Disease**

Limsuwan A. (1), Wattanasirichaigoon D. (2)

Division of Pediatric Cardiology(1); Division of Clinical Genetics (2), Department of Pediatrics, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand.

In patients with idiopathic pulmonary hypertension (iPAH) the mutations of bone morphogenetic protein receptor type 2 (BMPR2), which have an impact on the clinical outcome, was well defined. We investigated the occurrence of BMPR2 mutation in children with associate pulmonary arterial hypertension to congenital heart disease (aPAH/CHD) and correlate with the disease severity and the pulmonary vasoreactivity testing.

**Methods:** BMPR2 mutation types were determined in 30 aPAH/CHD children. All children underwent cardiac catheterization to obtain baseline hemodynamic data and responsiveness to pulmonary vasoreactivity testing. Determination of pulmonary vasoreactivity response in patients with aPAH/CHD is focus on the alteration of pulmonary vascular resistance (PVR) and pulmonary blood flow (Qp) while the pulmonary artery pressure(PAP) usually remain unchanged.

**Results:** Of 30 patients (median-age 90 months) with aPAH/CHD (mean PAP  $40 \pm$  mmHg, PVR  $6.7 \pm 4.2$ ), 23 (76%) were BMPR2 mutation-negative, and 7 (24%) were mutation-positive. Both BMPR2 mutations –positive and negative have similar response to pulmonary vasoreactivity test (86% vs 87%,  $p = 0.93$ ) and pulmonary hemodynamics (mean PAP  $44 \pm 15$  vs  $49 \pm 17$  mmHg,  $p = 0.45$ ; PVR  $5.7 \pm 4.1$  vs  $7.0 \pm 4.3$  WU.m<sup>2</sup>,  $p = 0.35$ ).

**Conclusions:** BMPR2 mutation-positive were found 24% of children with aPAH/CHD. In our cohort, these genetic mutations did not distinguish the responsiveness of pulmonary vasoreactivity testing or the severity of pulmonary hypertension in term of pulmonary hemodynamics. Therefore determination of BMPR2 mutation in children with aPAH/CHD is unlikely to predict the clinical severity of the disease and perhaps the disease outcome.

**PW2-5****The usefulness of scintigraphy with 99m Tc-Anti-Granulocyte antibody for diagnosis and follow-up in children with myocarditis.**

Ziolkowska L., Kawalec W., Biernatowicz M., Swiatek-Rawa E., Kaminska A., Tirska-Kmiec A., Boruc A., Czarnowska E., Brzezinska-Rajszys G., Raszek M.

The Children's Memorial Health Institute, Warsaw, Poland

**Objectives:** Clinical diagnosis of myocarditis (myo) is difficult. The aim of study was to investigate whether scintigraphy with 99mTcAGA is useful for diagnosis and follow-up in children with myo and what is its correlation with EMB.

**Methods:** From 2005 to 2009, 11 children, mean age  $13 \pm 3.8$  yrs presenting with symptoms of myo were evaluated at the time of initial presentation and 6, 12 and 24 months after the first study. In all pts heart scintigraphy with 99mTcAGA was performed. Estimation of antigranulocyte antibodies uptake was performed by calculation of the heart-to-lung ratio (HLR), value above 1.50 was used as a positive result. Control group consisted of 10 children without cardiovascular disease underwent scintigraphy with 99mTcAGA due to suspicion of enterocolitis (HLR 1.06–1.50). EMB was done in 10 pts at the time of initial presentation and after 6 months.

**Results:** The mean time from onset of symptoms of myo to diagnosis was 2,52 months. All pts presented with fatigue, heart failure ( $n = 4$ ), ventricular arrhythmias ( $n = 6$ ), syncope ( $n = 2$ ), chest pain ( $n = 6$ ). In 10 (91%) pts positive antigranulocyte uptake was observed (mean HLR 1,96; range 1,51 to 3,2), EMB was performed in 9 of them (no consent for EMB in 1 pt). EMB results in 8 pts showed evidence of myo, in 1 pt no features of myo. In follow-up scintigraphy after 6 months positive uptake was observed in 9 (82%) pts, the mean HLR was 1,97 (range 1,51 to 2,9), in EMB performed in 8 pts persistent inflammatory process in myocardium was observed. The scintigraphy after 12 months indicated positive uptake in 8 (73%) pts, mean HLR 1,88 (range 1,51 to 3,0). The scintigraphy after 24 months was positive only in 4 (36%) pts, mean HLR 2.03 (range 1,7 to 2,4).

**Conclusion:** (1) In 89% pts with positive scintigraphy results biopsy-proven myocarditis was observed. (2) The control scintigraphy performed in follow-up after 6, 12, 24 months enable to evaluate resolved or persistent myocarditis. (3) Scintigraphy with 99mTcAGA seems to be a useful diagnostic method in children with suspected myocarditis, but further studies are needed to establish its sensitivity and specificity.

**PW2-6****Effect of dexamethasone on the inflammatory response and myocardial protection during and after arterial switch operation in neonates: A double blind randomized controlled study**

Heying R. (1,3), Wehage E. (1), Tassani P. (2), Haas F. (2), Hess J. (2), Seghaye M.C. (1)

Department of Paediatric Cardiology, University Hospital Aachen, Germany (1); Department of Paediatric Cardiology, German Heart Centre at the Technical University Munich, Germany (2); Department of Paediatric Cardiology, KU Leuven, Belgium (3)

**Introduction:** Cardiac surgery with cardiopulmonary bypass (CPB) is associated with a systemic inflammatory response that, if uncontrolled, is associated with organ damage. The anti-inflammatory effects of glucocorticoids might be beneficial in the context of neonatal cardiac surgery. This study was designed to answer the question of whether preoperative treatment with dexamethasone would attenuate the perioperative inflammatory response and ameliorate postoperative outcome.

**Methods:** In 20 neonates (age: 8–21 days) with transposition of the great arteries undergoing arterial switch operation intramyocardial expression of the pro-inflammatory cytokines interleukin (IL)-6, IL-8, IL-1 $\beta$  and tumor necrosis factor  $\alpha$  (TNF- $\alpha$ ) as well as the anti-inflammatory cytokine IL-10 were quantified on mRNA level in right atrial tissue taken before institution of CPB. In addition plasma levels of IL-6, IL-10, lipopolysaccharide binding protein (LBP) and cardiac Troponin T (cTnT) were measured before, during and after CPB.

**Results:** Patients treated with dexamethasone showed lower myocardial expression of the mRNA encoding for the pro-inflammatory cytokines IL-8, IL-6, TNF- $\alpha$ , and IL-1 $\beta$  but not of the mRNA encoding for IL-10 in comparison to patients of the control group. They also showed lower plasma levels of IL-6 and higher levels of IL-10 during and after CPB than patients of the control group. cTnT levels were significantly lower and LBP levels higher after CPB in treated as compared to control patients. Postoperative catecholamine requirements were lower in treated patients than in controls.

**Conclusions:** Dexamethasone administration in neonates prior to arterial switch operation induces a significant reduction of myocardial mRNA expression of pro-inflammatory cytokines

already before connecting CPB, and a shift of the inflammatory response during and after CPB toward an increased anti-inflammatory reaction. This is associated with a significant reduction in postoperative release of cTnT, suggesting myocardial protection.

## PW2-7

### Rotational Mechanics and Left Ventricular Propagation Velocity Elucidate Mechanisms of Left Ventricular Diastolic Dysfunction in Children with Dilated Cardiomyopathy

Dillenburg R., Mertens L., Friedberg M.; Hospital for Sick Children, Toronto, Canada

**Objectives:** Left ventricular (LV) early diastolic filling is dependent on systolic torsion and subsequent LV untwisting. Therefore, decreased LV systolic function may impair diastolic filling in children with dilated cardiomyopathy (DCM). The objective of this study was to examine early LV filling in children with DCM in relation to LV rotational mechanics.

**Methods:** Echocardiograms of children (0–18 yr) with dilated cardiomyopathy (DCM) and LV EF < 50% were compared with age-matched controls. Mitral inflow velocities and deceleration time, isovolumic relaxation time (IVRT) and pulmonary vein flow were assessed by Doppler. LV inflow propagation velocity (Vp) was measured from the slope of a color M-Mode interrogation line placed from LV base to apex. Early diastolic mitral velocities (E') were assessed by pulsed tissue Doppler. LV apical and basal circumferential peak rotation, rotation rate, torsion and untwist rate were measured using 2-D speckle tracking from short axis views using frame rates of 60–90 fps (Echopac, GE).

Variable	Controls (n = 12)	DCM (n = 12)	p-value
Age (years), mean ± SD	11.6 ± 5	10.4 ± 5.7	0.14
BSA (m <sup>2</sup> ), mean ± SD	1.35 ± 0.45	1.27 ± 0.55	0.46
HR (bpm)	72.9 ± 12.2	80.7 ± 29.1	0.31
LV EF (%)	63.5 ± 7.1	37.1 ± 10.5	< 0.0001
LVED (cm)	4.2 ± 0.79	5.4 ± 0.98	0.0012
Mitral E (cm/s)	106 ± 15.6	104 ± 18	0.82
Mitral E/A ratio	2.4 ± 0.61	2.5 ± 1.0	0.66
Mitral E deceleration time (ms)	145 ± 15.6	146 ± 40.2	0.94
IVRT (ms)	70 ± 8.2	74 ± 15.1	0.47
Pulmonary vein systolic velocity (cm/s)	42.5 ± 7.3	46.2 ± 12.6	0.39
Pulmonary vein diastolic velocity (cm/s)	58.5 ± 12.9	65 ± 13.6	0.75
Pulmonary vein atrial reversal velocity (cm/s)	19.1 ± 3.5	20.3 ± 6.1	0.40
Pulmonary vein atrial reversal duration (ms)	95.6 ± 19.1	116.4 ± 21.3	0.22
E', lateral (cm/s)	18.7 ± 2.6	14.1 ± 6.1	0.007
E/E' ratio, lateral	5.8 ± 1.5	9.4 ± 6.8	0.06
E', septal (cm/s)	13.9 ± 2.1	11.7 ± 2.7	0.04
E/E' ratio, septal	7.4 ± 1.6	9.5 ± 2.6	0.03
Peak rotation, base (deg)	-5.14 ± 2.43	-3.49 ± 1.97	0.08
Peak rotation, apex (deg)	4.61 ± 1.34	1.61 ± 3.11	0.0054
Peak torsion (deg)	9.8 ± 1.8	5.1 ± 3.5	0.0008
Peak rotation rate, base (deg/s)	-64.3 ± 21.45	-37.55 ± 24.16	0.039
Peak rotation rate, apex (deg/s)	65 ± 22.3	26.9 ± 28.35	0.0032
Peak torsion rate (deg/s)	129.3 ± 24.1	64.54 ± 44.3	0.0016
Peak untwist rate, apex (deg/s)	-54 ± 36.4	-28.4 ± 28.7	0.044
Peak untwist rate, base (deg/s)	57.9 ± 19.5	38.7 ± 26.2	0.03
Peak untwist rate (deg/s)	-112.1 ± 39.2	-67.13 ± 38.39	0.019
LV propagation velocity (cm/s)	59.6 ± 17.6	42.8 ± 12.4	0.009

**Results:** Echocardiograms of 12 children with DCM and 12 controls were studied. Results are shown in the table. Peak early LV filling (E) and relaxation (deceleration time and IVRT) were equal in DCM patients and controls; while torsion, untwisting, early diastolic wall motion (E') and Vp were lower in DCM. The E/E' ratio, a marker of LV filling pressures, was elevated in DCM

while pulmonary venous Doppler parameters were not different between the groups.

**Conclusions:** These results suggest that in children with DCM, decreased LV systolic torsion is associated with decreased untwisting and decreased early diastolic suction measured by Vp. However, early LV filling remains normal due to elevated filling pressures. Our results suggest that rotational mechanics and Vp should be evaluated when assessing diastolic function in DCM, as conventional echo parameters are inadequate (due to pseudonormalization) to reveal ubiquitous diastolic dysfunction in these patients.

## PW2-8

### Assessment of myocardial performance index in fetuses with supraventricular tachycardia successfully treated in utero

Dangel J.H., Hamela-Olkowska A.

Perinatal Cardiology Dept., 2nd Dept of Obstetrics and Gynecology, Medical University of Warsaw, Poland

**Objectives:** Myocardial performance index (Tei index, MPI) is defined as a sum of isovolumic contraction and relaxation time divided by ejection time and it is used to measure the global cardiac function. The aim of this study was to assess MPI for the left (LV) and the right ventricle (RV) in fetuses with supraventricular tachycardia (SVT) at the time of diagnosis and at the day of cardioversion to sinus rhythm.

**Methods:** Doppler flow-derived measurements of ventricular performance were obtained with echocardiography in 22 fetuses with SVT successfully treated in utero with digoxin and amiodarone, in monotherapy or combination. All fetuses had normal heart anatomy, 9 were hydropic. There was 13 SVT with short VA conduction time and 9 with long VA. Mean gestational age at diagnosis was 29 ± 5 weeks. Cardioversion time ranged from 1 to 51 days (median 10 days). Fetal echocardiographic examinations were performed using transabdominal convex probe 3,5–6 MHz of Acuson Sequoia 512. Tei indexes were calculated off-line as a subtraction of time between closure and opening of the atrioventricular valve and the ejection time, divided by the ejection time. Normal values in our laboratory are: TeiLV = 0,47 ± 0,07, TeiRV = 0,48 ± 0,1.

**Results:** FHR at the time of diagnosis ranged from 202 to 300 bpm (mean 254 ± 29 bpm), at the day of cardioversion from 112 to 145 bpm (mean 127 ± 9 bpm). MPI for LV at the time of diagnosis ranged from 0,31 to 1,47 (mean 0,7 ± 0,27), at the day of cardioversion: from 0,3 to 0,56 (mean 0,46 ± 0,07), p < 0,001. MPI for RV at the time of diagnosis ranged from 0,37 to 1,11 (mean 0,71 ± 0,19) and at the day of cardioversion: from 0,4 to 0,61 (mean 0,49 ± 0,06), p < 0,001. MPI was increased in 11 fetuses at presentation, in comparison to just 4 fetuses in whom SF was decreased.

**Conclusions:** MPI is useful for monitoring the effectiveness of antiarrhythmic treatment in fetuses with SVT. Tei index is superior to SF probably due to more severe impairment of diastolic, than systolic function during fetal SVT. According to our knowledge, such findings were not published before.

## PW2-9

### Prevalence of anemia in children with congestive heart failure.

Mueller G.C. (1), Schlueter E.L. (1), Arndt F (1), Banhidai M. (1), Dodge-Khatami A. (2), Weil J. (1), Mir T.S. (1)

Department of Pediatric Cardiology, University Heart Centre Hamburg (1); Department for Pediatric Cardiac Surgery, University Heart Center Hamburg (2).

**Introduction:** Anemia is prevalent in adult patients with congestive heart failure (CHF) and appears to be an independent risk factor for higher morbidity and mortality in patients with heart failure. The purpose of this work is to determine the prevalence of anemia in children with heart failure and to evaluate its influence on morbidity and mortality.

**Methods:** Between 1998 and 2007, 3996 patients were retrospectively evaluated for congestive heart failure. CHF was diagnosed in 693 patients (17.3%), of which 58 children with dilative cardiomyopathy were included in the study group and analyzed for heart failure symptoms, brain natriuretic peptide levels, hemoglobin levels, hospitalization rate in days per year, age of first clinical symptoms, necessity of transfusion and death during medical attendance. Anemic and non-anemic congestive heart failure patients were divided into groups and analyzed for differences in age distribution, morbidity and mortality.

**Results:** Anemia was present in 64% of pediatric patients with congestive heart failure. Hospitalization days per year served as a surrogate for morbidity. Anemic CHF patients had a significantly higher hospitalization rate than non-anemic patients (mean  $35.1 \pm 40.5$  days per year vs.  $9.97 \pm 9.65$  days per year,  $p < 0.05$ ). Significant relations between the onset of heart failure symptoms and the risk of developing anemia were not found between anemic and non-anemic patients. The evaluation of the mortality rate showed no significant difference between anemic and non-anemic congestive heart failure patients.

**Conclusion:** Anemia is prevalent in pediatric patients with heart failure and is associated with higher morbidity. Higher mortality did not appear in our population of anemic patients. Further studies are needed to determine the causes of anemia in pediatric heart failure patients and to explore whether correction of anemia could reduce morbidity and improve quality of life.

#### PW2-10

##### **Quality of Life and Its Determinants in Parents of Children, Adolescents, and Young Adults with Congenital Heart Disease**

Hua, Y.C. (1), Chen, C.A. (2)

Cardiac childre's foundation, ROC (1); Division of Pediatric Cardiology, Department of Pediatrics, National Taiwan University Hospital, Taipei, Taiwan (2)

**Objectives:** Knowledge of parental quality of life (QoL) in children with congenital heart disease (CHD) is sparse. Therefore, we investigated parental QoL, as well as its determinants, including potential association with children's own QoL.

**Methods:** 195 patients with CHD ( $13.7 \pm 2.5$  years, range 9–20 years; 55% females) and their parents ( $43.3 \pm 4.8$  years, range 29–59 years; 79% females) were investigated. Both parental and patients' QoL were measured using Taiwanese version of Quality of Life questionnaire of the World Health Organization. Personality traits and psychological characters were assessed by the Maudsley Personality Inventory and the Brief Symptom Rating Scale, respectively.

**Results:** Physical, psychological, and social domains of QoL were similar between parents and the general population. Regardless of maternal ages, their satisfactions with the environmental health were even better than those in the general population ( $P < 0.01$ ). After converting each domain scores of parental QoL into the z scores based the general norms, 37% of parents had negative sum scores. Although linear correlation between parental and children's QoL sum score was weak ( $r = 0.317$ ,  $P < 0.001$ ), we

found that all 4 domains of children's QoL were significantly lower in parents whose QoL sum scores were negative. In stepwise multivariate linear regression analysis, parental QoL in each domain was largely associated with various underlying personality and psychological characters. However, children's level of environmental health positively linked to all 4 domains of parental QoL.

**Conclusions:** QoL in parents of children with CHD was preserved. Moreover, mothers had extraordinarily higher satisfactions with the environmental health, possibly related to broad coverage of Taiwan's universal national health insurance and high accessibility of health care facilities. Children's QoL were associated with parental QoL, with children's environmental health being the most important aspect of QoL related to parental QoL.

#### PW2-11

##### **Integrated analysis of atrio-ventricular interaction in tetralogy of Fallot**

Riesenkampff E. (1), Mengelkamp L. (1), Mueller M. (2), Kropf S. (3), Abdul-Khaliq H. (2), Sarikouch S. (4), Beerbaum P. (5), Hetzer R. (1), Steendijk P. (6), Berger F. (1), Kuehne T. (1)

German Heart Institute Berlin, Berlin, Germany (1); Universitätsklinikum des Saarlandes, Homburg/Saar, Germany (2); Otto-von-Guericke-University Magdeburg, Magdeburg, Germany (3); Medizinische Hochschule Hannover, Hannover, Germany (4); King's College London, London, United Kingdom (5); Leiden University Medical Center, Leiden, The Netherlands (6)

**Introduction:** The left atrium is known to play an important role in cardiac performance. However, data about right atrial function in TOF patients are sparse. This study was undertaken to evaluate atrial function and atrio-ventricular interaction in adolescent and adult patients with repaired tetralogy of Fallot (TOF).

**Methods:** Twenty patients who had undergone surgical repair of TOF and 7 controls were investigated. The patients had residual pulmonary but no major tricuspid valve insufficiency. Atrial and ventricular strain-rates were obtained by echocardiographic speckle tracking. Cine MRI derived volumetric analysis provided atrial and ventricular time volume and time volume change curves yielding emptying and filling parameters. In addition, on the atrial level, reservoir, conduit and pump function and cyclic volume change were calculated. On the atrio-ventricular-valve level, tricuspid and mitral annular plane systolic excursion (TAPSE and MAPSE) were measured by two-dimensional echocardiography.

**Results:** In the patients, compared to controls, right ventricular enddiastolic volumes were increased and biventricular ejection fraction decreased (all  $p < 0.05$ ). Biventricular measures of early diastolic ventricular filling were at control levels, but in late diastole, right ventricular filling parameters and strain-rates were decreased ( $p < 0.001$ ). Maximal right atrial size was slightly but not significantly diminished, but cyclic volume change was significantly reduced ( $p < 0.0001$ ). Pump and reservoir function were decreased ( $p < 0.05$ ), conduit function elevated ( $p < 0.001$ ). Left atrial function was not quite normal, with reduced reservoir function and cyclic volume change ( $p < 0.05$ ). TAPSE and MAPSE were also decreased ( $p < 0.05$ ). There were statistically significant interdependencies between RV ejection fraction, TAPSE and right atrial filling and emptying parameters (all  $p < 0.05$ ).

**Conclusions:** In TOF patients, moderate systolic and diastolic right ventricular dysfunction is associated with clearly impaired right atrial function. The left atrium is also affected but to a lesser extent.

**PW2-12****Fibroblasts support functional integration of purified embryonic stem cell-derived cardiomyocytes in avital myocardial tissue**

Khalil M. (1,2), Xi J. (2,3), Spitkovsky D. (2), Hannes T. (1), Pfannkuche K. (2), Bloch W. (4), Āarić T. (2,5), Brockmeier K. (1), Hescheler J. (2,5) and Pillekamp (2,6)

*Pediatric Cardiology, University of Cologne, Cologne, Germany (1); Institute for Neurophysiology, University of Cologne, Cologne, Germany; (2); Department of Physiology and German-Chinese Stem Cell Center, Tongji, Medical College, Huazhong University of Science and Technology, Wuhan, Hubei, China(3); Department of Molecular and Cellular Sports Medicine, German Sport University, Cologne, Germany (4); Center for Molecular Medicine Cologne, University of Cologne, Cologne, Germany (5); Department of Paediatric Cardiology, Children's Hospital, Heinrich-Heine-University of Duesseldorf, Duesseldorf, Germany (6)*

**Introduction:** Transplantation of pluripotent stem cell-derived cardiomyocytes (ES-CMs) into damaged myocardium might become a therapy to improve contractile function after myocardial infarction. However, current knowledge on the mechanisms of cell integration and processes of physiological reconstitution as well as mechanical and electrical coupling after transplantation into the host tissue is still fragmentary. Undifferentiated cells remain after *in vitro* differentiation and bare the risk of tumorigenicity when transplanted. This could be overcome by lineage-selection of differentiated cells. Puromycin-selection of ES-CMs gives rise to highly purified populations without tumor-forming progenitors. Previous own experiments suggested that purified murine ES-CMs did not integrate as well as the non-purified human ES-CMs. There is cumulating evidence reporting beneficial effects of cell transplantation strategies combining a source for CMs with other cell types. From these observations we hypothesized that non-myocytes might be necessary for integration.

Aim of this study was to investigate whether murine embryonic fibroblasts (MEF) support the functional integration of purified embryonic stem cell-derived cardiomyocytes ES-CMs.

**Methods:** Neonatal murine ventricular tissue slices were subjected to irreversible simulated ischemia using oxygen and glucose deprivation. Vital tissue slices served as control. Vital and avital tissue slices were co-cultured with puromycin-selected ES-CM clusters with or without MEF. Integration of ES-CM clusters was assessed morphologically by immunohistochemistry and electro-microscopy, functional integration by isometric force measurements. Migratory activity of ES-CMs with and without MEF was analyzed by time lapse microscopy.

**Results:** ES-CMs without MEF showed no motility at all whereas ES-CMs with MEF demonstrated a high motility with a velocity of  $10.30 \pm 1.41 \mu\text{m/h}$ . The mobility of the ES-CMs was caused by the attachment to the moving MEF. We observed that ES-CM clusters integrated morphologically into vital but poorly into avital slices. Adding MEF to the co-cultures improved morphological integration into irreversibly damaged slices and enabled purified ES-CMs to confer force.

**Conclusion:** We conclude that non-cardiac cells like MEF support morphological integration and force transmission. The adhesion of ES-CMs to fibroblasts and the subsequent enablement of migration could be an additional mechanism of improved engraftment.

**PW3-1****Isolated Coarctation of the Aorta: experience in 100 consecutive patients**

Ferreira Martins J.D. (1), Costa G. (1), Gouveia S. (1), Paramés F. (1), Freitas I. (1), Trigo C. (1), Fragata J. (2), Pinto F. (1).

*Pediatric Cardiology Service, Hospital de Santa Marta, Lisbon, Portugal (1); Cardiothoracic Surgery Service, Hospital de Santa Marta, Lisbon, Portugal (2)*

**Introduction:** Coarctation of the aorta (CoA) represents 5–8% of congenital heart disease. The treatment provides excellent immediate results, but a persistence of significant residual problems.

**Objective:** To describe the presentation, treatment and evolution in long term of a unselected population of about 100 consecutive patients with Isolated CoA followed in a single center.

**Methods:** Retrospective study of all patients with Isolated CoA treated during the past 21 years (1987–2008).

**Results:** Patients (n = 100, 68% male) were diagnosed with a median age of 94 days (1 day to 16 years). Clinical presentation differed between patients with age under and upper a year, the first having cardiac failure and the later being asymptomatic and with evidence of arterial hypertension (88 and 63%, respectively;  $p < 0.01$ ). Therapy performed in median 8 days after diagnosis, was surgical in 79 cases (20 end-to-end anastomosis, 31 sub-clavian flaps, 28 patch plasty) and percutaneous in the remaining 21 (15 balloon angioplasty, 6 stent implantations). Mean age of surgical patients was lower than in percutaneous (3.4 vs 7.5;  $p < 0.01$ ). Immediate mortality was 2% and occurred in two newborns, in the surgical group. There was no late mortality, with a mean follow-up of  $7.2 \pm 5.4$  years. Re-coarctation was found in 8 patients (6 surgical, 2 percutaneous). 46 patients have hypertension (19 in rest, 27 with effort), with a median age of diagnosis later than the others (23 vs 995 days;  $p < 0.01$ ).

**Conclusion:** Isolated CoA is a disease with excellent short term prognosis but significant long term complications that should not continue to be regarded as a simple obstruction of descendent aorta, rather as a complex pathology that requires close follow-up. The semiologic variability and the potential insidious presentation, warrants a high clinical suspicion, namely an accurate clinical examination, that includes the palpation of pulses in lower limbs. The delay on diagnosis and treatment has significant impact in late cardiovascular morbidity and mortality. The ultimate choice of the treatment technique depends of the age of the patient, associated findings and the experience of medico-surgical team. Special relevance must be given to hypertension in the follow-up of these patients, its risk factors and complications.

**PW3-2****Catheter Ablations in Children with Ventricular Tachycardia**

Pokushalov E., Romanov A., Artemenko S., Shugaev P., Tiurov A., Shirokova N.

*State Research Institute of Circulation Pathology, Novosibirsk, Russian Federation*

**Introduction:** The aim of this study was to expand data on the outcomes of catheter ablation of ventricular tachycardia (VT) in young patients and to identify obstacles to success.

**Methods:** In this study 143 children with monomorphic VT (mean age  $13.8 \pm 2.9$  years, ranging from 6 to 18 years) who underwent catheter ablation for different VT were observed.

**Results:** The structure of the arrhythmias was as following: right ventricular outflow tract tachycardia (RVOT; 24%), aortic sinus of valsalva (27%), arrhythmogenic right ventricular cardiomyopathy (ARVC; 14%), left VT (34%). VT originating from the OT: septum of the RVOT (41%), free wall of the RVOT (5%), near the His-bundle region in the RVOT (12%), endocardium of the LVOT (15%), left sinus of Valsalva (24%), LV epicardium

remote from the LSV (3%). During  $13 \pm 7$  months of the follow-up period after a single ablation procedure, the results were as follows: RVOT ablation ( $n = 50$ ) – 64% successful, 36% – unsuccessful, 8% showed recurrence of VT; LVOT ablation ( $n = 24$ ) – 83% successful, 17% – unsuccessful, 4% – recurrence of VT. The second ablation procedure (RVOT+LVOT) was performed in 22 patients. In  $16 \pm 9$  months after the last ablation procedure the success rate was 96%. Idiopathic left fascicular VT. During  $11 \pm 8$  months of the follow-up period after a single ablation procedure the results were as follows: ventricular activation guided ablation ( $n = 21$ ) – 33% successful, 67% – unsuccessful, 19% showed recurrence of VT; P potential guided ablation ( $n = 28$ ) – 71% successful, 29% – unsuccessful, 10% – recurrence of VT. The second ablation procedure (ventricular activation+P potential guided ablation) was performed in 22 patients. In  $9 \pm 8$  months after the last ablation procedure the success rate was 88%. ARVC. During  $8 \pm 7$  months of the follow-up after a single ablation procedure ( $n = 21$ ; endocardial ablation) – 91% of the procedures were successful, 9% – unsuccessful, 9% – showed recurrence of VT. The second ablation procedure (epicardial ablation) was performed in 5 patients. In  $7 \pm 6$  months after the last ablation procedure the success rate was 60%. **Conclusion:** Catheter ablation can be effective, safe, and life saving technique for children with medically resistant VT.

### PW3-3

#### Transseptal access to the left atrium following atrial septal device closure – feasibility and force needed with 3 different permanent and one absorbable closure device

Jux C. (1), Kröll M. (2), Steinmetz M. (2), Michaelis I. (3), Quentin T. (2)

Pediatric Heart Centre, Justus Liebig University, Giessen (1) and Dept. of Pediatric Cardiology University of Göttingen (2), IVK RWTH Aachen (3)

**Background:** An increasing number of transcatheter septal device closures are performed to treat patients with atrial septal defects (ASDs) and patent foramen ovale (PFOs) for hemodynamic reasons or for the prevention of paradoxical embolism. Some of these patients may require transseptal access later on for diagnostic or therapeutic electrophysiologic studies or left sided heart valve repair. We accessed for the first time the ability to gain transseptal access through 4 different well healed-in atrial septal closure devices in an in vivo model.

**Methods:** 3 different permanent (Amplatzer, Helex, Starflex) and 1 biodegradable (Biostar) septal closure devices were implanted in 8 sheep. Following a healing phase of 2 years transseptal access was aimed for by use of a Brockenborough transseptal needle as well as the Bayliss transseptal radiofrequency system. Thereafter, the force needed to perform a transseptal needle puncture was measured quantitatively for each device under defined conditions using an industrial dynamometer ex vivo.

**Results:** Transseptal access through a healed-in closure device was possible in 3/10 (Amplatzer), 2/10 (Helex), 3/10 (Starflex) and 8/10 (Biostar) attempts respectively. Radiofrequency perforation of the device was achieved only with the Biostar device (9/10 attempts) and failed in all other devices. The force to cross a healed-in device with a transseptal needle measured between 14 and 2.42 N (compared to  $1.57 \pm 0.6$  N in native atrial septa) and was significantly lower with the absorbable device compared to the permanent devices.

**Conclusion:** Healed-in permanent atrial septal closure devices are difficult to cross using a Brockenborough transseptal needle and virtually impossible to cross using a radiofrequency transseptal system. Thus, the current advice is to puncture the septum away

from the device which may result in an inappropriate angle towards the target structure or be difficult and more dangerous with larger devices. Bioresorbable devices allow for direct and easier transseptal access, should this be necessary during later life of the patient.

### PW3-4

#### Simplified 3-lead ECG as screening tool for LQT syndrome in children

Germanakis I., Androutsopoulou G., Perakaki R., Kakouri P., Kalmanti M.

Dpt of Pediatrics, University Hospital Heraklion, Crete, Greece

**Introduction:** As the first manifestation of LQT syndrome in children might be sudden cardiac death, a low cost ECG screening system that allows for reliable QTc interval measurement could represent a life saving screening tool. Aim of the study was to evaluate the accuracy of QTc estimation using a simplified 3-truncal lead ECG system compared to a standard 12 lead ECG system, in a population of school aged children.

**Methods:** Study population included a random sample of 8-year old children ( $n = 69$ , 41 boys, 28 girls) participating in a pilot screening survey of cardiovascular health among school age children (CPCS), approved by the Greek Ministry of Education. Participant children undergo both a standard 12-lead ECG as well as a digital phonocardiogram incorporated with a simplified 3-lead ECG recording. Simplified truncal ECG leads are placed on the right and left distal subclavian area and lower left abdomen, providing a single ECG tracking line as time reference for phonocardiographic data. A single experienced observer measured the QTc interval (Bazet formula) as the average of three consecutive heart cycles for each patient both a) on the printout of the original 12 ECG recording (lead II) b) on a laptop PC screen by viewing the digitally stored simplified ECG and using appropriate time calipers (QTcII and QTc3, respectively). Paired-T test was used to compare mean QTcII and QTc3 values.

**Results:** The mean QTcII was  $408.6 \text{ sec}^{-1}$  (SD 21.8, median 406 range 366–457) while mean QTc3 was  $409.7 \text{ sec}^{-1}$  (SD 18.4, median 410.6, range 377–468) (Fig. 1). The mean QTc difference (QTcII–QTc3) was  $-1.01 \text{ sec}^{-1}$  (95% C.I.: -5,5 to 3,4) and not statistically significant ( $t = -0,45$ ,  $df = 68$ ,  $p = 0,654$ ).

**Conclusions:** QTc estimation based on simplified 3-lead ECG compares well with QTc estimate based on standard 12-lead ECG recording. Incorporation of low cost 3-lead-ECG devices in cardiovascular screening programs could allow for a wide and cost effective screening for LQT syndrome in the school aged children.

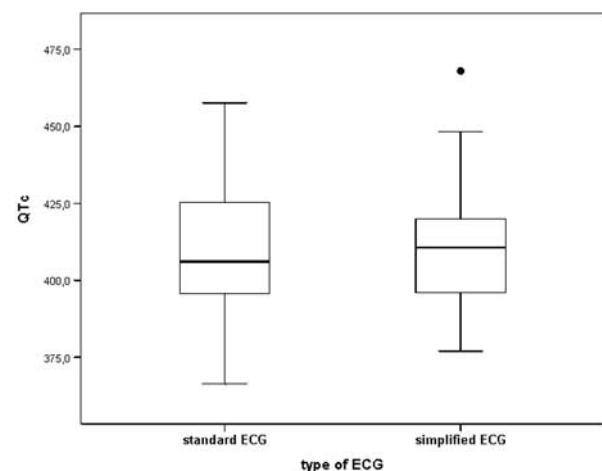


Fig 1. Standard and simplified ECG QTc values

## PW3-5

**The Importance of the Level of Maternal Anti-Ro/SSA Antibodies as a Prognostic Marker of the Development of Cardiac Neonatal Lupus Erythematosus: A Prospective Study of 186 Antibody-Exposed Fetuses and Infants**

Jaeggi E. (1), Laskin C. (2), Hamilton R.M. (1), Kingdom J. (2), Silverman E. (1)

The Hospital for Sick Children, Toronto, Canada (1); Mount Sinai Hospital, Toronto, Canada (2)

**Background:** Anti-Ro/SSA and anti-La/SSB autoantibody-positive mothers are frequently referred for serial echocardiography because of the elevated fetal risk of developing immune-mediated complications such as heart block. Little is known why only some and not all offspring are affected. The aim of this study was to determine if cardiac complications of neonatal lupus erythematosus (NLE) are related to maternal autoantibody-levels rather than their presence.

**Methods:** All cases referred since 2000 to our cardiac tertiary care center for serial fetal echocardiography or cardiac complications related to maternal antibodies were included. Patients without (group 1) and with (group 2) cardiac NLE were compared. Antibody-levels were measured by solid-phase enzyme-linked immunosorbent assay (ELISA) using recombinant human 60-kD Ro, 52-kD Ro and 48-kD La proteins (Phadia; Germany). The cut-off for a positive test result in our laboratory was 8 units/ml. **Results:** Group 1 included 146 serially screened fetuses with normal pregnancy outcomes. Group 2 consisted of 40 cases with fetal (N = 34) or neonatal (N = 6) diagnosis of heart block, endocardial fibroelastosis or both, and included 4 fetuses diagnosed during serial screening. All cardiac complications were associated with moderate (>50 U/ml; 15%) or high (>100 U/ml; 85%) maternal anti-Ro levels, independently of anti-La antibody titres. The event rate of complete heart block was 5% for prospectively screened fetuses with Ro-values >50 U/ml (odds ratio 7.8) and 0% for those fetuses with lower titres ( $p < 0.0001$ ). Infants with prenatal exposure to high-titre anti-La levels >100 U/ml were the most likely to develop non-cardiac features of NLE (event rate 57%; odds ratio 4.7).

**Conclusions:** Our findings support that the amount of maternal antibodies rather than their presence is associated with fetal tissue injury. As anti-Ro levels correlates with the risk of cardiac complications, serial echocardiography should be limited to those women with high anti-Ro-titres.

## PW3-6

**Randomized controlled trial comparing two heparinization protocols for prophylaxis of vascular thrombosis during cardiac catheterization in children – the HEARTCAT study**

Hanslik A. (1), Kitzmüller E. (1), Mlekusch W. (2), Salzer-Muhar U. (1), Michel-Behnke I. (1), Male C. (1)

Division of Pediatric Cardiology, Department of Pediatrics, Medical University of Vienna, Austria (1); Department of Internal Medicine, Medical University of Vienna, Austria (2)

**Introduction:** Thrombotic complications are serious complications of cardiac catheterization (CC) in children. Clinical signs of arterial and venous thrombosis are unreliable in children but few studies have used objective ultrasound testing. Moreover, only few studies have assessed unfractionated heparin (UFH) protocols for prophylaxis of CC-related thrombosis. Therefore, the optimal UFH dose for CC in children is not known.

**Objective:** To (i) describe the incidence of thrombosis during CC in children using ultrasound assessment, and (ii) compare a high-dose versus a low-dose UFH protocol for thromboprophylaxis. **Methods:** Randomized controlled trial comparing high-dose UFH (100 u/kg bolus, followed by 20 u/kg/h continuous infusion) versus low-dose UFH (50 u/kg bolus) during elective CC in children with congenital heart disease at a tertiary care pediatric cardiology centre. Outcome assessment was by i) documentation of clinical signs of thrombosis and ii) vascular ultrasound performed before and within 48 hours after CC. Patients without consent for randomization received standard of care heparin, but received the same outcome assessment and were followed in a cohort study (cohort study).

**Results:** 201 children were included in the study (median age 5.6 years, 23% infants, 39% females). Ninety-eight patients (49%) underwent interventional CC. Overall, 9 (5%) patients developed thrombosis (6 (3%) arterial, 3 (2%) deep venous); 5 patients had other vascular complications (3 arteriovenous fistulae, 2 pseudoaneurysm). Arterial thrombosis occurred in infants only, whereas deep venous thrombosis was only seen in older children. Minor bleeding at puncture site occurred in 13 (7%) patients, and only 1 patient needed red cell transfusion. Of 201 patients, 120 children were randomized to one of the two heparinisation protocols (58 low-dose UFH, 62 high-dose UFH). There was no significant difference in incidence of vascular thrombosis or minor bleeding between the two groups.

**Conclusions:** Incidence of thrombosis during CC in children was 5% using ultrasound screening. Infants were at particular risk for arterial thrombosis. The incidence of bleeding was 7% which was mostly minor. High-dose UFH was not superior to low-dose UFH for prophylaxis of cardiac-catheterization associated vascular thrombosis.

## PW3-7

**NT-pro-brain natriuretic peptide (N-BNP) reflects strongly left ventricular volume load before and after PDA closure in children and adolescents.**

Mueller G.C. (1), Mir T.S. (1), Arndt F. (1), Samson-Himmelstjerna P. (2), Dodge-Khatami A. (2), Weil J. (1)

Department of Pediatric Cardiology, University Heartcenter Hamburg, Germany (1); Department of Pediatric Cardiac Surgery, University Heartcenter Hamburg, Germany (2)

**Introduction:** The role of plasma N-BNP levels in screening and treatment of persistent ductus arteriosus (PDA) in preterm neonates are well reported. Less is known about the value of cardiac natriuretic peptides during interventional closure of relevant PDA in children and adolescents. The purpose of this study was to evaluate cardiac natriuretic peptides as a parameter of left ventricular volume load during closure of isolated PDA.

**Methods:** Thirty children aged 2 to 17 years were studied during transvenous closure of PDA by Amplatzer Ductal Occluder. N-BNP and pro-atrial natriuretic peptide (pro-ANP) levels were measured before, 5 min after and 24 hours after successful PDA-closure. Intracardiac dimensions were measured by echocardiography (GE Vivid 3) before and 24 hours after PDA-closure.

**Results:** Pro-ANP concentrations increased significantly at 5 min after PDA-closure (mean  $\pm$  SD; pre 56  $\pm$  30 vs. 5 min post 81  $\pm$  71 pg/ml;  $p < 0,05$ ), whereas N-BNP concentrations did not change (pre 260  $\pm$  378 vs. 5 min post 255  $\pm$  445 pg/ml;  $p = 0,86$ ). At 24 hours after PDA closure pro-ANP and N-BNP concentrations decreased significantly compared to preclosure and directly postclosure concentrations (pro-ANP 24h post 29  $\pm$  15;  $p < 0,05$ ; N-BNP 24h post

123 ± 200 pg/ml;  $p < 0,05$ ). Concerning left atrial and ventricular dimension, LA/Ao-ratio decreased significantly within 24 hours after PDA closure (mean 1,44 vs. 1,23;  $p < 0,05$ ), whereas left ventricular dimensions did not change early.

**Conclusion:** For the first time we describe N-BNP as a stable parameter during interventional closure of isolated PDA in children and adolescents. In contrast to stable N-BNP pro-ANP showed an increase temporarily due to interventional PDA-closure. In the early postinterventional period N-BNP reflects significantly left ventricular volume load reduction and downsizing of left atrial diameters.

### PW3-8

#### **Complications after pulmonary artery stenting in patients with hypoplastic left heart syndrome (HLHS) – what to be afraid of.**

*Moszura T. (1), Mazurek-Kula A. (1), Dryżek P. (1), Górczynski S. (1), Moll JA. (1), Moll JJ. (2), Sysa A. (1)*

*Cardiology Department Polish Mother's Memorial Hospital, Research Institute Lodz, Poland (1); Cardiosurgery Department Polish Mother's Memorial Hospital, Research Institute, Lodz, Poland (2)*

**Background:** Stenosis or hypoplasia of pulmonary arteries are frequent during multistage treatment of HLHS. The mechanism of hypoplasia and stenosis of pulmonary arteries after first stage Norwood operation is connected with changes in geometry of pulmonary arteries and significantly deteriorates prognosis in this group of patients. Use of interventional techniques is a method of choice in pulmonary arteries stenosis treatment in HLHS. Balloon angioplasty is efficient only in patients with short-distance pulmonary stenosis and totally ineffective in the group of children with tubular stenosis or pulmonary hypoplasia. This group of patients requires intravascular stent implantation. Good long-term results and possibility of redilatation of previously implanted stent allows for use of these techniques in children.

**Material:** In our material pulmonary arteries stenosis and hypoplasia remains to be a crucial complication in HLHS treatment. Implantation of 93 stents in 87 patients with HLHS and pulmonary arteries stenosis or hypoplasia resulted in pulmonary pressure reduction and increase of vessel diameter.

We present three types of severe complications after stent implantation in our 5 patients:

- I. two patients with bronchial compression.
- II. one patient with compression of native ascending aorta with coronary circulation deterioration resulted in acute myocardial ischemia with circulatory arrest (required resuscitation and inotropic drugs infusion).
- III. two patients with stent fracture and dislocation (in one case after cardiac massage in second one due to local stress connected with change of geometry of LPA due to Sano anastomosis).

**Methods and results:** In group I we performed successful balloon plasty of left bronchus. Control bronchography revealed unconstrained contrast flow through the extended bronchus. In group III the two parts of fractured and dislocated stents were stabilized and connected by implantation of an additional CP stent.

**Conclusions:** We conclude that tracheobronchial obstruction, native aorta compression and stent fracture and dislocation might be rare and unexpected adverse effects of endovascular stent placement in children with HLHS. In our cases balloon plasty of bronchial obstruction was an efficient emergency therapeutic option. If the pulmonary stent implantation or redilatation is necessary computed tomography might be very useful to show potential collision of thoracic structures.

### PW3-9

#### **Diagnostic and interventional percutaneous catheterization in the early postoperative period (≤30 days) after congenital heart surgery in children – a single-centre experience**

*Kretschmar O. (1), Balmer C. (1), Prêtre R. (2), Dave H. (2), Knirsch W. (1)*

*University Children's Hospital Zurich, Department of Pediatric Cardiology (1); and Cardiothoracic Surgery (2), Zurich, Switzerland*

Catherizations in the early period after congenital heart surgery (CHS) are considered to be associated with an elevated rate of morbidity and mortality due to the hemodynamic instability and vulnerability of these patients. Therefore, they are often avoided or delayed. The purpose of our study was to evaluate the feasibility and safety of diagnostic and interventional percutaneous procedures in this critical period (≤30 days) after CHS in children. Method: Retrospective evaluation of all early postoperative catheterizations in our institution between 01/2001 and 12/2009. Patients: We performed in total 140 catheterizations – 134 diagnostic and 66 interventional procedures. Patients age ranged from 1 day to 18.8 (median 0.3) years, their weight from 2.5 to 70 (median 4.7) kg. 80 patients (57%) were male. Catheterizations were performed between 0 and 30 (median 11) days after CHS. Previous surgery was partial/total cavopulmonary-anastomosis (33.6%), shunt- or Norwood/hybrid procedures (18.6%), implantation of a pulmonary graft (12.9%), arterial switch-operation (8.6%), repair of coarctation (5.7%), and others.

**Results:** Indications for catheterization were prolonged postoperative course (25%), decrease of saturation (22.1%), ECG-changes (13.5%), elevated pulmonary-arterial pressure (12.1%), assumed re-coarctations (5%), and others. 4 patients were catheterized under extracorporeal mechanical support. 81 diagnostic procedures (60.4%) revealed a significant pathology leading to a direct catheter-intervention ( $n = 60$ ) or early redo-surgery ( $n = 21$ ). Interventions ( $n = 86$ , 66 patients) included stent-implantations/balloon-dilatations in pulmonary arteries (25.6%) or (re-)coarctations (10.5%) or restrictive ASDs (10.5%), coil-embolizations of aortopulmonary (11.6%) and venovenous collaterals (9.3%), manipulations of shunts and fenestrations (12.8%), and others. Immediate success rate was high (97%). Complications (arrhythmias, vessel thrombosis, pneumothorax, intracerebral bleeding) occurred in 12 patients (8.6%). 30-day survival rate after catheterization was 95%.

**Conclusion:** If indicated, diagnostic catheterizations can be performed safely and with a high diagnostic value in the early postoperative course. Postoperative decrease of saturation had the highest predictive value for a postoperative problem needing acute treatment (68%). Transcatheter interventions can be successfully performed leading to a significant and lasting improvement of these critically ill patients. Therefore, diagnostic and interventional catheterizations should not be withheld from the patients at any time after CHS. An experienced multidisciplinary team approach is needed to ensure the successful realization of these procedures.

### PW3-10

#### **First experience with the epicardial application of a transvenous 4.1 Fr lumenless pacemaker lead in patients with congenital heart disease**

*Peters B., Miera O., Ewert P., Hacke P., Alexi-Mekishvili V., Stamm C., Huebler M., Hetzer R., Berger F.*

*German Heart Institute Berlin, Germany*



**Introduction:** To date opinion on the best approach for pacemaker implantation in small children is favouring epicardial implantation. In addition intraoperative placement of epicardial pacing leads during corrective surgery in patients with congenital heart disease (CHD) is well established. Certain conditions like high tissue resistance to electrical current (multiple pre-operated patients or difficult accessibility may lead to failure of standard epicardial leads. Due to its favourable and unique construction characteristics like short tip-to-ring spacing, low electrode surface polarization, extremely slim lumenless design (4.1 Fr) and superior tensile strength we chose the recently developed transvenous model 3830 (Medtronic Inc. MN, USA) as an alternative candidate. We therefore report our first experience using this lead in an epicardial manner in patients with CHD.

**Methods:** The surgical approach was either left or median thoracotomy. For ventricular pacing a small incision into the myocardium of the desired pacing site was done to house the electrode tip. For atrial placement, the electrode was screwed into the left atrial appendage (LAA) after direct puncture, secured by a purse-string suture or directly screwed tangentially into the surface of the LAA and fixed with sutures.

Implantation		
	Threshold	Sensing
atrial n = 5	1.1 ± 0.8 V (0.8–2.7)	2.1 ± 1.9 mV (1.1–4.3)
ventricle n = 8	1.25 ± 1.07 V (0.9–3.7)	12.5 ± 8.2 mV (5.1–30.0)
Last Follow up		
	Threshold	Sensing
atrial n = 5	0.5 ± 0.48 V (0.25–1.5)	4.0 ± 2.9 mV (1.0–8.0)
ventricle n = 8	1.25 ± 1.49 V (0.25–5.0)	11 ± 4.0 mV (4.0–15.3)

**Results:** 13 Electrodes were implanted in 10 patients whenever no acceptable pacing parameters with conventional bipolar epicardial leads could be achieved. Initial pacing thresholds were significant lower than with standard epicardial leads. Thresholds remained stable in all cases (follow up 83 d median, range 4–472 d) except for 2 leads with significant increasing values (see table); in one of those, the electrode later had to be explanted due to infection of the system.

**Conclusions:** Our results demonstrate that the 3830 lumenless pacing lead may be effectively serve as epicardial pacing lead even under unfavourable conditions for standard epicardial lead placement ("rescue treatment"). The slim electrode design combined with an extreme mechanical stability is advantageous compared to standard epicardial designs especially in growing children. Further adjustment of the electrode, e.g. shorter tip-to-ring distance, availability in shorter lengths and the development of dedicated application mechanisms might establish this electrode as a primary choice in epicardial lead placement. Of course, good mid- and long term performance has to be proven.

### PW3-11

#### Long-term survival after paediatric cardiac surgery for congenital heart defects

Sunnegårdh J.T., Berggren H., Synnergren M.

The Queen Silvia Children's Hospital, Sahlgrenska University Hospital, Göteborg University, Sweden

**Objective:** To gather reliable and complete data for evaluation of long-term survival after paediatric cardiac surgery.

**Methods:** All 1822 patients (835 girls/987 boys) operated due to congenital heart defects in our institution before the age of 18 years

from Jan 1st 1994 to Jan 1st 2005 were included. During the study period nearly 50% of all paediatric cardiac surgery in Sweden was performed at our institution. Patient files were cross-checked as of Jan 1st 2009, against the National Population Registry in Sweden, allowing for reliable and complete data on long-term survival.

**Results:** Median age at surgery was 0.36 years (0–17.8 years). 1591 patients (87%) had surgery for biventricular correction and 231 patients (13%) had univentricular heart palliations. Median age of survivors at follow up was 12.1 years (4–31.6), and median follow up time after first surgery was 10.4 years (4–30.4). 144 deaths (7.9%) occurred with a median age at death of 0.5 years (0–23.3). Median survival time in the deceased patients was 36 days (0–13.5 years) after the last major surgical procedure. 81 deaths (5.1%) occurred after surgery for biventricular correction and 63 (28.2%) after palliation for univentricular heart defects ( $p < 0.001$ ). 31/59 (52.5%) patients died after Norwood surgery for classic hypoplastic left heart syndrome compared to 32/172 (18.6%) after surgery for other univentricular heart defects ( $p < 0.001$ ). In children with univentricular heart palliation (classic hypoplastic left heart syndrome included) being born 1994–1999 and 2000–2004 respectively, mortality was 42/114 (36.8%) and 18/82 (21.9%,  $p < 0.05$ ). 17 patients had a heart transplantation of whom 7 (0.4%) had previous biventricular and 10 (4.3%) univentricular heart surgery.

**Conclusions:** Total survival was 92.1%, with most deaths occurring later than 30 days after the last major surgical procedure, reflecting the need for long-term follow up. Total survival after surgery for biventricular correction was 94.9%, after Norwood surgery in classic hypoplastic left heart syndrome 44.5% and in all other types of univentricular heart patients 81.4%. Survival after univentricular heart surgery improved significantly over time.

### PW3-12

#### Device Occlusion of Atrial Septal Defects (ASD) Without Balloon Sizing: Results of a Multicenter Registry

Radtke W.A.K. (1), Holzer R. (2), Owada C. (3), Bandisode V.M. (4), Lim S (5), Everett A (6)

Nemours Cardiac Center, Wilmington, DE, USA (1); Nationwide Children's Hospital, Columbus, OH, USA (2); Children's Hospital of Central California, Madera, CA, USA (3); MUSC Children's Hospital, Charleston, SC, USA (4); University of Virginia, Charlottesville, VA, USA (5); Johns Hopkins University, Baltimore, MD, USA (6)

Device occlusion of atrial septal defects has become a routine procedure. In the process, some operators have omitted balloon sizing from their standard protocol. We have reviewed our multicenter database to determine the safety and efficacy of this approach.

All ASD device occlusions submitted from the 16 institutions to the datacenter from April 2004 until December 2009 were included in the analysis. The de-identified data include patient age and weight, diagnoses, procedural parameters, complete hemodynamics, and pertinent information on attempted and accomplished ASD occlusion, complications and immediate results. We analyzed 910 such procedures with patient ages ranging from 1 month to 83 years (median 6 yrs). Patient weight ranged from 2.3 to 160 kg (median 22 kg) and defect diameter from 1 mm to 55 mm by transesophageal or intracardiac echocardiogram. Balloon sizing was not performed in 235 procedures: these patients were younger (mean 10.6 versus 16.6 yrs;  $p < 0.0001$ ), weighed less (mean 28.0 versus 40.5 kg;  $p < 0.0001$ ) and had slightly smaller ASDs (mean 11.4 versus 13.3 mm;  $p < 0.0001$ ). Three centers accounted for the procedures

without balloon sizing. Fluoroscopy time was slightly shorter when no balloon sizing was performed (mean 15.0 versus 18.9 min;  $p < 0.0001$ ). Concomitant additional interventions were included in the fluoroscopy time which for the entire cohort ranged from 1.9 min to 102 min with a median of 14.2 min. Omission of balloon sizing had no negative impact on complication rates, in particular there was only one device embolization versus four in the balloon sizing group. Also, residual shunt appeared less frequent without balloon sizing ( $p = 0.011$ ). Our retrospective analysis suggests that ASD device occlusion can be performed safely and effectively without balloon sizing.

#### PW4-1

##### **Systolic right ventricular (RV) function in pediatric patients and adolescents with tetralogy of Fallot (TOF): Tricuspid annular plane systolic excursion (TAPSE), tricuspid annular peak systolic velocity (TAPSV), and RV ejection fraction (EF) in comparison to age-matched normal subjects**

Koestenberger M. (1), Nagel B. (1), Stueger HP (2), Ravekes W. (3), Everett AD. (3), Heinzl B. (1), Sorantin E. (4), Cvirn G. (5), Gamillscheg A. (1)

Division of Pediatric Cardiology, Department of Pediatrics, Medical University Graz, Austria (1); Data, Statistics & Risk Assessment; Austrian Agency for Health and Food Safety, Graz, Austria (2); Division of Pediatric Cardiology, Johns Hopkins University School of Medicine, Baltimore, MD, USA (3); Division of Pediatric Radiology, Department of Radiology, Medical University Graz, Austria (4); Institute of Physiological Chemistry, Centre of Physiological Medicine, Medical University Graz, Austria (5)

**Objectives:** The TAPSE and the TAPSV have not been thoroughly investigated in pediatric patients and adolescents with TOF following transannular patch repair in early childhood.

**Methods:** TAPSE and TAPSV values were prospectively determined in 105 patients (0–28 years; 58 male; 47 female) with TOF, and 643 age-matched normal subjects. TAPSE values were compared to RVEF measured by both, MRI and echocardiography.

**Results:** The TAPSE values showed a positive correlation with age in normal subjects. A significant reduction of TAPSE and TAPSV values with an increasing time interval following corrective surgery was observed when compared to the normal subjects. After a mean of 7 years TAPSE values become significantly reduced compared to the lower bound of the –2 SD of age-matched control patients. The TAPSE and TAPSV values showed a significant positive correlation with RVEF determined by MRI and echocardiography. There was no significant difference of TAPSE or TAPSV values between male or female subjects.

**Conclusions:** We found a significant reduction of TAPSE and TAPSV values with an increasing time interval following corrective surgery of TOF. The impaired RV function could be confirmed in MRI and echocardiography measurements of RVEF and could serve as reference data.

#### PW4-2

##### **Beneficial effects of vasopressors in the treatment of experimental acute right ventricular failure induced by pulmonary artery constriction**

Apitz C. (1,4), Honjo O. (2), Assad R. (3), Humpl T. (1), van Arsdell G. (2), Redington A.N. (1)

Division of Cardiology, The Hospital for Sick Children, Toronto, Canada (1); Division of Cardiac Surgery, The Hospital for Sick

Children, Toronto, Canada (2); University Heart Institute, Sao Paolo, Brazil (3); Paediatric Heart Centre, Giessen, Germany (4)

**Introduction:** An acute increase in right ventricular (RV) afterload leads to RV dilation, reduced systolic function and low cardiac output. It has previously been shown, experimentally, that aortic constriction can reverse some of these changes, presumably via beneficial ventricular-ventricular interactions or improved myocardial perfusion. We studied the more clinically relevant effects of intravenous vasopressors on this phenomenon.

**Methods:** Adjustable banding devices were placed on the main pulmonary artery (PA) and the descending aorta respectively via sternotomy in adult new Zealand white rabbits (3.5–5 kg). Cardiac output was measured by a flowprobe placed on the ascending aorta. Acute RV failure was induced by severe PA constriction. We assessed the effect of an additional aortic constriction ( $n = 10$ ) by pressure-volume loop-analysis using load-independent indexes of global ventricular contractile performance by the end-systolic pressure-volume relationship (ESPVR). We assessed the impact of aortic constriction and the effects of 0.05, 0.1, 0.5, and 1 mcg/kg  $\times$  min<sup>-1</sup> norepinephrine and epinephrine ( $n = 5$ ) by paired t-test.

**Results:** With additional aortic constriction biventricular contractile performance increased significantly, as indicated by an increased slope of the ESPVR (RVESPVR 3.17 vs. 5.16 mmHg/ml ( $p = 0.0002$ ), and LVESPVR 13.9 vs. 18.8 mmHg/ml ( $p = 0.02$ )). There was a non-significant increase in cardiac output from 131 to 134.8 ml/min, and heart rate was unchanged. Norepinephrine and epinephrine resulted in similar effects on biventricular contractility, the most significant effect being achieved with 0.5 mcg/kg  $\times$  min<sup>-1</sup> norepinephrine (RVESPVR 2.8 vs. 4.8 mmHg/ml,  $p = 0.007$ ; LVESPVR 14.9 vs. 16.6 mmHg/ml,  $p = 0.08$ ). In contrast to aortic constriction, cardiac output was also increased, the most significant effect being achieved by 1 mcg/kg  $\times$  min<sup>-1</sup> epinephrine (143 vs. 215.7 ml/min,  $p = 0.04$ ) and 1 mcg/kg  $\times$  min<sup>-1</sup> norepinephrine (143 vs. 191 ml/min,  $p = 0.07$ ).

**Conclusions:** Aortic constriction improves biventricular contractility but does not increase cardiac output in an experimental model of acute right heart failure using PA constriction. An equipotent effect on biventricular contractility, but with additional benefit in terms of cardiac output was achieved by the use of systemic vasopressors and so they may be a useful clinical tool in acute right heart failure.

#### PW4-3

##### **Longitudinal evaluation of hepatic function profile following total cavopulmonary anastomosis**

Kaulitz R., Haber P., Sieverding L., Ziemer G., Hofbeck M.

Department of Paediatric Cardiology and Paediatric Intensive Care Medicine, Tuebingen University Hospital, Germany

Long-term consequences of chronic congestion for the developing liver in children after total cavopulmonary connection (TCPC) are still unclear. Longitudinal data on liver function during individual follow-up are not available.

**Methods:** 52 patients were assessed according to a clinical protocol  $4.1 \pm 2.1$  yrs (age  $10 \pm 4.9$  yrs) (I) and  $9.1 \pm 3.1$  yrs (age  $14.2 \pm 5.4$  yrs) (II) after TCPC (time interval mean  $5.5 \pm 1.5$  yrs). Primary diagnosis: TA 13, DIV 9, complex UH 30 pts. 9 pts (17%) developed protein-losing enteropathy (PLE), 12 pts (23%) had chronic ascites. Haemodynamic data were available in 41 pts, hepatic sonography and Doppler ultrasound in 34 pts. 17 pts (35%) received anticoagulation treatment.

**Results:** The most important laboratory findings are summarized in the table (in 79% with moderately elevated  $\gamma$ -glutamyltransferase ( $\gamma$ GT), in 13%  $>100$  U/l).

	$\gamma$ GT (U/l)	AST (U/l)	ALT (U/l)	Bili (mg/dl)	Prot (g/dl)
I (%abn)	34 $\pm$ 25 (32)	27 $\pm$ 10 (21)	20 $\pm$ 8 (2)	1 $\pm$ 0.5 (11)	7.1 $\pm$ 0.9 (7)
II (%abn)	61 $\pm$ 47 (79)	35 $\pm$ 9 (57)	31 $\pm$ 12 (17)	0.9 $\pm$ 0.5 (17)	7.2 $\pm$ 1.1 (15)

Serum protein level was normal in all pts except those with PLE; INR was slightly elevated in only 2 pts. There was no correlation of biochemical parameter to haemodynamic data except for  $\gamma$ -glutamyltransferase-level and mean pulmonary artery pressure ( $p < 0.05$ ) and enddiastolic ventricular pressure ( $p < 0.01$ ). Hepatic sonography revealed hepatomegaly in 5/34 and splenomegaly in 15/34 patients; increased echogenicity, inhomogeneity or liver surface nodularity were present in 18/34 patients with significantly longer postoperative follow-up ( $> 8$  yrs). Doppler ultrasound revealed inspiratory dependence of hepatic venous flow in 90%, antegrade portal venous flow in all patients with respiratory variability in 78% and low flow velocity ( $< 0.2$  m/s) without undulation in 3 patients with  $\gamma$ -GT-level  $> 100$  U/l.

**Conclusion:** Continuous evaluation of liver function revealed persistent elevation and further increase of  $\gamma$ -GT during long-term follow-up. There was no compromise of synthetic function and only mild signs of hepatocellular damage. We found abnormalities in portal vein flow pattern in patients with elevated  $\gamma$ -GT-level and clinical signs of ascites, respectively. Sonographic structural abnormalities occurred with increasing time interval since Fontan procedure. Hepatic morphological and functional changes will become an increasingly important aspect during long-term follow-up in these patients even in presence of favourable haemodynamic parameters.

#### PW4-4

##### Patients with Repaired Tetralogy of Fallot Undergoing Pulmonary Valve Replacement – What Happens to Tricuspid Regurgitation?

Silva M. (1), Inuzuka R. (2), Li W. (2), Tei E. (2), Dimopoulos K. (2), Gatzoulis M. (2), Anjos R. (1), Martins F. (1)

Centro Hospitalar de Lisboa Ocidental, EPE, Hospital de Santa Cruz, Serviço de Cardiologia Pediátrica, Lisbon, Portugal (1); Royal Brompton Hospital and the National Heart & Lung Institute, Imperial College, London, UK/Adult Congenital Heart Disease Centre and Centre for Pulmonary Hypertension (2)

**Introduction:** A significant number of patients with repaired Tetralogy of Fallot (rTOF) undergoing pulmonary valve replacement (PVR) have tricuspid valve regurgitation (TR). While tricuspid annuloplasty (TAP) is associated with an additional surgical risk, persistent significant TR after PVR may cause long term RV dysfunction and possibly alter outcome. The purpose of the study was to investigate whether TR improves spontaneously after PVR without TAP and to identify predictors of improvement in this setting.

**Methods:** We screened 186 consecutive patients with rTOF who underwent PVR between January 2005 and October 2009. Patients with significant (moderate or severe) TR prior to the PVR but did not have TAP at the time of PVR were included in the study. 2D-echocardiographic parameters pre and post-procedure were compared between patients with and without improvement.

**Results:** Twelve patients with significant TR were studied. All patients had significant pulmonary regurgitation. Mean age at

PVR was  $39 \pm 12.3$  years, 50% male. Mean age at Fallot repair was  $9 \pm 7.8$  years. Pulmonary valve was implanted surgically in 7 patients and percutaneously in 5 patients. Before PVR, 10 patients had moderate and 2 had severe TR. Seven patients (58%) had improvement in TR severity after PVR. Patients with improvement of TR had larger mid-right ventricle diameters ( $p = 0.008$ ) and lower Tei index ( $p = 0.004$ ). There was a trend for a greater reduction in RV size in patients who had significant improvement of TR ( $p = 0.09$ ).

**Conclusions:** TR significantly improved after PVR without TAP in patients with rTOF. Larger RV size and better RV function before procedure were related to improvement in TR, suggesting that the RV dilatation is the main driver for TR in this identity, rather than intrinsic tricuspid valve disease.

#### PW4-5

##### Relationship between temporal sequence of right ventricular deformation pattern and right ventricular performance in patients with corrected tetralogy of Fallot

Roest A.A.W. (1), Van der Hulst A.E. (1), Delgado V. (2) Holman E.R. (2), De Roos A. (3), Blom N.A. (1), Bax J.J. (2)

Division of Paediatric Cardiology, Dept of Paediatrics, Leiden University Medical Center, Leiden, The Netherlands (1); Dept of Cardiology, Leiden University Medical Center, Leiden, The Netherlands (2); Dept of Radiology, Leiden University Medical Center, Leiden, The Netherlands (3)

**Introduction:** RV dysfunction is common in cToF patients. Abnormalities in the temporal pattern of RV mechanical activation have been observed in cToF patients, but the relationship with RV performance remains unclear. Therefore, we evaluated right ventricular (RV) performance and the temporal sequence of RV deformation in corrected Tetralogy of Fallot (cToF) patients with 2-dimensional (2D) speckle tracking analysis.

**Methods:** Results: Thirty-seven cToF patients were compared to eighteen controls. Using 2D speckle tracking analysis, global RV strain was assessed. In addition, time to peak strain, as well as the time difference between RV inlet and RV outlet (RV time delay) was assessed.

**Results:** RV strain was reduced in patients as compared to controls ( $-20.9 \pm 4.3\%$  vs.  $-30.7 \pm 3.4\%$ ,  $p < 0.001$ ). Time to peak strain at the RV inlet showed no differences between cToF patients and controls (RV inlet:  $406 \pm 55$  ms vs.  $405 \pm 67$  ms,  $p = 0.429$ ), whereas time to peak strain at the RV outlet was significantly delayed in cToF patients (RV outlet:  $339 \pm 75$  ms vs.  $262 \pm 85$  ms,  $p = 0.003$ ). Consequently, RV time delay was significantly shorter in cToF patients (RV time delay:  $66 \pm 48$  ms vs.  $143 \pm 53$  ms,  $p < 0.001$ ). A strong relation between RV time delay and RV strain was observed ( $r = -0.70$ ,  $p < 0.001$ ).

**Conclusions:** Deformation of the RV outlet precedes deformation of the RV inlet in healthy children. In cToF patients, RV outlet deformation is delayed, causing a reduction of RV time delay which is strongly related to impairment in RV performance.

#### PW4-6

##### Innominate Vein-to-Common Atrium Fenestration at Fontan Completion: A Safe and Reproducible Option

von Samson P. (1), Mir T.S. (2), Arndt F. (2), Philipp L. (2), Schneck C. (2), Mueller G.C. (2), Dodge-Khatami A. (1)

Division of Congenital Cardiac Surgery, University Heart Center Hamburg-Eppendorf, Hamburg, Germany (1); Division of Pediatric Cardiology, University Heart Center Hamburg-Eppendorf, Hamburg, Germany (2)

**Background:** Adaptation to total cavopulmonary connection (TCPC) may be improved by a fenestration, through decreased venous pressure and augmented cardiac output. With the hypothesis of lower thrombo-embolic risk and a higher rate of late post-operative spontaneous closure, a new fenestration technique was attempted.

**Methods:** From 2008–2009, fourteen consecutive patients (mean age 3.4 years) were prospectively enrolled for an innominate vein-common atrium 5 mm Goretex graft fenestration during extra-cardiac TCPC completion. Studied variables included intubation time, N-BNP, arterial saturation, amount and duration of pleural effusion, and length of hospital stay. Fenestration monitoring was performed by contrast bubble echocardiography at hospital discharge and up to 6 months post-operatively.

**Results:** There was no mortality or significant morbidity. In 5/14 patients (43%), early (3 weeks) fenestration closure occurred. In this group saturations averaged 97% vs. 90% in patients with an open fenestration, and pleural effusion averaged 342 ml/kg (8 days) vs. 98 ml/kg (3 days;  $p < 0.05$ ). Maximal N-BNP levels were higher in patients with early fenestration closure (3556 vs. 1779 pg/ml;  $p < 0.05$ ). There was a positive correlation ( $p < 0.001$ ;  $r = 0.80$ ) between the durations of cardiopulmonary bypass and pleural effusion.

**Conclusions:** During TCPC, a 5 mm innominate vein-common atrium Goretex graft was a safe and reproducible technique, did not add to surgical difficulty or time, and provided reliable fenestration of up to at least 3 weeks, with a high rate of spontaneous closure during intermediate follow up. The modification provided lesser amount and shortened duration of post-operative pleural effusions, and potentially has a lower risk of paradoxical embolism from the upper extremities.



#### PW4-7

##### En Bloc Rotation of the Truncus Arteriosus – Experience With a Complete Anatomic Repair of TGA/VSD/LVOTO or DORV/TGA/VSD/LVOTO.

Mair R., Sames-Dolzer E., Innerhuber M., Steiner J., Gitter R., Tülzer G. Children's Heart Center Linz, AUSTRIA

**Introduction:** Standard method of reconstruction in TGA/VSD/LVOTO or DORV/TGA/VSD/LVOTO is the Rastelli procedure. Inherent problems are a long intracardiac tunnel with a tendency to subaortic obstruction and an allograft in the RVOT. Other options of repair of this malformation are: reparation a l'etage ventriculaire (Lecompte), aortic translocation procedure (Nikaidoh), the Ross-Konno switch procedure and the en-bloc rotation of the truncus arteriosus (Yamagishi). The latter procedure allows a complete anatomic repair with autologous material.

**Methods:** Aorta and pulmonary artery are divided at the same height. The coronaries are excised and mobilized. Aortic root, pulmonary root and part of the conusseptum are excised en bloc, rotated by 180° and reimplanted. The VSD is closed with a pericardial patch, thereby enlarging the LVOT. The coronaries are reimplanted. A Lecompte-maneuver is performed. Ascending aorta and aortic root are reanastomosed, Pulmonary root and main pulmonary artery are reconnected. 10 patients between 4d–5,7years – median age 74d [33; 367], median weight 3,8 kg [3,35 kg; 4,95 kg] (median, quartiles) – underwent this procedure since July 2003. The underlying anatomy was a DORV in 5 patients and a TGA in the other 5 patients Obstruction of the LVOT was subvalvar in 5 patients and valvar in the other 5.

**Results:** Hospital mortality was 0. Patients were discharged from the hospital after 12–31 days. The pulmonary valve could be preserved in 8 patients. A trans-annular patch was necessary in 2 cases. Postoperative echocardiography showed an unobstructed LVOT in all patients. A gradient between 16–36 mm Hg was detected across the pulmonary valve. Maximum follow up is 5,3years, median follow up is 0,3 yrs [0,03; 2,36] (median, quartiles). No gradient across the LVOT could be detected. 2 cases showed a mild aortic valve insufficiency (II°). A pacemaker had to be implanted in 1 patient after 2 months due to an AV-block II°. The gradient across the RVOT did not increase.

**Conclusions:** En bloc rotation of the truncus arteriosus is an excellent option for complete anatomic repair in TGA/VSD/LVOTO or DORV/TGA/VSD/LVOTO, preserving growth potential in all tubular structures. The pulmonary valve can be kept intact in the majority of cases.

#### PW4-8

##### Extracardiac Fontan operation without cardiopulmonary bypass reduces complications and blood transfusions

Ovroutski S., Redlin M., Miera O., Peters B., Ewert P., Berger E., Hetzer R., Hübner M. German Heart Institute Berlin.

**Objective:** Use of modified extracardiac Fontan operation (ECFO) for total cavo-pulmonary connection (TCPC) allows to avoid cardiopulmonary bypass (CPB) and seems to improve early postoperative results. We evaluated our experience with the off-pump technique for ECFO.

**Methods:** Out of the total of 130 who underwent ECFO (1997–2009) the last 6 consecutive patients (median age 4 (2.6–11) years, median weight 16 (11.7–36) kg) were operated on without CPB. Preoperative data were in median: mean pulmonary artery pressure (mPAP) 11 mmHg, pulmonary artery index  $234 \text{ mm}^2/\text{m}^2$ , oxygen saturation 78% and Hb 15.2 g/dl. The main diagnoses were hypoplastic left heart syndrome in 2 patients, tricuspid atresia (TA) in 2 and other complex univentricular heart in 2. The non-fenestrated Gore-Tex® graft (16–20 mm) was implanted during temporary passive inferior vena cava (IVC)-to-atrial bypass through 18 or 22 french cannulas. In 2 children (2.6 and 3.4 years) primary complete

TCPC without prior Glenn operation could be performed with off-pump technique.

**Results:** Perioperatively we observed slight arterial pressure drop of 15 mmHg (MAP in median from 57 to 42 mmHg), as well as IVC pressure increase of 8 mmHg (in median from 14 to 22 mmHg) after IVC clamping, but no further hemodynamic disturbances. Mean PAP on the first postoperative day was in median 12 (9–14) mmHg and the postoperative course was uneventful in all patients with extubation during the first 24 hours and discharge within 10 days. No patient had prolonged pleural effusions or postoperative cyanosis. With our specific off-pump technique we were able to avoid blood transfusions; the lowest postoperative Hb-level on discharge was 9.1 g/dl.

**Conclusions:** Both secondary and primary TCPC using extra-cardiac Fontan operation may be performed in selected patients without CPB. This results in improved postoperative hemodynamics, very low incidence of complications, and rapid recovery, and allows the use of blood products to be significantly reduced.

#### PW4-9

##### **Dilated cardiomyopathy presenting in childhood: etiology, diagnostic approach and clinical course**

*Gesute V (1), Ragni L. (1), Mariucci E. (1), Oppido G. (2), Formigari R. (1), Prandstraller D. (1), Gargiulo G. (2), Picchio F.M. (1)*

*Pediatric Cardiology Unit, S.Orsola-Malpighi Hospital, Bologna, Italy (1); Pediatric Cardiac Surgery Unit, S-Orsola-Malpighi Hospital, Bologna, Italy (2)*

**Introduction:** To determine the outcome of dilated cardiomyopathy (DCM) presenting in childhood and the features that might be useful for prognostic stratification.

**Methods:** Retrospective study of 41 consecutive children affected by DCM (age 0–14 years; median 33.4 ± 49.25 ranging from 0 to 14 years between 1993 and 2008). The medical history was reviewed to determine age at diagnosis, family history, previous viral illness, etiology, symptoms and signs at presentation, treatment and outcome. The diagnosis was made on the basis of cardiomegaly and evidence of poor left ventricular function by echocardiography. A metabolic evaluation including blood lactate, pyruvate, carnitine, amino acids and urine organic acids, assessment of respiratory chain enzymes and analysis of histopathological material was also carried out. Survival curves were constructed by the Kaplan-Meier method.

**Results:** Follow-up ranged from 10 days to 162 months (median 45.25 ± 41.15 months). Freedom from death or heart transplantation was 68.3% at five years. At presentation, 20 patients were under the age of six months, 6 were between 6 months and two years, 8 were between 2 and 5 years and 7 were above the age of five years. Seven children (17%) had a family disease while 34 patients (83%) had a non-familial form. Signs of overt congestive heart failure were present in 28 pts. (68.3%). The primary end-point of death/heart transplantation was associated with the need for iv. inotropic support (5 out of 12 pts, 41.6% vs. 2 out of 16 pts., 12.5%,  $p = 0.011$ ). A trend towards a poorer prognosis was found for age at diagnosis more than 5 years (57.1% vs. 20.5%,  $p > 0.05$ ) and for a metabolic etiology of DCM (50.0% vs. 25.7%,  $p > 0.05$ ). For the children affected by DCM as a part of a multisystemic involvement, the mortality was 50%.

**Conclusions:** In children, DCM is a diverse disorder with outcomes that depend on cause, age and heart failure status at presentation. Overt heart failure at presentation is a major prognostic factor for death or heart transplantation. Older age at presentation and metabolic etiology may be associated with a poorer prognosis.

#### PW4-10

##### **Treatment of terminal heart failure and cardiogenic shock in 26 neonates and infants assisted by Berlin Heart Excor device**

*Miera O. (1), Hübler M. (2), Hetzer R. (2), Berger F. (1)*  
*Dept. of Congenital Heart Disease/Pediatric Cardiology, Deutsches Herzzentrum Berlin, Germany (1); Dept. of Cardiothoracic and Vascular Surgery, Deutsches Herzzentrum Berlin, Germany (2)*

**Introduction:** Since the first implantation of a pulsatile paediatric ventricular assist device (VAD, Berlin Heart) in an infant in 1992 at our institution outcome has significantly improved, with the implementation of optimized cannulas, modified anticoagulation and optimized surgical and intensive care management since 1999. We present our clinical experience with the Berlin Heart Excor device over the last 10 years in infants under one year of age.

**Methods:** We performed a retrospective case analysis of all infants under 1 year of age who were treated with the Berlin Heart Excor between January 1st 1999 and December 31st 2009.

**Results:** Twenty-six infants, age 11–356 d (median 127 d), weight 3.2–9.5 kg (median 5.5 kg), were treated with VAD. Median time on support was 31 days (1–175 d). Twenty-one infants had left VAD and five infants biventricular VAD support systems. All infants suffered from terminal heart failure with multiorgan failure or cardiogenic shock. Fifteen had cardiomyopathy, four suffered from fulminant myocarditis and seven had a preceding operation on cardiopulmonary bypass. Procedural success was achieved in 80% of infants: seven could be weaned from the system, another 13 patients received successful heart transplantation, five infants died and one is still on mechanical support. All patients with fulminant myocarditis were weaned from the assist system.

At follow-up 1 year after explantation ( $n = 21$ ) the statomotoric development was normal or near normal in 13 of the 16 surviving children. Thromboembolic events occurred in five infants: one had mild hemiparesis, two severe statomotoric retardation and two recovered completely.

**Conclusions:** With a total survival rate of 80% the outcome of VAD support in small infants is comparable to that in older children. The VAD is now a well-established treatment for infants suffering from cardiogenic shock of any aetiology.

#### PW4-11

##### **Fœtal double discordance : a series of 65 foetuses**

*Bah G. (1), Fermont L. (2), Le Bidois J. (2), Bonnet D. (2)*  
*Neonatalogie, CH Le Mans, France (1); Centre de Référence Malformations Cardiaques Congénitales Complexes – M3C, AP-HP, Université Paris Descartes, Paris, France (2)*

**Background:** Fœtal diagnosis of double discordance (DD) raises a variety of questions with regards to post-natal outcome and prediction of prognosis.

**Aim of the study:** To review retrospectively all consecutive foetuses with a diagnosis of DD in our institution.

**Methods:** All foetuses transferred in utero with a final diagnosis of DD were reviewed for outcome of pregnancy and short-term postnatal outcome.

**Results:** Sixty five foetuses were identified. Sex ratio was 2.64. Term of diagnosis was 25 weeks gestation. Reason for referral to tertiary centre were: structural heart defect at foetal echoscan in 46 cases, atrioventricular block (AVB) in 9 cases, situs inversus/dextrocardia in 4 cases and screening for family history of congenital heart defect in 6 cases. Twenty/65 foetuses were referred for transposition of the great arteries (TGA). Foetal diagnosis of DD was confirmed in 53 cases while in the

remaining 13 cases the diagnosis was tetralogy of Fallot or pulmonary atresia-VSD in 11, TGA in 1 and hypoplastic left heart syndrome in 1. Additional cardiac anomalies were: VSD in 22, pulmonary atresia with VSD in 11, obstructive left heart disease in 5, Ebstein anomaly in 10. Parents chose termination of pregnancy in 20 cases (all had DD with associated cardiac anomalies). Forty-five foetuses were live born. Associated cardiac anomalies were confirmed in all but 12 cases of VSD where the interventricular septum was found intact. Significant tricuspid valve regurgitation was found in 7 neonates and was consistent with prenatal findings in only 3. During first year follow-up, 5 patients died (all with associated cardiac anomalies); 17 were operated either for a Blalock shunt or for pulmonary artery banding; and 3 had a pace-maker implanted. All 20 non operated patients are doing well but 3 have significant tricuspid regurgitation.

**Conclusion:** Foetal diagnosis of DD remains difficult with a high rate of incomplete or erroneous diagnoses. Prenatal counselling is a challenge as postnatal outcome in isolated DD, while favourable in the short term follow-up, remains uncertain for the long term. Preventive banding of the pulmonary artery might be interesting in the prevention of progressive tricuspid regurgitation.

#### PW4-12

##### Long-term Follow-up of Pulmonary Valve Replacement Following Repair of Tetralogy of Fallot

Ruzmetov M., Yurdakok O., Rodefeld M.D., Turrentine M.W., Brown J.W.

Indiana University School of Medicine, Indianapolis, Indiana, USA

**Background:** Pulmonary insufficiency appears to be well tolerated following repair of tetralogy of Fallot (TOF); however, it may result in progressive right ventricular (RV) dilatation requiring pulmonary valve replacement (PVR). We report our experience with late PVR after repair of TOF.

**Methods:** There were 92 patients reoperated between 1978 and 2009. Mean age was 16.2  $\pm$  12.9 years (range, 2–59). The mean interval between repair of TOF and PVR was 11.9  $\pm$  9.4 years (range, 1–47). Indication for reoperation was decreased exercise tolerance in 62% of patients, right heart failure in 38%, RV dilatation with arrhythmia and syncope in 24%, and progressive RV dilatation in 21%. Tissue prosthetic valves were used in 52 patients, homografts in 16, and Gore-Tex transannular patching in 24. Fifty-two patients underwent PVR as the sole procedure. Concomitant procedures in the other 40 patients included pulmonary artery patch angioplasty (n = 13), tricuspid valve repair (n = 10), residual VSD closure (n = 6), MAZE procedure (n = 5), ASD closure (n = 4), and aortic valvuloplasty (n = 2). Preoperatively, 57% (52/92) of patients were in NYHA class III-IV.

**Results:** There were 2 late deaths, both were cardiac related. Survival was 98% at 5, 10 and 20 years. Mean follow-up was 10.2  $\pm$  6.9 years (range, 6 months to 20 years). Functional status of patients was improved significantly. Following PVR, 97% (87/90) of surviving patients were in NYHA class I-II (p < 0.001). Nine patients underwent pulmonary valve re-replacement (re-PVR) with a mean interval of 9.2  $\pm$  3.8 years (range, 3–16 years). Freedom from re-PVR was 96% at 5 years, and 92% at 10 years, and 90% at 20 years. The only factor predicting re-PVR was young age at the time of PVR (p = 0.02).

**Conclusions:** Late PVR after repair of TOF can be done with low mortality and results in improved functional status. Subsequent pulmonary valve re-replacement may be necessary to maintain functional improvement.

#### P-1

##### Beta-blockers in the management of Congenital Long QT Syndrome

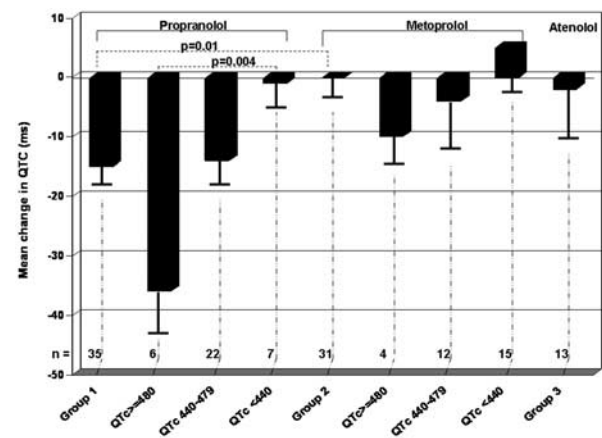
Chockalingam P. (1), Beckmann B.M. (2), Clur S.A.B. (1), Heuvel E van den. (3), Kaab S. (2), Blom N.A. (1), Wilde A.A.M. (1) Academic Medical Centre, Amsterdam, Netherlands (1); Ludwig-Maximilians-University, Klinikum Grosshadern, Munich, Germany (2); University Medical Centre, Groningen, Netherlands (3)

**Introduction:** Beta-blockers (BB) form the mainstay in managing Congenital Long QT Syndrome (LQTS). Based on recent findings that propranolol blocks late non-inactivating sodium currents while metoprolol does not, we aimed to study the electrocardiographic differences among patients using the different BB.

**Methods:** Study population included genotype-positive pediatric LQTS patients (n = 79), divided into 3 groups based on initial BB treatment (propranolol [P] = 35, metoprolol [M] = 31, atenolol [A] = 13). Subset 1 included 17 subjects in group P who changed from P to M during follow-up and subset 2 included all 13 subjects in group A who changed from A to M. Manual QTc measurements were made from lead II by an investigator blinded to therapy details.

**Results:** Gender distribution, type of LQTS and mode of presentation were not significantly different among the 3 groups. Baseline (BL) heart rate and QTc of group P were significantly higher compared to group M (93  $\pm$  21 vs 79  $\pm$  21 bpm, p = 0.01, 462  $\pm$  33 vs 442  $\pm$  35 ms, p = 0.02). More QTc-shortening was produced by P (462  $\pm$  33 to 447  $\pm$  29 ms, p = 0.047, time interval 9  $\pm$  9 months) compared to M (442  $\pm$  35 to 438  $\pm$  40 ms) and A (460  $\pm$  29 to 458  $\pm$  36 ms). Longer the BL QTc, more pronounced was the QTc-shortening (mean-SEM, see figure) in group P. The QTc of subset 1 was significantly shorter prior to change from P to M (450  $\pm$  29 vs 463  $\pm$  41, p = 0.038), an effect not seen in subset 2.

**Conclusions:** Propranolol has a significantly better QTc-shortening effect compared to metoprolol and atenolol and this effect is stronger in patients with longer QTc. Propranolol should probably be first-line therapy in LQTS.



#### P-2

##### Dyssynchrony and ventricular function improve following catheter ablation of non-septal accessory pathways in children

Abadir S. (1,2), Fournier A. (1), Dubuc M. (2), Hugues N. (1), Garceau P. (2), Khairy P. (2)

Hopital Sainte Justine, Montreal, Canada (1); Montreal Heart Institute, Montreal, Canada (2)

**Introduction:** Prior studies assessing ventricular preexcitation-induced dyssynchrony in children have focused on septal or paraseptal pathways and described paradoxical or hypokinetic septal motion. Data regarding non-septal pathways is limited, as these have generally been perceived to carry lesser risk for ventricular dysfunction.

**Methods:** We characterized and compared the degree of left ventricular (LV) dyssynchrony and LV systolic and diastolic function by echocardiographic analyses with tissue Doppler imaging (TDI), prior to and following catheter ablation of septal and non-septal accessory pathways.

**Results:** A total of 16 consecutive children, age  $14.2 \pm 3.7$  years, weight  $53 \pm 17$  kg, were systematically assessed. All had Wolff-Parkinson-White syndrome, structurally normal hearts, and underwent successful ablation (cryoenergy in 4; radiofrequency in 12). Septal or paraseptal pathways were present in 6 (37.5%) and non-septal pathways in 10 (62.5%): left lateral (N = 5), right lateral/anterolateral (N = 3), left posterior (N = 2). Following ablation, LV ejection fraction (EF) (Simpson's method) increased by  $4.9 \pm 2.1\%$  ( $p = 0.038$ ) from a baseline value of  $57.0 \pm 7.8\%$ , with a decrease in the difference between aortic and pulmonary pre-ejection times ( $11.0 \pm 3.3$  ms,  $p = 0.017$ ). By TDI, the interval between QRS onset and peak systolic velocity decreased from 33.0 [interquartile range (IQR) 20.0, 18.0] to 18.0 (IQR 5.0, 24.0) ( $p = 0.013$ ). No significant changes in septal-to-posterior wall motion delay or diastolic parameters were noted. LVEF increased to a greater degree following ablation of non-septal ( $5.9 \pm 2.6\%$ ,  $p = 0.023$ ) versus septal ( $2.5 \pm 4.1\%$ ,  $p = 0.461$ ) pathways. The 4 patients with LVEF < 50% prior to ablation, 2 of whom had left lateral pathways, improved to >50% post ablation. Similarly, the magnitude of improvement in LV dyssynchrony was more marked in patients with non-septal versus septal pathways, e.g. difference between septal and lateral wall motion delay before and after ablation of  $20.6 \pm 7.1$  ms ( $p = 0.015$ ) versus  $1.4 \pm 11.4$  ms ( $p = 0.655$ ).

**Conclusion:** LV systolic function and dyssynchrony improve after successful ablation of antegrade-conducting accessory pathways in children, with more pronounced changes noted for non-septal pathways.

### P-3

#### **Sudden cardiac death: clinical evaluation of paediatric family members**

*Tomaske M., Bauersfeld U.*

*Division of Paediatric Cardiology, University Children's Hospital Zurich, Switzerland*

**Introduction:** The annual incidence of premature sudden cardiac death (SCD) is estimated as 1 in a 100 000. A number of SCDs are caused by inherited heart diseases, such as cardiomyopathies, channelopathies or conduction system disorders with predominant autosomal dominant inheritance.

**Patients and Methods:** A total of 36 consecutive referred families after sudden cardiac death ( $n = 29$ ) or life-threatening event ( $n = 7$ ) of a family member younger than 50 years were analyzed. Mean age of the 58 paediatric family members was  $8.3 \pm 6.1$  years. Degree of relationship to index patient was either first- ( $n = 37$ ) or non-first-degree ( $n = 21$ ). Stepwise evaluation included history, 12-lead and 24-hour electrocardiogram (ECG) as well as 2D echocardiography in all. Additional investigations were signal average ECG, cardiac magnetic resonance imaging (cMRI), event recorder and genetic testing when indicated.

**Results:** Inherited heart disease in the index patient was identified or suspected due to autopsy, post-event or family evaluation in

23/36 (64%) families: cardiomyopathies ( $n = 12$ ), channelopathies ( $n = 9$ ), and sinus node dysfunction ( $n = 2$ ). Eight (22%) families had a history of additionally unexplained premature SCDs. A total of 18 (31%) of the evaluated paediatric relatives were diagnosed with proven or likely inherited heart disease: cardiomyopathies ( $n = 6$ ), channelopathies ( $n = 8$ ), and sinus node dysfunction ( $n = 4$ ). The following evaluations contributed either separately or combined to the diagnosis: 12-lead ECG ( $n = 12$ ), 24-hour ECG ( $n = 10$ ), 2D echocardiography ( $n = 5$ ), signal average ECG ( $n = 1$ ) and cMRI ( $n = 1$ ). Channelopathies were genetically proven in 3 children. Prevention of SCD or of a cardiac event was initiated in 16/58 (28%) by implantation of an anti-bradycardia device ( $n = 3$ ), an implantable cardioverter defibrillator (ICD,  $n = 6$ ) and/or medication with antiarrhythmic drugs ( $n = 8$ ). Subsequently, no cardiac event was noted in any patient during a follow-up period of 3.6 (0.8–6.7) years. Appropriate and successful ICD discharges occurred in 4 (22%) of the affected patients.

**Conclusions:** A stepwise, comprehensive clinical investigation of SCD or life-threatening event families identifies a substantial number of paediatric relatives at risk for sudden cardiac death. This allows for targeted prevention by effective treatments and evaluation of further relatives.

### P-4

#### **On the origin of the Y111C/KCNQ1 founder mutation- a major cause of the Long QT Syndrome in Sweden**

*Winbo A. (1), Diamant U.B. (2), Rydberg A. (1), Jensen S.M. (2), Stattin E.L. (3)*

*Department of Clinical Sciences, Division of Pediatrics, Umeå, Sweden (1); Department of Public Health and Clinical Medicine, Division of Medicine, Umeå, Sweden (2); Department of Medical Biosciences, Medical and Clinical Genetics, Umeå, Sweden (3)*

**Introduction:** In the Swedish population, a single mutation (Y111C/KCNQ1) has been identified as a major cause of the Long QT Syndrome (LQTS) – a disease causing preventable paediatric cardiac death. The Swedish Y111C carriers exhibit a surprisingly mild clinical phenotype, in spite of the mutations' dominant-negative electrophysiological properties demonstrated in vitro. The aim of this study was to investigate the origin and possible founder-nature of the Y111C/KCNQ1 mutation.

**Methods:** In all Swedish Y111C families, ancestors' geographical origin, migration and possible interrelation were investigated using parish registers and genealogical databases. In 26 index families haplotype analysis was performed using 15 satellite markers, 6 upstream and 9 downstream of the KCNQ1 gene (distance ~ 8 cM). To identify the mutation-associated allele, two mutation-carriers in separate generations in each family were analyzed. Forty-eight chromosomes from healthy Swedish controls were included to calculate allele frequency. The ESTIAGE computer software was used for estimating the age of the mutation.

**Results:** The Swedish Y111C population comprises >150 identified mutation-carriers in >30 index families. Their ancestors apparently originated from a single northern region, from where the population spread, migrating along the Ångerman river valley during the 17th–19th century. Twenty-five index cases are genealogical descendants of a founder couple born ~ 1600, whose two sons themselves are founders of two separate branches of the Y111C pedigree.

The 26 haplotyped Y111C families share 3–15 (median 13) uncommon allele-variants surrounding the Y111C locus, with frequencies ranging between 0.02–0.69 (median 0.17) in the general population. Familial haplotypes co-segregate within the subdivisions of the pedigree, supporting the genealogical data.

The estimated age of the mutation is 31 generations (95% CI 23; 41). Assuming that one generation is 25 years, the mutation is 775 years old (95% CI 575; 1,025).

**Conclusions:** The Y111C mutation was probably introduced in the inland of northern Sweden by the first colonists. The subsequent population development within the relatively isolated river valleys caused strong founder effects that in combination with the mutations' mild phenotype could enable the enrichment of this LQT1 mutation in the Swedish population. This Y111C founder population constitutes an invaluable asset for future genetic and clinical studies.

#### P-5

##### **Feasibility and reproducibility of conventional and color-coded tissue Doppler M-mode echocardiography in the assessment of septal-to-posterior wall motion delay in children and adolescents with right ventricular pacing.**

*Udink ten Cate F.E.A. (1), Schmidt B.E. (2), Wiesner N. (1), Krüssell M.A. (3), Sheta S.S. (4), Breuer J. (5), Trieschmann U. (3), Brockmeier K. (1), Sreeram N. (1)*

*Departments of Pediatric Cardiology of the Heart Center Cologne, University Hospital Cologne, Cologne, Germany (1); Children's Hospital Cologne, Cologne, Germany (2); Department of Anesthesiology and Postoperative Intensive Care Medicine, University Hospital of Cologne, Cologne, Germany (3); Cairo University Children's Hospital, Cairo, Egypt (4); University of Bonn, Bonn, Germany (5)*

**Objectives:** Right ventricular pacing has detrimental effects on cardiac function. Assessment of left ventricular (LV) dyssynchrony may be helpful in identifying children at risk for pacing-induced heart failure. This study evaluated the feasibility and reproducibility of M-mode echocardiographic methods in the assessment of LV dyssynchrony in children with permanent pacing.

**Methods:** Septal-to-posterior wall motion delay was measured using conventional M-mode (SPWMD) and color-coded tissue Doppler M-mode echocardiography (ccSPWMD) in 22 children and adolescents (mean age  $13.9 \pm 6.8$  years (yrs), male 59%) with right ventricular pacing and 22 healthy age-matched controls. Indications for pacing included congenital heart block ( $n = 8$ , 36%) or surgical acquired heart block ( $n = 16$ , 64%). Intra- and interobserver variability, and agreement was obtained. LV dyssynchrony was defined as a septal-to-posterior wall motion delay of  $> 130$  ms.

**Results:** After long-term pacing (median 7.4 yrs, range 1.8–18.4 yrs), LV dyssynchrony was demonstrated in 55% and 50% of the patients, using SPWMD and ccSPWMD, respectively. Septal-to-posterior wall motion was not assessable in 6 patients (SPWMD  $n = 2$  (9%), ccSPWMD  $n = 4$  (18%)), due to hypokinesia ( $n = 1$ ), paradoxical septal motion ( $n = 2$ ), or poor image quality ( $n = 3$ ). Intraobserver variability was relatively good (SPWMD:  $13.5 \pm 10.1\%$ , ccSPWMD:  $8.5 \pm 8.1\%$ ,  $p < 0.001$ ). Interobserver variability was acceptable (SPWMD:  $19.7 \pm 7.8\%$ , ccSPWMD:  $14.2 \pm 8.7\%$ ,  $p < 0.001$ ). There was disagreement in presence of LV dyssynchrony in 2 patients (9%). Patients and controls were similar in age. Paced children had a significantly decreased ejection fraction compared with controls ( $63.8 \pm 7.6$  vs  $69.0 \pm 5.6$ ,  $p < 0.001$ ). There was no relation between QRS duration and LV dyssynchrony ( $r = 0.04$ ,  $p = 0.8$ ).

**Conclusions:** LV dyssynchrony can be identified in more than 50% of children after long-term pacing. Color-coded tissue Doppler M-mode echocardiography is more reliable than conventional M-mode echocardiography in the assessment of septal-to-posterior wall motion delay. Applying these methods in pediatric patients can be challenging.

#### P-6

##### **Can primary health care physicians interpret pediatric ECGs?**

*Germanakis I., Nasikas D., Kalmanti M.*

*Dpt of Pediatrics, University Hospital Heraklion, Crete, Greece*

**Introduction:** ECG has been recommended by ESC for pre-participation screening in asymptomatic competitive young and adult athletes. However, given the limited availability of pediatric cardiologists, the feasibility of preparticipation screening of young children will largely depend on the clinical skills of primary health care physicians to correctly interpret pediatric ECGs. We aimed to evaluate the background knowledge in pediatric ECG of primary health care physicians and the potential benefit from the attendance of a structured teaching course.

**Methods:** A two-day intensive teaching course on pediatric ECG, with theoretical and practical exercise was offered to 38 medical doctors providing primary health care services: 12 pediatricians, 8 general physicians (board certified and trainees) and 12 medical school graduates. Participants were asked to validate a series of 10 representative pediatric ECGs (5 with axis/amplitude abnormalities, 5 with arrhythmias) (pre-test), while a different series of 10 pediatric ECGs (with similar though distribution of abnormalities) was provided at the end of the course (post-test). Mean score of correct answers was documented. The overall quality and usefulness of the course was anonymously assessed by participants by a 5-level scale at its end (1:very bad to 5: excellent).

**Results:** The mean (SD) pre-test and post-test scores were 42.2 (20.8) and 73.4(14.1) respectively ( $p < 0.001$ , paired T test). The training level (medical graduates: 40.5, trainees: 45, board certified: 41.2) or the specialty (general physicians: 51.4, pediatricians: 39.5) had no significant impact on pre-test score ( $p = 0.861$  and  $p = 0.444$ , ANOVA). Although all participants demonstrated an improvement (defined as difference between the two scores through pre-test score) following the course, this improvement was more pronounced to participants with the lowest pre-test score ( $r = -0.796$ ,  $p < 0.001$ , Pearson cor. Coef.). Participants validated the course with a mean score 4.68 and 4.85 for overall impression practical usefulness, respectively.

**Conclusions:** The overall basic skills of primary health care physicians regarding pediatric ECG interpretation are suboptimal and independent from specialty or training level. Structured teaching courses can demonstrate a measurable improvement of participant's skills, contributing in the improvement of pediatric heart disease detection from primary health care physicians and allow for a cost effective preparticipation screening.

#### P-7

Withdrawn

#### P-8

##### **Ventricular automatic capture (AC) is feasible and safe with unipolar epicardial leads in young patients with congenital heart disease**

*Kerst G. (1), Gass M. (2), Apitz C. (1), Nagy Z. (3), Ziemer G. (3), Hofbeck M. (1)*

*Pediatric Cardiology, University Children's Hospital Tuebingen, Germany (1); Heart Centre Bodensee, Konstanz, Germany (2); Congenital Heart and Pediatric Cardiac Surgery, University Children's Hospital, Tuebingen, Germany (3)*

**Introduction:** Permanent pacing therapy in young patients requires high lower rate limit settings and high pacing rates resulting in



augmented energy consumption and shortened battery life. Ventricular automatic capture (AC) can reduce energy consumption by automatic adjustment of pacing thresholds and pacing output amplitudes. Pacemakers with “beat-to-beat” capture verification technology use evoked response analysis. Appropriate ventricular AC has not been investigated for feasibility and safety when small heart size or scar tissue necessitates the use of unipolar epicardial leads not specifically designed for low polarizing potentials.

**Methods:** 15 patients with congenital heart disease (age 3.6 to 20.4, median 10.2 years) and a unipolar suture-on or a suture-less screw-in epicardial lead (lead #1: Medtronic 4965 CapSure Epi, Medtronic Inc., Minneapolis, MN, USA, n = 5; lead #2: Biotronic ELC-UP, Biotronic Inc., Berlin, Germany, n = 10) and a pacemaker with AC (Insignia I Ultra, Guidant Inc., St. Paul, MN, USA) were enrolled in this retrospective study. AC in this pacemaker model has specifically been designed for unrestricted usage regarding pacing lead models and pacing/sensing configurations. Leads were implanted via a subxyphoidal incision or via midline sternotomy in case of concomitant cardiac surgery. Intraoperative mean pacing thresholds/R-wave sensing values were  $1.1 \pm 0.36 \text{ V}@0.5 \text{ ms}/7.4 \pm 4.1 \text{ mV}$  and  $1.2 \pm 0.41 \text{ V}@0.5 \text{ ms}/7.5 \pm 5.8 \text{ mV}$  for lead #1 and lead #2, respectively.

**Results:** AC was successfully activated at pre-discharge in all 15 patients. An early exit block precluded further analysis in two patients (with lead #1 and lead #2 respectively). AC worked in 12 of 13 patients tested at 6, 12 and 24 months and in 10 of 11 patients tested 36 months after pacemaker implantation. In the particular patient without adequate AC during the first 36 months, AC could be activated when retested 54 months after implantation and has remained stable since then (last follow-up 60 months after implantation). During follow-up no adverse effects of AC were seen. Follow-Up and further analysis is ongoing.

**Conclusion:** Ventricular automatic capture (AC) is feasible and safe using Insignia I Ultra in combination with the above mentioned epicardial lead systems in young patients with congenital heart disease.

## P-9

### 6-year experience with implanted cardioverter-defibrillators (ICD) in children in Department of Cardiology at Children's Memorial Health Institute, Warsaw, Poland.

Brzezinska-Paszke M. (1), Bieganowska K. (1), Miszczak-Knecht M. (1), Rękawek J. (1), Kaszuba A. (1), Raszek M. (1), Kawalec W. (1), Birbach M. (2)

Department of Cardiology, Children's Memorial Health Institute, Warsaw, Poland (1); Department of Cardiosurgery, Children's Memorial Health Institute, Warsaw, Poland (2)

**Introduction:** Implementation of ICD was a milestone in saving lives of patients with life-threatening ventricular arrhythmias. However, potential complications may severely impact patients' life.

**Method:** We analysed 36 patients (18 male), age 1–17 years (mean 13 years), hospitalized in our Department of Cardiology during last 6 years, who received ICD.

**Results:** Indications for implantation were: aborted sudden cardiac death (SCD) –21pts, primary prevention of SCD –15 pts (long QT syndrome with ventricular tachycardia (VT) in holter-ECG and syncope in anamnesis–10pts, ventricular tachycardia/ventricular fibrillation (VT/VF) in holter-ECG –3pts and hypertrophic cardiomyopathy with syncope and ventricular arrhythmia –2pts). We implanted 38 ICDs including: 30 endocardial (26 DDD, 4 VVI) and 8 epicardial (7 using endocardial leads implanted epipleural, 1 – using epicardial leads). 3 pts required ICD replacement system because of lead

damage and 1 needed reoperation due to battery wear. In the follow-up period which lasted from 1 month to 6 years (mean 26 month) we noted shocks in 14 pts. In 6 pts shocks were delivered because of ventricular arrhythmia and successfully terminated ventricular fibrillation. Nine children received inappropriate shocks because of sinus tachycardia (5 pts), atrial fibrillation/supraventricular tachycardia (2pts), T-wave oversensing (1 time), artifacts from the damaged lead (2pts). We have eliminated inappropriate shocks by setting just 1 therapeutic zone – ventricular fibrillation >240 bpm, optimized T-sensing and changed damaged leads. In 3 pts we have observed electrical storms causing multiple ICD shocks. Due to ICD shocks-related trauma some patients required assistance of psychiatric therapists. All patients are alive except a boy who had a heart transplantation followed by ICD removal. All patients are on high doses of beta blockers.

**Conclusions:** ICD is a well known life saving method in children with life-threatening ventricular arrhythmias of non reversible causes. After implantation patients still need to take medicines and control the devices every 3–4 months. Only properly programmed ICD can reduce occurrence of inappropriate shocks.

## P-10

### Drug-refractory arrhythmias: radiofrequency ablation in neonates

Brugada G., Bartrons J., Jimenez L., Rissech M., Carretero J., Prada F., Brugada J.

Arrhythmia Unit, Cardiology Section, Hospital Sant Joan de Déu, Barcelona, Spain

**Introduction:** Incessant tachycardia can lead to hemodynamic instability. Fetus and neonates are especially sensitive to incessant tachycardias, eventually mortal, even in utero. Drugs, alone or in combination, can revert or slow down tachycardia, allowing hemodynamic improvement. But when poly-therapy is required, severe secondary effects can be observed. Then, the only available treatment is ablation. Single institution experience in neonatal tachycardia ablation is presented.

**Patients and methods:** Inclusion criteria are neonates referred to our institution with diagnostic of hemodynamically significant incessant tachycardia, refractory to intensive antiarrhythmic treatment. Previous detailed explanation of technique risks to the family, especially in this age group. Under general anesthesia, we perform electrophysiological study introducing single 5-French catheter for detection, stimulation and ablation, from venous approach, preferably femoral. Once the origin of the tachycardia is located, radiofrequency ablation is performed under radioscopic surveillance.

**Results:** In the last ten years, we have performed 18 procedures in 17 neonates (age 5–to–28 days), with mean weigh of 3,3 kg (range 1,5–4,9 kg). Three patients had associated congenital heart disease (hypertrophic cardiomyopathy, significant duct and non-compaction of left ventricle). Mean procedure time was  $41,76 \pm 21,69$  minutes, and mean radiation time of  $6,4 \pm 5,2$  minutes. Single catheter technique was used for all patients but one (who required two catheters to perform the procedure). Diagnostics were: 6 patients had concealed accessory pathway, 5 patients had Coumel type tachycardia, 4 patients with atrial flutter, 1 junctional ectopic tachycardia and 1 WPW. Immediate efficacy was obtained in 16 out of 18 procedures: in one case study had to be cancelled due to pericardial effusion, and was finally ablated in a later procedure; in another case, tachycardia could not be induced. Two patients had complications: one with

pericardial effusion requiring drainage with residual mild mitral regurgitation; and the second has residual moderate mitral regurgitation, stable after 7 years of follow-up.

**Conclusions:** In our institution, radiofrequency ablation is indicated in severely ill neonates with tachycardia refractory to drugs. We perform ablation using so called single-catheter technique. Efficacy is very high, with acceptable moderate complications in this group of patients, otherwise compromised by their arrhythmia.

#### P-11

##### **Impact of the individual circadian QTc variance for the validation of QT prolongation in children and adolescents**

*Fischer M., Ulmer H.E.*

*University Children's Hospital, Department of Paediatric Cardiology, Heidelberg, Germany*

**Introduction:** In children the duration of the QT interval depends on several well known factors, e.g. gender, age, heart rate. The importance of the circadian variance of the QT interval seems to be widely underestimated. Therefore we compared the variance of the QT interval in young patients with long QT syndrome (LQTS) and probands.

**Methods:** In 142 children and adolescents, 76 patients with LQTS (mean age 10.1 [0.4–19.9] years) and 66 probands (mean age 11.7 [2.1–18.8] years) a 12-lead Holter recording (Mortara Instruments®, H-Scribe-II-System) was performed. QTc intervals were measured every quarter of an hour in leastwise 6 leads of one heart beat. The mean value of all measured QTc intervals for this selected heart beat was used to determine maximal and minimal QTc and finally the QTc range for each Holter recording. QTc was calculated according to the Bazett formula. **Results:** Mean QTc differed significantly between patients and probands ( $490 \pm 44$  vs  $415 \pm 29$  ms;  $p < 0,0001$ ). The mean range of the QTc values were  $119 \pm 32$  ms and  $114 \pm 27$  ms (not significant).

**Conclusions:** As well as in adults the frequency adjusted QT interval (QTc) is the mainstay of diagnosis of long QT syndrome in children. Our analysis of 24-hour ECG's found a high range of QTc values during this time period in children with LQTS. Surprisingly probands in the same age showed equal ranges of QTc values. Hence, single QTc values in the normal range measured in the standard ECG are not sufficient to exclude the diagnosis of QT prolongation.

#### P-12

##### **Diagnostic Accuracy of Brain Natriuretic Peptide for Congenital Heart Disease in the First Month of Life.**

*Cantinotti M., Arcieri L, Margaryan R, Maizza A, Storti S, Prontera C, Luisi S, Bernabei M, Crocetti M, Assanta N, Clerico A, Murzi B. Pediatric Cardiac Surgery Unit, G Monasterio Foundation and Institute of Clinical Physiology-National Research Council, Heart Hospital, Massa, Italy*

**Introduction:** to evaluate the diagnostic accuracy of BNP assay in children with congenital heart disease (CHD) in the first month of life.

**Methods:** BNP was measured in 152 neonates with untreated CHD. Neonates with corrected/palliated CHD were excluded. 154 healthy children, matched for age, were used as controls. BNP was measured by a fully automated platform (Triage-BNP reagents, Access Immunoassay Systems, Beckman Coulter, Inc.). The diagnostic accuracy of BNP was assessed by ROC analysis taking into account 3 groups of age.

Group-1: all CHD patients and controls as a whole (from birth to the 30th day of life); Group-2: 1st-3rd day of life; Group-3: 4th–30th day of life.

**Results:** BNP values were significantly higher ( $p < 0.0001$ ) in CHD than controls (patients: mean  $\pm$  SD  $3429.7 \pm 6165.7$  ng/L, median  $1167.5$  ng/L, range  $25$ – $54447$  ng/L; controls: mean  $\pm$  SD  $189.4 \pm 174.4$  ng/L, median  $150.5$  ng/L, range  $5$ – $866$  ng/L). Group-1: BNP levels were greatly higher in patients than controls. The area under the ROC curve (AUC) was  $0.843$  (SE  $0.02$ ,  $p < 0.001$ ). The BNP cut-off values for rule-out and rule-in were of CHD  $73$  ng/L (sensitivity  $0.95$ ; specificity  $0.34$ ) and  $487$  ng/L (specificity  $0.95$ ; sensitivity  $0.64$ ).

Group-2: BNP levels in patients were higher than controls (AUC  $0.769$ , SE  $0.04$ ,  $p < 0.001$ ). The BNP cut-off values for rule-out and rule-in were  $64$  ng/L (sensitivity  $0.95$ ; specificity  $0.04$ ) and  $531$  ng/L (specificity  $0.95$ ; sensitivity  $0.59$ ).

Group-3. BNP levels were greatly higher in patients than controls (AUC:  $0.935$  SE  $0.02$ ,  $p < 0.001$ ). The BNP cut-off values for rule-out and rule-in were  $66$  ng/L (sensitivity  $0.95$ ; specificity  $0.66$ ) and  $281$  ng/L (specificity  $0.95$ ; sensitivity  $0.75$ ). AUC of Group-3 ( $0.935$ ) was significantly higher than that of Group-1 ( $0.843$ ,  $p = 0.009$ ) and Group-2 ( $0.769$ ,  $p = 0.0003$ ), while the AUC values of Group-1 and Group-2 were not significantly different ( $p = 0.191$ ).

**Conclusions:** BNP may be considered a useful marker in the integrated approach of children with suspected CHD. However, the accuracy of BNP varies greatly during the first month of life, showing the lowest diagnostic accuracy in the first 3 days after birth. After the second week of life BNP becomes more accurate in the ruling in or out CHD.

#### P-13

##### **Percutaneous Transcatheter creation of an aortopulmonary shunt in an animal experimental model**

*Sabi T. (1), Schmitt B. (1), Sigler M. (2), Ovroutski S. (1), Peters B. (1), Miera O. (1), Nagdyman N(1), Kuchne T. (1), Berger F. (1), Ewert P (1).*

*Department of pediatric cardiology, German Heart Institute Berlin, Germany (1); Department of pediatric Cardiology, Georg August University, Goettingen, Germany (2)*

**Introduction:**The surgical creation of an aortopulmonary shunt is an important tool in the therapy of complex congenital heart defects. We evaluated a transcatheter approach to establish an aortopulmonary shunt in an animal model.

**Methods:** In 10 piglets with a median body weight of  $10.5$  kg ( $8$ – $12$  kg), a central aortopulmonary shunt was created by radiofrequency perforation from the aorta to the pulmonary trunk, followed by stent implantation. The procedures were performed via the femoral vessels through  $5F$  sheaths under biplane fluoroscopy guidance. A total of six bare metal coronary stents and five polytetrafluoroethylene-covered coronary stents of  $3$ – $4$  mm diameter were deployed. Four animals were sacrificed immediately after intervention; six pigs were reevaluated  $4$ – $5$  weeks later for stent patency and measurement of shunt volume.

**Results:** The procedure was successful in all piglets. Median shunt volume was Qp:Qs  $5.24:1$ . At re-evaluation median body weight had increased to  $18$  kg ( $15.5$ – $27.5$  kg)  $P < 0.028$ . Four of six stents were completely obstructed due to tissue ingrowth in the bare metal stents (two cases) and thrombus formation in the covered stents (two cases). A third bare metal stent had a residual lumen, and a third covered stent was fully open without any thrombus formation or tissue ingrowth.

**Conclusions:** Transcatheter creation of an aortopulmonary shunt by radiofrequency perforation and stent implantation is feasible. The use of appropriate covered stents and an effective anticoagulatory regimen seem to be crucial to keep the shunts open.

#### P-14

##### **The Relation of Serum Levels of Matrix Metalloproteinases and Their Tissue Inhibitors with Cardiac Functions in Congenital Heart Diseases**

Kilic Z. (1), Ucar B. (1), Erdogan P. (2), Colak O. (3), Demir T. (1), Ertugrul T. (4)

Eskisehir Osmangazi University Faculty of Medicine, Departments of Pediatric Cardiology (1), Pediatrics (2), and Biochemistry (3), Eskisehir, Turkey; Istanbul University Istanbul Medical Faculty, Department of Pediatric Cardiology (4), Istanbul, Turkey

**Introduction:** Matrix metalloproteinases (MMP) are endopeptidases responsible for extracellular matrix (ECM) degradation. Tissue inhibitors of metalloproteinases (TIMP) are specific inhibitors of MMPs. In this study, we aimed to investigate the serum levels of MMP-2, MMP-9, TIMP-1, and TIMP-2 and their relations to ventricular functions in congenital heart diseases (CHDs).

**Methods:** Eighty-seven patients with CHD aged between 3 months–18 years were included in this study. The patients were evaluated by separating them into four groups: right ventricular (RV) volume overload (ASD) (n: 14), left ventricular (LV) volume overload (VSD or PDA) (n: 30), left to right shunt who developed pulmonary hypertension (n: 19), and cyanotic CHDs (tetralogy of Fallot and the other complex cyanotic CHDs) (n: 24) groups. The control group was consisted of 47 healthy children aged between 3 months–18 years. Besides the evaluation of LV systolic and diastolic functions by the conventional echocardiographic methods, systolic and diastolic peak flow velocities of the both ventricles and myocardial performance indexes (MPI) were measured by tissue Doppler echocardiography. Serum levels of MMP-2, MMP-9, TIMP-1, and TIMP-2 were analyzed with enzyme-linked immunosorbent assay (ELISA) method.

**Results:** Serum TIMP-1 levels of the patients with pulmonary hypertension were significantly higher than those of the patients without pulmonary hypertension and than those of the control group ( $905.3 \pm 191.2$ ;  $370.1 \pm 76.7$  and  $344.1 \pm 76.3$  pg/ml, respectively) ( $p < 0.01$ ). There were some differences also between the other study and control groups for serum MMP and TIMP levels, but these differences were not statistically significant. In the patients with left to right shunt associated with pulmonary hypertension, serum MMP-2 levels negatively correlated with Qp/Qs ratio and positively correlated with Rp/Rs ratio; TIMP-1 levels positively correlated with RV MPI; TIMP-2 levels positively correlated with diastolic pressure of the RV. In the cyanotic CHD group, serum levels of both MMP-9 and TIMP-1 positively correlated with Qs but negatively correlated with Rs, however TIMP-2 levels positively correlated with MCV values ( $p < 0.05$ ).

**Conclusions:** TIMP-1 has an important role in the pathogenesis of pulmonary hypertension. MMPs and TIMPs may have a role in the cardiac remodeling observed in CHDs.

#### P-15

##### **Endothelial pro-inflammatory and pro-coagulant activity in a model of S. aureus endocarditis – impact of matrices used in tissue-engineered heart valves**

Heying R. (1,3), Wolf C. (2), Beekhuizen H. (4), Jockenhoevel S. (5), Moelleken M.L. (2), Hoylaerts M. (6), Schmidt K.G. (1), Schrotten H. (2,7)

Department of Paediatric Cardiology (1); Department of Paediatric Infectiology (2); University Children's Hospital, Duesseldorf, Germany; Department of Paediatric Cardiology, UZ Leuven, Belgium (3); Department of Infectious Diseases, LUMC, Leiden, The Netherlands (4); Department of Applied Medical Engineering, RWTH Aachen, Germany (5); Department of Molecular and Cellular Medicine, KU Leuven, Belgium (6); Department of Paediatrics Mannheim, University Heidelberg, Germany (7)

**Introduction:** Staphylococcus aureus is among the most important bacterial pathogens responsible for infective endocarditis. Bacterial metastasis involves the preferential interaction of disseminating bacteria with endothelial cells (ECs) and monocytes leading to subsequent endothelial inflammatory activation and pro-coagulant activity. Especially patients requiring heart valve prosthesis are at high risk to develop endocarditis. Autologous matrix-based tissue engineered heart valves are thought to have excellent potential of tissue remodelling expected to lead to an increased biocompatibility as well as a lower risk of developing endocarditis.

**Methods:** In the present study we compare the influence of a fibrin and collagen gel matrix. ECs were cultivated on the two gels and polystyrene tissue culture plates as a control matrix. Bacterial-endothelial interaction was measured by endothelial adhesion-molecule expression, cytokine production and tissue-factor derived activation of the coagulation.

**Results:** We verified a high capability of S. aureus to infect endothelial cells seeded on the fibrin and collagen matrix compared to ECs cultured on tissue culture plates ( $4.2 \pm 0.5\%$  and  $3.7 \pm 0.8\%$  vs.  $1.2 \pm 0.3\%$  of the inoculation dose;  $p < 0.01$ ). Subsequent endothelial pro-inflammatory response was demonstrated by a marked increase of adhesion molecule expression showing similar results to the values after stimulation with IL-1 without differences between the two matrices and the control surface (ICAM-1: 3.2–3.5 fold increase and VCAM-1 (1.9–2.9 fold increase). Interestingly, S. aureus stimulated production of IL-8 and MCP-1 shows about 30% higher levels after seeding ECs on fibrin gel vs. collagen gel. Despite a similar monocyte adhesion, monocyte enhanced pro-coagulant activity do not differ between the two matrices (2.5 fold increase on fibrin and 2.3 fold increase on collagen gel) but showed a marked decreased endothelial coagulation activity compared to results on tissue culture plates (10.2 fold increase).

**Conclusion:** The investigated underlying gel matrix seems to have an impact on endothelial activation. Current further investigations might state if the surface structure of the matrices influence monocyte-endothelial contact and subsequent interaction to evoke pathways of inflammation, tissue damage and fibrin deposition at the infected endovascular sites.

#### P-16

##### **Hypothermia decreases apoptosis after oxidative stress in cardiomyocytes**

Drescher C. (1), Diestel A. (2), Berger F. (1,2), Schmitt K. (1) Department for Congenital Heart Disease and Pediatric Cardiology, Deutsches Herzzentrum Berlin, Germany (1); Pediatric Cardiology, Charité-Universitaetsmedizin, Berlin, Germany (2)

**Background:** Hypothermia protects organs during cardiopulmonary bypass (CPB) in pediatric heart surgery. Therefore we investigated hypothermia-mediated cell protection in H<sub>2</sub>O<sub>2</sub> damaged cardiomyocytes.

**Methods:** For hypothermic treatment we cooled cardiomyocytes to 20°C and maintained 20°C for 20 min, during which short-term

oxidative damage was inflicted with 200  $\mu$ M H<sub>2</sub>O<sub>2</sub>, followed by rewarming to 37°C. Later on we analyzed cell viability (trypan blue staining), lactate dehydrogenase release (LDH assay), caspase cleavage (western blot technique), M30 CytoDeath expression (immunohistochemistry), mitochondrial activity (MTT assay) and DNA damage via H2AX (western blot technique).

**Results:** Hypothermia decreased H<sub>2</sub>O<sub>2</sub> damage in cardiomyocytes as demonstrated in higher cell viability and lower LDH release. After rewarming H<sub>2</sub>O<sub>2</sub> damaged cells demonstrated significantly higher cell viability compared to normothermic H<sub>2</sub>O<sub>2</sub> damaged cardiomyocytes. This was in line with less apoptosis as shown by diminished caspase cleavage and significantly lower M30 CytoDeath staining in cooled and rewarmed cells. Moreover, hypothermia preserved cell viability by up-regulation of the anti-apoptotic protein Bcl-2. Phosphorylation of histone H2AX on Ser 139 as a sensitive reporter of DNA damage was significantly reduced after hypothermia and rewarming. Therefore, this finding indicates that deep hypothermia at 20°C not only protects the mitochondria from oxidative stress but also defends the DNA against oxidative damage after H<sub>2</sub>O<sub>2</sub> treatment, resulting in decreased apoptosis.

**Conclusion:** In this cell culture study we showed that hypothermia protects cardiomyocytes during H<sub>2</sub>O<sub>2</sub> treatment by preventing apoptosis via inhibiting mitochondrial dysfunction and DNA damage.

#### P-17

##### **Deep hypothermia prevents inflammation but leads to cellular stress in a coculture model of endothelial cells and macrophages**

*Wöllersheim S. (1), Diestel A. (2), Berger F. (1,2), Schmitt K. (1)*  
Department for Congenital Heart Disease and Pediatric Cardiology, Deutsches Herzzentrum Berlin, Germany (1); Pediatric Cardiology, Charité-Universitätsmedizin, Berlin, Germany (2)

**Background:** During cardiac surgery with cardiopulmonary bypass (CPB), patients are often cooled to a temperature of 14–18°C in order to protect organs from ischemia. Cardiopulmonary bypass triggers a systemic inflammatory response. We hypothesized that dynamic temperature changes may influence this inflammation. The aim of our studies was therefore to investigate the effects of deep hypothermia and rewarming on the inflammatory and cellular stress response in a coculture model of endothelial cells and macrophages.

**Methods:** Primary human umbilical vein endothelial cells (HUVECs) and macrophages (THP-1 cell-line) were exposed to dynamic temperature changes analogous to the clinical settings found during pediatric cardiac surgery: 20 min deep hypothermia (20°C), slow rewarming (30 min up to 37°C) and normothermia (72h at 37°C). To imitate inflammatory response after CPB, the coculture was stimulated with 500 U/ml TNF- $\alpha$ . Cell viability, expression of IL-6, IL-8 and MCP-1 as well as intracellular reactive oxygen species (ROS) and adenosine triphosphate (ATP) content were investigated.

**Results:** Deep hypothermia had no influence on cell viability but suppressed inflammation, as shown in lower IL-6 and IL-8 release and significant MCP-1 release. Moreover, hypothermia led to an increase in intracellular ROS in the comparison with the stimulated normothermic coculture. This is in line with a significant decrease in ATP content.

**Conclusion:** In a new coculture cell model of simulated CPB, hypothermia diminishes the inflammatory response. Otherwise, hypothermia and rewarming increase mitochondrial oxidative stress combined with a decrease in intracellular ATP content after

24 hours. Therefore, hypothermic CPB should be performed with caution as underlying hypothermia induced mechanisms are not fully understood.

#### P-18

##### **Fibroblasts facilitate the engraftment of embryonic stem cell-derived cardiomyocytes on three dimensional collagen I matrices**

*Attia W. (1,3), Pfannkuche K. (2), Hescheler J. (2), Brockmeier K. (3)*  
Clinic of Pediatric Cardiology, Cairo, Egypt (1); Institute for Neurophysiology, Cologne, Germany (2); Clinic of Pediatric Cardiology, Cologne, Germany (3)

There is growing interest in purified cardiomyocytes derived from embryonic stem cells for in vitro or in vivo tissue engineering. Up to date, the role of non-cardiac cells in cardiomyoplasty remains elusive. We established a three-dimensional tissue culture model based on a freeze-dried collagen matrix with tubular structure, which was seeded either with highly purified cardiomyocytes, that have been purified by genetic selection, alone or in combination with fibroblasts. The collagen sponges that were transplanted with embryonic stem (ES) cell derived cardiomyocytes alone did neither show morphological nor functional integration of the cells. However, when cocultured with embryonic fibroblasts cardiomyocytes formed fibre-like structures of rod-shaped cells with organized sarcomeric structure. Electrical coupling between cardiomyocytes was demonstrated by strong expression of connexin43. We conclude that fibroblasts are needed for morphological and functional engraftment of purified cardiomyocytes on collagen matrices.

#### P-19

##### **Closure of Muscular Ventricular Septal Defects with a New Self Centering Patch System in a Large Animal Model**

*Lang N. (1), Mever A. (1), Merkel E. (1), Thalmann R. (1), Lehner A. (1), Freudenthal F. (2), Vasilyev N. (3), DelNido P. (3), Netz H. (1), Kozlik-Feldmann R. (1)*  
Kinderherzzentrum, Ludwig-Maximilians-Universität, München, Germany (1); Kardiozentrum, La Paz, Bolivia (2); Children's Hospital /Harvard Medical School, Cardiac Surgery, Boston, USA (3)

**Objective:** Therapies for closure of muscular ventricular septal defects (mVSDs) beyond the moderator band have still limitations. Recently, we established a new hybrid technique for closure of mVSDs on the beating heart. Here, we evaluated a self-centering patch system for this technique.

**Methods:** 9 pigs underwent left anterolateral thoracotomy to expose the left ventricle (LV). mVSDs were created under epicardial echocardiographic control. The self-centering patch system consists of a removable centering system and a releasable attached patch. After creation of an arterio-venous circuit, an implantation sheath was forwarded from the venous side via the guide wire into the LV. The system was loaded, advanced and deployed in the LV and pulled back into the VSD. Then, the patch was attached to the septum with Nitinol anchors under epicardial echocardiographic and fluoroscopic guidance. Finally, the patch system is drawn back into the sheath. All hearts were explanted for macroscopic evaluation.

**Results:** Closure of mVSDs was successful in 6 out of 9 pigs without residual shunting. Macropathology revealed adequate positioning and fixation of the patch. In three cases, the patch is fixated to the delivery system by nitinol anchors. Consequently,

the patch could not be detached from the delivery system. After using smaller anchors, this complication did not occur anymore. **Conclusions:** Closure of mVSDs can be successfully performed with our new patch system. A big advantage is the self centering mechanism. However, further modifications and improvement of imaging is needed before clinical application is possible.

#### P-20

##### **Pulmonary artery banding as a novel therapeutic option for dilative cardiomyopathy (DCM): Thoughts about interaction between RV and LV in children with isolated RV pressure load due to pulmonary valve stenosis.**

Mueller G.C. (1), Arndt F. (1), Eman H.M. (3), Schaefer A.-L. (1), Mir T.S. (1), Dodge-Kathami A. (2), Weil J. (1)

Department of Paediatric Cardiology, University Heart Center Hamburg, Germany (1); Department for Paediatric Cardiac Surgery, University Heart Center Hamburg, Germany (2); Azadi General Hospital, Kirkuk, Iraq (3)

**Introduction:** Pulmonary artery banding is reported to be a possible innovative treatment option in DCM (Schranz et al., 2007). The beneficial effect could be caused by the shift of the interventricular septum into the left ventricle and the induction of left ventricular myocardial hypertrophy. Children with isolated pulmonary valve stenosis (PS) could serve as a useful natural model to study the effect of chronic right ventricular pressure load on left ventricular geometry and function.

**Method:** Examination of 10 children (15–216 months) undergoing balloon valvuloplasty for isolated PS by invasive measurements and echocardiography before and after intervention.

**Results:** RV-pressure was significantly elevated in all patients (65–180 mmHg; mean 116 mmHg). Echocardiographic measurements showed a significant correlation of increasing right ventricular pressure load and increasing thickness of left ventricular posterior wall ( $r = 0,783$ ;  $p < 0,01$ ). Left ventricular enddiastolic diameter decreases significantly due to increasing right ventricular pressure load ( $r = -0,776$ ;  $p < 0,01$ ).

**Conclusion:** Left ventricular myocardial growth and left ventricular geometry are significantly affected by right ventricular pressure load in this study population. These interactions of right and left ventricle could be reasons for the reported successful treatment of DCM by pulmonary artery banding.

#### P-21

##### **Ventricular assist devices: initial experience in a spanish center with the Berlin Heart EXCOR**

García-Guereta L., Aroca A., Villar MA., Menéndez JJ., Carton A., Polo L., Oliva P., Cerro MJ., Rodríguez E., Gutiérrez-Larraya F, Villagrà F

Hospital Universitario La Paz. Madrid. Spain

Ventricular assist devices are replacing ECMO in the treatment of children with refractory heart failure as a bridge to transplantation. Berlin Heart (BH) EXCOR® provides different sized pumps to treat children of any age. We report our initial experience with BH EXCOR® in critically ill children with refractory heart failure.

**Method:** Retrospective review of indications, type of device (left ventricular -LVAD- or biventricular -BiVAD-), length of assistance and outcome.

**Results:** From 10/2006 to 11/2009 5 systems were implanted, 4 BiVAD and 1 for a univentricular heart. Indications were dilated cardiomyopathy (3), restrictive cardiomyopathy with pulmonary

hypertension (1) and failed fontan (1). There was no surgical mortality. Patient #1 could not be weaned from bypass because of severe hypoxia due to pulmonary hemorrhage. A membrane oxygenator (Jostra Cuadro®) was successfully intercalated in the LV outflow cannula, and withdrawn after 5 days when pulmonary function was stable. A LVAD was inserted to a single ventricle patient 9 year after Fontan who developed cardiogenic shock after influenza H1N1 infection. 3/5 patients were bridged to transplantation after 10 to 210 days of assistance. They are all now in functional class I. Patient #2 had a complex course with brain haemorrhage, multisystemic failure and massive hemorrhagic enterocolitis after an enteroinvasive E Coli infection; there was asystolia and the child was disconnected from the system. Patient #5 recovered from influenza infection but did not recover contractility. He had severe persistent thrombocytopenia and frequent bleeding despite withdrawal of anticoagulation drugs; he had severe intracranial bleeding and EXCOR was discontinued.

	Age	Diagnosis	Type	ECMO	Extubation	Days	Outcome
1	5 y	DCM	BiVAD	10 d	11 d	210	Heart Transpl.
2	3 y	RCM + PHT	BiVAD	no	14 d	90	Exitus
3	7 m	DCM	BiVAD	10 d	15 d	24	Heart Transpl
4	5 m	DCM	BiVAD	10d	6 d	10	Heart Transpl
5	14 y	Failed Fontan	LVAD	6d	-	20	Exitus

**Conclusions:** Berlin Heart EXCOR is a very useful device to bridge critically ill children, including infants, to transplantation. Neurological and hemorrhagic sequelae are frequent and limit recovery or transplantation.

#### P-22

##### **Eisenmenger Syndrome and Bosentan Therapy: a single centre experience**

Agnetti A., Tchana B., Carano N., Allegri V., Bo I., Dutto M., Quadrini I., Macchi C., Al-Obeyde M., Squarcia U.

Paediatric Cardiology Unit – Department of Paediatrics- University of Parma, Italy

**Background:** Eisenmenger syndrome, the most advanced form of pulmonary hypertension associated with congenital heart defects, is a condition that has a severe effect on patients clinical status and quality of life. Bosentan, an orally active dual endothelin – receptor antagonist, has shown to improve symptoms, exercise capacity and hemodynamics in patients with pulmonary hypertension secondary to congenital heart disease, with the same safety profile as reported in other groups of patients with pulmonary hypertension. We present a single centre experience.

**Objective:** The purpose of this study was to investigate the therapeutic effects of Bosentan in patients with Eisenmenger syndrome.

**Materials and methods:** In this single centre study, 7 patients with Eisenmenger syndrome (3 with Down syndrome and 4 without) were treated with Bosentan (62.5 mg twice daily for 4 weeks then 125 mg twice daily). Their mean age was 36.7 years. All patients were evaluated at baseline and during follow-up with laboratory tests, six-minute walk test (6-MWT), Doppler echocardiography, strain, strain rate and hemodynamic study in cathlab.

**Results:** The follow-up of the patients treated with Bosentan ranges from 15 to 30 months. Induction and treatment with Bosentan was well tolerated. No serious adverse drug reactions were noted. The 6-MWT showed an increase of 45.6 meter ( $p = 0.05$ ) after 6 months and of 44.2 meter ( $p < 0.05$ ) after 12 months, whereas at 3 months the result was not significant. On echocardiography in one patient with Down syndrome

after 6 months of treatment a low velocity flow appeared on Doppler in main pulmonary artery, most likely a patent ductus arteriosus which became evident with reduction of the pulmonary resistances following the Bosentan therapy.

**Conclusions:** Our observations confirmed the significant response to Bosentan treatment in patients with Eisenmenger syndrome. Even Down syndrome patients, in which pulmonary hypertension has been suggested to develop earlier with a more aggressive course, may benefit from Bosentan treatment.

#### REFERENCES

- Duffels MG et al. *Int J Cardiol.* 2009 May 29;134(3):378–83.  
 Duffels MG, Vis JC, et al. *Int J Cardiol.* 2008 Jun 23.  
 Galie N, Beghetti M et al. *Circulation* 2006; 114: 48–54

#### P-23

##### **15 year follow-up in renal function after pediatric heart transplantation in a single center**

*Behnke-Hall K., Bauer J., Thul J., Logeswaran T., Horn M., Akintürk H., Valeske K., Schranz D. Pediatric heart center, Giessen, Germany*

**Introduction:** Survival after pediatric heart transplantation (HTX) has improved substantially in the last decades. However, the long term outcome is still affected by morbidities mostly caused by side effects of immunosuppressive therapy. Among these, renal dysfunction is an emerging problem before and after transplantation. In the post transplantation period, the nephrotoxicity of calcineurin-inhibitors (CNI) especially influences the long term outcome. Therefore, we analyzed the long term outcome of renal function, in our patients group.

**Methods:** We estimated the glomerular filtration rate (GFR) by Schwarz-formula at time of HTX, at 6–12 months, and then every 5 years up to 15 years after HTX. Results were analyzed in relation to time after HTX, immunosuppressive therapy, age at time of HTX, diagnosis leading to HTX and gender.

**Results:** Starting 1988, 99 patients (pts) who are still in present follow-ups, were included (Group A), 36 of which are female. Median age at HTX was 2.6 (+/-4.44) years with 60 pts less than age 2. All pts received CNIs since HTX. 23 of the 99 pts were later converted to an mTOR-Inhibitor (Group B). Mean GFR at time of HTX was reduced (72.09 (+/- 34.0)ml/min/1.73 m<sup>2</sup>) and improved 6–12months to a mean GFR of 87.32 (+/-26.32)ml/min/1.73 m<sup>2</sup> and remained stable at 5, 10 and 15 years after HTX (84.66 (+/- 20.32), 78.99 (+/- 21.9) and 88.75 (+/-20.57)ml/min/1.73 m<sup>2</sup>) in all pts. Male gender and older age at HTX were associated with a better outcome. Diagnosis leading to HTX had also an impact on renal function, with the best outcome in pts with cardiomyopathy. In group B, GFR increased at least 6 months after conversion from 69.67 (+/- 17.39) to 96.04 (+/-24.09)ml/min/1.73 m<sup>2</sup>.

**Conclusion:** Renal dysfunction has an important impact on morbidity after HTX. Most of the patients have a mild renal insufficiency and remain stable despite CNI therapy. Change in immunosuppressive therapy to an mTOR-Inhibitor can improve renal function.

#### P-24

##### **Recurrence rate of cardiac disease in cases with family history of cardiomyopathy referred for fetal echocardiography**

*Fesslova V (1), Brankovic J. (1), Piazza L. (1), Villa L. (2), Meli V. (3)*

*Dpt. Ped. Cardiology/Center of Fetal Card., Policlinico San Donato IRCCS, Milan, Italy (1); 1.st Gyn.Obstet.Clinic, University of Milan (2); Dpt. Materno-Infantile, Policlinico Universitario, Palermo, Italy (3)*

**Background:** The large variety of onset and presentation of different types of cardiomyopathies (CMP) makes very difficult to assess the possibility of recurrence in fetal life, in cases with positive familial history.

**Objectives:** to evaluate the recurrence of cardiac disease in fetuses of women with familial risk of dilated or hypertrophic cardiomyopathy (DCMP, HCMP), screened prenatally.

**Material and methods:** retrospective/prospective study of 70 pregnancies in 65 pregnant women with family history of CMP referred between Jan. 1995 – July 2008 for fetal echocardiography. Forty women, in 45 pregnancies had family history of HCMP (single risk 29 index cases, multiple risk 16 in 2–5 relatives); affected mother in 10, in 5 with another relative, father in 4, in one with another relative, 1 previous child in 13 and 2 previous children in 3 cases, distant relatives in 12 (9 with 2–5 relatives). Family history of DCMP was present in 25 women, 3 affected, with multiple risk in one; father was affected in 3, previous child in 8, distant relatives in 11 cases (with 2–3 relatives in 6). Fetal diagnosis was compared with postnatal findings at 6–12 months and diseased infants were followed-up for a median of 5 years (2–12).

**Results:** Group HCMP: 5/45 (11.1%) showed HCM already in utero at 22–35 w.g.: 2/10 (20%) of affected mothers, 2/17 (11.8%) with 1–2 previous children affected and 1/12 (8.3%) with 2nd degree relatives affected. Another fetus, with father and a 2nd degree relative affected – had tricuspid atresia. Of 16 cases with multiple risk 1 proband was affected.

Group DCMP: No form of DCMP was revealed, while one proband with index case of a distant relative had univentricular heart (1/25 = 4%). A total recurrence rate for cardiac anomaly in the population was 7/70 (10%).

**Conclusions:** Our limited data show a recurrence rate of 11.8–20% in fetuses with family history of HCMP in first degree relatives and no recurrence in a smaller series with family history of DCMP; interestingly, two probands with both forms of CMP had congenital heart defects, to suggest a possible genetic link between the two types of cardiac anomalies.

#### P-25

##### **Does Prenatal Anti-inflammatory Treatment prevent from Late-Onset Dilated Cardiomyopathy (DCM) in Children with Immune-Mediated Congenital Complete AV Block (CAVB)? Results of a Retrospective Multicenter Experience**

*Hutter D.M. (1), Hornberger L. (2), Cuneo B. (3), Fouron J.-C. (4), Grady A. (2), Friedberg M. (1), Silverman N. (5), Jaeggi E. (1) The Hospital for Sick Children, Toronto, Canada (1); San Francisco Children's Hospital, San Francisco, USA (2); Heart Institute for Children, Hope Children's Hospital, Chicago, USA (3); Sainte-Justine Hospital, University of Montreal, Montreal, Canada (4); Pediatric Heart Centre, Stanford, Palo Alto, USA (5)*

**Background:** Maternal anti-Ro/La antibodies may trigger inflammation of the atrio-ventricular (AV) node and the myocardium in the susceptible fetus. The inflamed tissue then may heal with fibrosis which may lead to heart block and endocardial fibroelastosis (EFE). Of particular concern is the development of myocardial dysfunction months to years after birth in a subset of patients with CAVB and apparently normal cardiac function at

birth: 5–10% of untreated fetuses will develop DCM during childhood. We postulated that the risk of late-onset DCM is reduced with in-utero anti-inflammatory treatment.

**Methods:** We reviewed the findings, management and outcome of 70 treated cases (mean age: 24.3 weeks) with a prenatal diagnosis of antibody-associated CAVB since 1990. Prenatal treatment included maternal administered dexamethasone (dexa) to birth, addition of a  $\beta$ -sympathomimetic ( $\beta$ S) if fetal heart rates declined below 50–55 beats/minute, and, more recently, intravenous immune-globuline (IVIg) if EFE was detected. Late-onset DCM was defined as progressive left ventricular dilatation in association with progressive cardiac dysfunction.

**Results:** Of the 70 cases, 46 had isolated CAVB and 24 also had EFE. Prenatal treatment included dexa (N = 67; 96%),  $\beta$ S (N = 26; 37%), and/or IVIg (N = 14; 17%). At a mean follow-up of 5.9 years, 42 (60%) underwent implantation of a permanent pacemaker and 81% remain alive. None of the deaths in the treatment group was related to late-onset DCM. The table shows the management and outcome of fetuses with prenatal treatment.

Treatment	N	Paced	Unpaced	EFE fetal	EFE last echo	Death
Dexa	36	21	11	7	3	2 (6%)
IVIg	2	2	0	2	2	0 (0%)
$\beta$ S	1	1	0	1	1	1 (100%)
Dexa + $\beta$ S	17	12	2	5	0	4 (24%)
Dexa + IVIg	6	2	1	4	2	0 (0%)
Dexa + $\beta$ S + IVIg	8	4	0	5	4	6 (75%)
Total	70	42	14	24	12	13 (18%)

**Conclusions:** Of the prenatally treated cases with CAVB none developed late onset DCM in this study. Fetal survival was poor for cases presenting extensive EFE and low heart rates despite treatment with dexa, IVIg and  $\beta$ S.

Perinatal survival was poor for cases presenting low heart rates and extensive EFE despite treatment with dexa,  $\beta$ S and IVIg. Of fetal and neonatal survivors none developed late onset DCM to this day.

## P-26

Withdrawn

## P-27

### Can mid-trimester Doppler flow echocardiography predict an abnormal postnatal ECG?

Sonesson S.-E. (1), Bergman G. (1), Wahren-Helenius M. (2)

Department of Women's and Children's Health (1); Department of Medicine (2); Karolinska Institutet, Stockholm, Sweden

**Objectives:** Having recently demonstrated that the isovolumetric contraction time (ICT) was an important contributor to prolongation of atrioventricular (AV) time intervals, this study was performed to investigate the diagnostic precision of three Doppler methods, two including and one excluding the ICT, in their ability to predict postnatal first-degree (AV) block.

**Methods:** Prospective, observational study from December 1999–March 2008 including 95 fetuses of anti-SSA/Ro positive mothers, examined with weekly fetal echocardiograms at 18–24 weeks of gestation. Doppler derived AV time intervals measured on recordings from left ventricular inflow (MV), inflow and outflow (MV-Ao) and superior vena cava and aorta (SVC-Ao) were compared with the PR interval in postnatal ECG. Reference values for MV intervals were established from

102 healthy fetuses. Another 284 fetuses served as controls for the two other methods. Bayesian analysis was performed and ROC curves constructed.

**Results:** Newborns with first-degree AV block in ECG had longer mid-trimester AV time intervals, using all three Doppler methods, than those with sinus rhythm. Prevalence of first-degree AV block at birth was 13.8%. MV-Ao and SVC-Ao time intervals with a 95% reference range had a sensitivity of 91.7%, NPV and LR- of 98.4% and 0.10 respectively. Corresponding PPV and LR+ values for MV-Ao and SVC-Ao were 42.3%, 4.5 and 47.8%, 5.7 respectively. ROC curve AUC for MV-Ao and SVC-Ao were 0.87 and 0.89 ( $p < 0.0001$ ) with generated optimal cut offs for AV time intervals at 134–138 and 132–138 ms. MV time intervals with a 95% reference range had a sensitivity of just 50% and an AUC of 0.75 ( $p < 0.01$ ).

**Conclusions:** The MV-Ao and SVC-Ao Doppler methods make it possible to identify nearly all fetuses with first-degree AV block at birth and to exclude conduction disturbances in the case of a normal AV time measurement but at the cost of a positive predictive value of 50%.

## P-28

### Gene Expression In Atrioventricular Junction From Fetal Hearts

Hamilton M. R. (1), Su B.B. (1), Lung K. (1), Capocchi M. (2), Yan J.Y. (1), Cui R. (1), Liu L. (1)

The Hospital for Sick Children, Toronto, Canada (1); University of Pisa, Pisa, Italy (2)

**Introduction:** Congenital heart block (CHB) is a frequent complication of pregnancies to autoimmune mothers. CHB should be an entirely preventable condition, but knowledge of the pathogenesis of CHB is lacking. Up to now, there are very few publications assessing fetal heart gene expression, and the specific gene expression profile in human fetal heart conductive tissue has not reported. In this study we identify genes differentially expressed in specific tissue atrioventricular junction (AVJ) as compared to apex (Apx) microarray.

**Methods:** RNA was extracted using Qiagen RNA Fibrous Tissue Mini kit and quantified by NanoDrop 2000 and Agilent 2100 BioAnalyzer. Total RNA were applied on Affymetrix human U133 plus 2 arrays and the data were confirmed on Real-Time PCR using the same samples for microarray. The microarray data was analyzed by Parquet Genomic Suite.

**Results:** We found 3012 genes specifically expressed in AVJ in the human fetal hearts at 20–22 weeks of gestation ( $p < 0.05$ ). There are 703 genes with fold changes either greater than 2 (588 genes) or less than 2 (115 genes). A total of 85 genes found with fold change either great than 2 or less than 2 with significantly different expression in AVJ compared to Apx ( $p < 0.05$ ). Gene ontology analysis scored 34 genes for molecular function and 33 genes for biological process with fold changes great than 2. We compared these 85 significant genes from AVJ of human fetal hearts to mouse AV canal/node cells that recently reported by Horsthuis et al. (Horsthuis et al., *Circ. Res.* 2009;105:61–69), we found genes SLC26 family of transporter member 7, transcription factor T-BOX 3 and potassium channel-interacting protein with different splice variants expressed on AV conductive tissue in both mouse and human. Some genes from the microarray data were confirmed on Real-Time PCR.

**Conclusions:** up-regulation of developmental genes in AVJ than Apx are responsible for structure biological process and for the extracellular matrix organization. High-throughput technique is a good tool to identify candidate targets for CHB.

**P-29****Prenatal diagnosis and outcome of fetuses with transposition of the great arteries (TGA)**

Rasih S.V. (1), Ewer A.K. (1), Tonks A. (2), Miller P. (3), Wright J.G. (3), Kilby M.D. (1)

Birmingham Women's Hospital NHS Foundation Trust, Birmingham, United Kingdom (1); West Midlands Perinatal Institute, Birmingham, United Kingdom (2); Birmingham Children's Hospital NHS Foundation Trust, Birmingham, United Kingdom (3)

*Introduction or Basis or Objectives:* To understand the spectrum and outcome following prenatal diagnosis of transposition of the great arteries (TGA).

*Methods:* Over an 8-year period all pregnancies in which the fetus was identified as having TGA on ultrasound examination at a tertiary fetal medicine centre were reviewed. This data was compared with the regional Congenital Anomalies Register (CAR).

*Results:* Prenatally, 75 fetuses were diagnosed with TGA. 16 cases were classified as simple (with intact septum or ventricular septal defect only). The remaining 59 were classified as complex i.e. they had additional intra-cardiac malformations. Following prenatal counselling, 18 (24%) couples chose termination of pregnancy. Of the 57 (76%) continuing pregnancies, 3 were stillbirths and 5 were lost to follow-up. Of the 49 live births, 7 were neonatal deaths without surgery. 42 babies underwent surgery and 34 have survived to date. At the time of prenatal diagnosis, the overall survival for simple and complex TGA were 92.8% and 55.3% respectively (excluding terminations and lost to follow-up). During the same period, there were 198 cases of TGAs identified by the Regional CAR. Of these 122 were simple and 76 were complex based on the above definition. Only 11% of the simple TGAs were identified prenatally. The one year survival was 92.2% for simple and 64.3% for complex TGAs (excluding terminations).

*Conclusions:* The majority of prenatally diagnosed TGAs were complex and these lesions had a poor overall outcome. The antenatal detection rate for simple TGAs is very low. This emphasises and supports the importance of including outflow tract assessment during mid-trimester scanning.

**P-30****Diagnosis and outcome of prenatally diagnosed atrio-ventricular septal defect**

Kraska A. (1), Własienko P. (2), Kuran J. (2), Dangel J.H. (2)

(1) Students' Scientific Group of Perinatal Cardiology, Medical University of Warsaw; (2) Perinatal Cardiology Unit, 2nd Dept. of Obstetrics and Gynecology, Medical University of Warsaw, Poland

*Objectives:* AVSD is the second most common congenital defect in prenatal life. Knowing this the aim of this study was to evaluate frequency of chromosomal aberrations (ChA), other intracardiac and extracardiac malformations (ECM) in relation to types of AVSD in fetuses in two periods of time: until and after 2002, due to different organization of cardiac perinatal care.

*Methods:* Retrospective analysis of computer database and exams evaluation of a Reference Centre for Perinatal Cardiology between 1995–2009. Out of 144 fetuses with AVSD 49 with heterotaxy syndrome were excluded. We evaluated 95: 20 until 2001, 75 after 2002.

*Results:* Mean maternal age was 30 yrs. Mean GA: 28 weeks (+/- 6,6 SD). Only 21 fetuses (22%) had isolated AVSD, 13 (14%) had extracardiac malformations, 61 (64%) had chromosomal anomalies (T21-48, T18-10) and among them 19 had also

ECM. 75 (79%) fetuses had balanced AVSD, 51 (68%) of them with chromosomal abnormalities. 20 (21%) had unbalanced AVSD, and 10 (50%) of them had chromosomal aberrations. Before 2002 3 (14% of all, 75% diagnosed before 24 weeks) pregnancies were terminated. All of them had ECM and 2 also T18. Out of the rest 17 fetuses 2 died in utero, 15 were live-born. 11 died in the neonatal period, 4 were operated on (3 survived). Since 2002 15 of 27 (20% of all, 56% of those diagnosed before 24 WOG) were terminated. These were fetuses with ChA10, ECM 2, ChA+ECM 2 and one with normal karyotype. Out of 60: 6 fetuses 7 (10%) died in utero; 50 (69%) were live-born. 4 died in the neonatal period, 33 were operated on, 30 survived, 13 are scheduled for treatment. In 3 pts follow-up is unknown.

*Conclusions:* AVSD is a strong sonographic marker of chromosomal abnormalities. Chromosomal aberrations, mostly T21, were more common in fetuses with balanced than unbalanced AVSD, however our research did not confirm that unbalanced AVSD was associated with normal karyotype. We noticed a significant rise of survival rate in children with AVSD over the last 7 years what is due to an improvement of the perinatal care.

**P-31****Fetal critical aortic stenosis – with or without pharmacological transplacental treatment?**

Moszura T. (2), Michalak K.W. (2), Respondek-Liberska M. (1), Janiak K. (1), Ostrowska K. (2), Sysa A. (2)

Department of Diagnosis and Prevention of Congenital Malformations, Polish Mother's Memorial Hospital in Lodz, Poland (1); Cardiology Department, Polish Mother's Memorial Hospital in Lodz, Poland (2)

Critical aortic stenosis is a heart defect, which occurs in 3% of all congenital heart defects among newborns, but probably is more frequent in fetuses as not all of them can reach term delivery.

Current procedure of choice for critical aortic stenosis in percutaneous balloon aortoplasty just after delivery and in selected cases the only way of successful treatment is Norwood procedure. There is also a possibility of transplacental, prenatal pharmacological treatment, which can improve cardiac function and prevent heart failure before and after the delivery. The aim of our study was to assess one centre experience in pharmacological transplacental treatment of critical aortic stenosis. 28 consecutive fetuses with critical aortic stenosis were divided into two groups – first one (14 pts) consist of patients with early diagnosis and echocardiographic monitoring and pharmacologic transplacental treatment in prenatal period; in second group subjects were not treated in utero – because of late diagnosis or short monitoring period. On the basis of each fetal echocardiographic examination the following indexes were evaluated (HA/CA, Ao flow, LVSF, RVSF, RA SF, LA SF, Tein index LV, Tein index RV) and each index changes during follow up were measured. Patients in first group were monitored and in each case transplacental digoxin treatment was introduced. After the delivery the newborns were transported within the same hospital to the cath lab, where percutaneous aortoballoonoplasty was performed. The general mortality in first group (27%) was statistically significant lower than in a second (57%) ( $p = 0,047$ ). Time between delivery was shorter in a first group (average 14h, standard deviation 3,7h) than in a second one (average 37,4h, SD=8,7h) ( $p < 0,01$ ). In our material increase of reduce fraction of right ventricle (RFRV) was correlated with the length of pharmacologic treatment in utero ( $p < 0,01$ ). In first group increase of heart area to chest area ratio (HA/CA) was statistically significant lower than in a second group ( $-0,03$  Gr. 1;  $+0,08$  Gr.2) ( $p = 0,034$ ). Received results showed positive influence of



pharmacotherapy in prenatal period on late results of interventional treatment in children with critical aortic stenosis and give reasons for further study to establish optimal doses and time of drug administration.

### P-32

#### **An elevated value of a drug-induced lymphocyte stimulation test for immunoglobulin is one of the immunological abnormalities of Kawasaki disease.**

Nomura Y. (1), Masuda K. (2), Eguchi T. (1), Morita Y. (2), Arata M. (1), Ueno K. (1), Kawano Y. (1)

Division of Pediatrics, Kagoshima University Graduate School of Medical and Dental Sciences, Kagoshima, Japan (1); Department of Pediatrics, Kagoshima Medical Association Hospital, Kagoshima, Japan (2)

**Background:** Kawasaki disease (KD) is an acute febrile vasculitis in childhood. Currently, treatment with 2g/kg of intravenous immunoglobulin (IVIG) is recommended to reduce the incidence of coronary artery abnormality. We encountered a patient with KD who showed persistent fever and a severe eruption after IVIG treatment. Using a drug-induced lymphocyte stimulation test (DLST), he was positive for an immunoglobulin product. His DLST values for material contained by immunoglobulins were not elevated. A positive DLST value for the immunoglobulin product observed in our patient may indicate an allergic reaction to immunoglobulin products, but it may also represent an immunological abnormality of KD. To illuminate this issue, DLST for immunoglobulin products were examined in KD patients. The aim of this study was to clarify the importance of a positive DLST value for immunoglobulin products in KD patients.

**Methods:** Subjects were 30 confirmed KD patients treated with IVIG at the Kagoshima Medical Association Hospital. DLST values were compared between patients with additional events and those without additional events using stimulation index (SI). The SI was calculated by the following formula and the SI value beyond 180% was defined as positive:  $SI = \text{value of } ^3\text{H-thymidine absorption of patient's mononuclear cells with antigen/without antigen}$ . Additional events were defined as symptoms observed after IVIG that were considered unexplainable by the symptoms of KD alone.

**Results:** DLST results were evaluated in 13 patients with additional events and 17 patients without additional events. DLST for immunoglobulin products were examined on day  $13 \pm 8$  after IVIG initiation. Elevated DLST values were observed not only in patients with additional events but also in those without additional events. Elevated SI values were observed in initial 14 days from IVIG and the SI values in this period were significantly higher than those after day 14 (initial 14 days,  $n = 20$ ,  $194 \pm 112\%$ , after day 14,  $n = 10$ ,  $117 \pm 66\%$ ,  $P = 0.010$ ).

**Conclusions:** Elevated SI values of DLST for immunoglobulin products are not related with additional events. Our results show they may be one of the immunological abnormalities of KD.

### P-33

#### **Dynamics of Oxidative Stress in the Chronic Stage of Kawasaki Disease**

Yahata T., Suzuki C., Hamaoka A., Fujii M., Ozawa S., Hamaoka K. Department of Pediatric Cardiology and Nephrology, Graduate School of Medical Science, Kyoto Prefectural University of Medicine, Kyoto, Japan

**Background:** It has recently reported in many studies that patients in the chronic stage of Kawasaki disease (KD) are likely to show defective endothelial function. Such patients show activation of

inflammatory and smooth muscle cells and increased oxidative stress. Increased activity of the antioxidant systems is one of the homeostatic mechanisms that function against increased oxidative stress in vivo. Few studies have been conducted on the dynamics of oxidative stress in KD patients and the results have been rather unclear.

**Subjects and methods:** The subjects were 27 outpatients (age,  $13.5 \pm 8.1$  years) who had had the history of KD for the last 5 years. They were divided into the following 3 groups: (a) 7 patients without coronary artery lesions (CALs), (b) 6 patients with temporary CALs, and (c) 14 patients with CALs. The oxidative stress regulation system was analyzed on the basis of the following variables; d-ROM (reactive oxygen metabolites) test and BAP (biological antioxidant potential) test using FRAS4 (Free Radical Analytical System). In this study, we evaluated the ratio of oxidative stress state by using the modified BAP/d-ROM ratio (modified ratio, (BAP value/d-ROM value)  $\div 7.541$ ) (normal range,  $1.00 \pm 0.13$ ).

**Results:** The ratio in Group (c) seemed high, but there were no significant difference in the modified ratio between 3 groups. Next, we studied the correlation between the follow-up months and modified ratios; no correlation was found in Groups (c) and (b), which comprised patients with CALs, and a weak correlation was found in Group (c). We further studied whether the administration of anti-platelet drugs affected the abovementioned ratios. Then, we found that the modified ratios in patients who were treated with not only anti-platelet drugs but also warfarin and/or statin are significantly higher than those in patients who were treated with only anti-platelet drugs.

**Conclusion:** In KD patients with CALs, the homeostatic mechanisms may persistently function against the increase in oxidative stress. Moreover, this study suggested that warfarin and statin may enhance the relative tolerance to oxidative stress.

### P-34

#### **The long-term outcome of cardiac sequelae with special reference to myocardial lesions and dysrhythmias in Kawasaki patients who reached adult**

Yonesaka S. (1), Takahashi T. (2), Otani K. (2), Jun S. (2), Konno Y. (2), Sato T. (2), Kitagawa Y. (2), Eto S. (2), Ueda T. (2), Sato A. (2), Iehinose K. (2), Kinjo M. (2)

(1) Department of Nursing, Hirosaki University School of Health Sciences, Hirosaki, Japan; (2) Department of Pediatrics, Hirosaki University School of Medicine, Hirosaki, Japan

**Purpose:** Kawasaki disease has been the leading cause of acquired heart disease in childhood. Despite treatment with intravenous gamma globulin, about 5% of patients have coronary abnormalities. Those with large coronary artery aneurysm are at risk for stenosis and myocardial ischemia or infarction. Even those with less severe coronary involvement are needed long-term follow-up because fate of myocardial sequelae remain unclear. Fatal ventricular dysrhythmias requiring ICD were reported in late stage of KD. The future impact of the cardiac sequelae is not certain particularly in the setting of KD patients who reach adult. The study objectives were to assess the long-term outcome of KD patients who reached the age of 20 years with special reference to myocardial lesions and KD-related dysrhythmias.

**Patients and method:** They included 33 KD with coronary artery lesions (CAL). Male to female ratio was 21 to 12. Ages at onset of disease ranged 0.3 to 11 years. Follow-up interval was 13 to 27 years. Follow-up evaluation of coronary arterial lesions was evaluated with coronary arteriography or 64-ray multidetector computed tomography (MDCT), endomyocardial biopsy (EMB), SPECT and biochemical markers for myocardial lesions.

**Results:** Most KD patients who reached adult were free from cardiac symptoms but 3 with ventricular arrhythmias and one angina. Patients with drug free were 11, calcified CAL 6, surgical intervention 3 and 3 with CAL experienced delivery. EMB showed interstitial fibrosis, degeneration, disarray and inflammatory cell infiltration. Accumulation of myelin bodies in the myocytes, disarray of myofibrils, vacuoles and microangiopathy were found in some KD patients. In cases with TL/BMIPP discrepancy, ultrastructural changes revealed massive myelin bodies.

**Conclusion:** The long-term outcome of most KD patients who reached adult was favorable and free of cardiac symptoms even with cardiac sequelae. SPECT image in long-standing KD suggested not only myocardial ischemia but also disordered myocardial fatty acid metabolism. MDCT are feasible and reliable for monitoring young children with KD. Further long-term follow-up studies are required to clarify residual cardiac sequelae. Because the mortality rate among the KD patients with cardiac sequelae is unclear comparing with general population.

### P-35

#### **Tricuspid annular plane systolic excursion (TAPSE) in pediatric and adolescent patients with tetralogy of Fallot (TOF), patients with atrial septal defect (ASD), and age-matched normal subjects**

Koestenberger M. (1), Nagel B. (1), Stueger HP. (2), Ravekes W. (3), Everett AD. (3), Heinzl B. (1), Sorantin E. (4), Cvirn G. (5), Gamillscheg A. (1)

Division of Pediatric Cardiology, Department of Pediatrics, Medical University Graz, Austria (1); Data, Statistics & Risk Assessment; Austrian Agency for Health and Food Safety, Graz, Austria (2); Division of Pediatric Cardiology, Johns Hopkins University School of Medicine, Baltimore, MD, USA (3); Division of Pediatric Radiology, Department of Radiology, Medical University Graz, Austria (4); Institute of Physiological Chemistry, Centre of Physiological Medicine, Medical University Graz, Austria (5)

**Objectives:** The TAPSE is an echocardiographic tool to assess right ventricular (RV) systolic function. RV systolic function in patients with TOF has not been investigated in the pediatric age group to date.

**Methods:** A prospective study was conducted in pediatric and adolescent patients with TOF (n = 105), with small secundum ASD (n = 200), and with normally structured heart (n = 643).

**Results:** The TAPSE values showed a positive correlation with age in both, ASD patients and normal subjects. No significant difference of TAPSE and RVEF values, measured by echocardiography, was seen between control subjects and patients with ASD. In our TOF patients TAPSE values become significantly reduced after a mean of 7 years compared to the lower bound of the -2 SD of age-matched control patients. The decreased TAPSE values showed a positive correlation (r = 0.90) with RVEF determined by MRI and echocardiography.

**Conclusions:** In pediatric patients with a small ASD the systolic RV function is preserved compared to normal subjects. In contrast a significant reduction of TAPSE values with increasing time interval following corrective surgery compared to normal subjects was observed. This was confirmed in MRI and echocardiography measurements of a reduced RVEF. TAPSE is a useful simple complementary tool for assessment of RV systolic function.

### P-36

#### **Alveolar To Arterial Oxygen Tension Difference Increases Due To Mild Impediment Of Pulmonary Circulation In Patients After Bidirectional Glenn Anastomosis**

Hamamichi Y., Ishii R., Terashima Y., Kadota A., Kawazu Y., Inamura N., Kayatani F., Kawata H., Kishimoto H.

Osaka Medical Center And Research Institute For Maternal And Child Health, Osaka, Japan

Increase of alveolar to arterial oxygen tension difference (AaDO<sub>2</sub>) results from venous to arterial shunt, scatter of ventilation/perfusion ratios and impairment of diffusion. In patients after bidirectional Glenn anastomosis increase of AaDO<sub>2</sub> is common because of venous to arterial shunt. However we think disturbance of pulmonary circulation can induce increase of AaDO<sub>2</sub>. This study assessed whether impairment of pulmonary circulation affect increase of AaDO<sub>2</sub> in patients after bidirectional Glenn anastomosis. The medical records of 92 patients after Glenn anastomosis were reviewed. Cardiac catheterization was performed under intravenous sedation between 1993 and 2008. Arterial carbon dioxide pressure (PCO<sub>2</sub>) and arterial oxygen pressure (PO<sub>2</sub>) was assessed by analyzing arterial blood gas during cardiac catheterization. The value of AaDO<sub>2</sub> was calculated with the use of PCO<sub>2</sub> and PO<sub>2</sub> in the following equation: AaDO<sub>2</sub> = 150-PCO<sub>2</sub>/0.8-PO<sub>2</sub> (mmHg). We defined AaDO<sub>2</sub> 50 mmHg or more as increase of AaDO<sub>2</sub> (n = 35). End-diastolic pressure and end-diastolic volume of major ventricle were almost same in both groups. Saturation of oxygen and Ejection fraction of major ventricle were apparently lower in patients with increase of AaDO<sub>2</sub>. Indexes for impediment of pulmonary circulation, consisting of mean pulmonary pressure, diameter of pulmonary artery, and oxygen saturation of pulmonary vein were significantly different in both groups. After the multiple logistic regression analysis increase of AaDO<sub>2</sub> was independently associated with an odds ratio of 59.5 (p = 0.001) for rise of mean pulmonary pressure at least one side (≥15 mmHg), 8.0 (p = 0.006) for narrow diameter of pulmonary artery at least one side (<75% of normal), 4.4 (p = 0.03) for low degree of oxygen saturation of pulmonary vein at least one side (<95%), and 17.0 (p = 0.004) for earlier study age (< 48 months). These four factors explained 55% of reason for increase of AaDO<sub>2</sub> (R-square = 0.55). Our study showed that AaDO<sub>2</sub> increased in response to pulmonary circulation impediments. After bidirectional Glenn anastomosis driving pressure of pulmonary artery is low. Therefore a little obstacle of pulmonary circulation may cause scatter of ventilation/perfusion ratios and impairment of diffusion. AaDO<sub>2</sub> is easily assessed by analyzing arterial blood gas. We suggest that increase of AaDO<sub>2</sub> may serve a useful tool as simple detection of pulmonary circulation disorder in patients after bidirectional Glenn anastomosis.

### P-37

#### **Moderate Hypoventilation Is Risk Factor For Residual Pulmonary Hypertension In Down Syndrome With Left-to-right Shunts Defects**

Hamamichi Y., Ishii R., Terashima Y., Kadota A., Kawazu Y., Inamura N., Kayatani F., Kawata H., Kishimoto H.

Osaka Medical Center and Research Institute for Maternal and Child Health

Hypoventilation due to upper airway obstruction often causes pulmonary hypertension in Down syndrome. Pulmonary hypertension can exacerbate obstructive pulmonary vascular change in Down syndrome patients with left-to-right shunts defects before operation. If hypoventilation persists after definitive surgery, it may intensify residual pulmonary hypertension. However there is no report to describe relation between the degree of hypoventilation and residual pulmonary hypertension after definitive repair in Down syndrome patients. We attempted to determine how much hypoventilation affects residual pulmonary

hypertension. The medical records of 48 Down syndrome patients who had undergone definitive surgery for left-to-right shunt defects were reviewed. Catheterization study was performed under intravenous sedation between 1993 and 2008. Supplemental oxygen was not supplied through pressure measurement. Arterial carbon dioxide pressure (PCO<sub>2</sub>) was assessed by analyzing arterial blood gas during cardiac catheterization. The value of PCO<sub>2</sub> was employed as index of hypoventilation. Residual pulmonary hypertension was defined as mean pulmonary pressure 25 mmHg or over. First operation was defined as primary complete repair or palliative pulmonary artery banding. The number of patients with residual pulmonary hypertension was 16 (ventricular septal defect 8/20, complete atrioventricular septal defect 5/17, patent ductus arteriosus 3/3). In the univariate analysis residual pulmonary hypertension was significantly related to type of cardiac anomalies, timing of first surgical intervention, mean pulmonary artery pressure before first operation, arterial carbon dioxide pressure and pulmonary capillary wedge pressure after definitive repair. After multiple logistic regression model, residual pulmonary hypertension was independently associated with an odds ratio of 10.8 ( $p = 0.015$ ) for moderate hypoventilation (PCO<sub>2</sub>  $\geq 50$  mmHg), 8.0 ( $p = 0.038$ ) for severe pulmonary hypertension before first operation ( $\geq 56$  mmHg). These two factors explained 53% of reason for residual pulmonary hypertension (R-square = 0.53). In this study Hypoventilation after radical surgery intensified residual pulmonary hypertension. Pulmonary hypertension before surgery also engaged with residual pulmonary hypertension. Generally hypoventilation before operation induces pulmonary hypertension in Down syndrome patients. In this view hypoventilation has twofold impact on residual pulmonary hypertension after definitive repair. It may be critical to control hypoventilation from pre-operation to post-operation to prevent residual pulmonary hypertension in Down syndrome patients with left-to-right shunt defects.

#### P-38

##### **Risk factors for postoperative subaortic stenosis: a serious long-term complication to detect early.**

Kalfa D., Ghez O., Metras D., Kreitmann B.

Department of Thoracic and Cardiac Surgery, La Timone Children's Hospital, CHU Marseille

**Background:** Secondary subaortic stenosis (SSS) can occur after surgery for various congenital heart defects with or without initial left ventricular outflow tract obstruction (LVOTO). Objective: to highlight the risk factors for the development of SSS after surgery on defects without initial LVOTO.

**Methods:** A retrospective study of 4710 patients was performed (1984–2005). The criterion for inclusion was a fixed subaortic obstruction requiring surgery, after an open- or closed-heart operation. The criterion for exclusion was an LVOTO at the time of the first operation.

**Results:** Thirty one patients were studied. The mean age at initial surgery was 31 months (4 days – 47 years; median: 2 months). SSS occurred after three main types of surgery: repair of coarctation of the aorta, repair of AVSD and LV – aorta rerouting for double outlet right ventricle or transposition of great arteries. The mean delay of occurrence was 4.3 years (2 months – 19 years). Frequently associated initial anatomical conditions were coarctation of the aorta (44%), lesions of the mitral valve (33%), bicuspid aortic valve (20%) and left superior vena cava (LSVC) (14%). Preoperative anatomical lesions of the LVOT were present in 91% of the cases. SSS was most frequently a subaortic membrane ( $n = 25$ ). The mean pressure gradient across SSS at the time of reoperation was  $47 \pm 29$  mmHg. Five patients

developed a second SSS after 7.4 years (mean). One patient developed a third SSS. No patient died. When compared with patients without SSS, significant risk factors for SSS were low age at surgery (31 vs 74.9 months,  $p < 10_{-4}$ ), pre-existing coarctation of the aorta (44 vs 10%,  $p < 10_{-4}$ ), bicuspid aortic valve (20 vs 6%,  $p = 0.002$ ) and LSVC (14 vs 4%,  $p = 0.02$ ).

**Conclusions:** SSS development is multifactorial, depending on initial anatomical lesions and initial surgery. Low age at initial surgery, coarctation of the aorta, bicuspid aortic valve and LSVC significantly increase the risk of SSS. These elements warrant long-term follow-up for early detection of SSS.

#### P-39

##### **Markers of endothelial dysfunction in youths with essential arterial hypertension**

Plotnikova I.V., Kovalev I.A., Suslova T.E., Churilova V.O.,

Bezlyak V.V.

Institute of Cardiology, Tomsk, Russia

**Objective:** We aimed to assess laboratory findings of endothelial dysfunction (ED) in children with essential arterial hypertension (AH) at different stages of its formation and correlate them with risk factors of cardiovascular diseases, data of daily blood pressure (BP) monitor, structural brain abnormalities.

**Methods:** There were examined 299 youths with essential AH at the age of 12–18 years old. Average age of patients was  $14.9 \pm 2.0$  years old. The control group involved 27 healthy youths comparable with examined group by sex and age. Every patient was made BP daily monitoring (BPDm), according to results of which study groups were formed: 1 group – patients with phenomenon of “white coat hypertension” (WCH) – 98 persons; 2 group – youths with labile atrial hypertension (LAH) – 108 persons; 3 group – stable atrial hypertension (stable AH) – 93 persons. There were assessed: stable metabolites of nitric oxide concentration in blood plasma (NO<sub>2</sub> and NO<sub>3</sub>), level of von Willebrand factor (vWF). Magnetic resonance imaging (MRI) of brain was made by tomography Magnetom – OPEN (Siemens AG, Germany).

**Results:** Significant increase of vWF concentration in relation to control was revealed in patient groups with labile and stable AH. Clinically significant difference of vWF average values in relation to control was 29,33% (95% CI 8,7:49,97,  $p = 0,0056$ ) in group with LAH, and 32,77% in group with stable AH (95% DI 12,31:53,24,  $p = 0,0018$ ). Burdened maternal and paternal inheritance of hypertensive disease and low weight at birth make a significant contribution in increase of vWF concentration in youths with essential AH ( $p = 0,049$ ,  $p = 0,49$  respectively). Logistic regression disclosed that at increase of vWF on 1 unit (%), average figures of pulse arterial pressure level increased at night by 0,36 mm Hg ( $p = 0,048$ ). Besides, made analysis showed that vWF level in group of patients with hypertensive encephalopathy was significantly higher by 18,83% (95% CI 2,39:35,28,  $p = 0,025$ ) then in group of patients without structural brain anomalies.

**Conclusion:** Signs of endothel dysfunction are observed in youths. Willebrand factor can be considered to be verified marker of endothelial dysfunction along with essential AH, even at early stages of AH formation.

#### P-40

##### **Radiofrequency ablation influence on intracardiac hemodynamics in children with idiopathic ventricular premature beats**

Chernushev A.A., Zavadovsky K.V., Kovalev I.A., Popov S.V.,

Lishmanov Yu.B.

Institute of Cardiology, Tomsk, Russia

**Objective:** We aimed to assess state of intracardiac hemodynamics in children with idiopathic ventricular premature beats (IVPB) before and after radiofrequency ablation (RFA).

**Methods:** 12 patients ( $14 \pm 3$  years old) with IVPB were included into study group. They were performed RFA of arrhythmia focus. Control group involved 12 patients comparable with sex and age without structural and functional cardiac changes.

**Results:** End diastolic volume (EDV) and end systolic volume (ESV) of RV were increased by 10–15% on average, stroke volume (SV) of RV was declined by 8%, and ejection fraction (EF) was decreased by 13% according to the quantitative bloodpool SPECT (QBS) data. Rate parameters of LV are increased by 25–29% on average. Time to peak filling (TPPF) of LV is declined by 13%. Rate parameters of RV are increased by 27–38% on average. TTPF of RV is declined by 17.5%. From one to three zones of ventricular premature contraction were also discovered. Their distribution coincides with EPS findings in 86%. Hemodynamics indexes after RFA were assessed in 3–7 days. RV decrease till its size in control group was observed. SV and EF of RV exceeded similar indexes by 15 and 16% respectively in control group. Rate parameters of both R and LVs exceeded in comparison with control group original: peak ejection rate (PER), peak filling rate (PFR) and mean filling rate for 1/3 diastole (MFR/3) of LV by 29–41%; PER, PFR and MFR/3 of RV by 34–50%. TTPF of LV was lesser by 20% in comparison with control group, TTPF of RV – by 27%. Number of zones of ventricular premature contraction decreased to one or disappeared at all after RFA.

**Conclusion:** RV hemodynamics is undergone changes in a greater degree than LV. This fact can be explained by most frequent distribution of arrhythmia focuses in RVOT. Higher rate parameters of LV and RV may have compensatory character under conditions of IVPB. RFA of arrhythmia focus leads to heart size reduction and decrease or disappear of ventricular premature contraction zones. Further increase of rate parameters predominant of RV is observed secondary to regular sinus rhythm. This requires follow-up.

#### P-41

##### **Pharmacological test with sildenafil at pulmonary hypertension assessing in children with congenital heart diseases**

*Yanulevich O.S., Ivanov S.N., Kovalev I.A., Krivoshechekov E.V.  
Institute of Cardiology, Tomsk, Russia*

**Objective:** We aimed to assess acute medical test with sildenafil at pulmonary hypertension (PH) diagnostics in children with congenital heart diseases (CHDs).

**Methods:** Acute medical test with phosphodiesterase inhibitor of V type – sildenafil (Viagra®, Pfizer) was conducted to 52 patients with CHDs and PH. Their systolic pressure (SP) in pulmonary artery (PA) is higher than 70% of systemic arterial pressure. At Echo SP in PA, Qp/Qs and transintra-ventricular pressure gradient with VSD were assessed. Arterial oxygen saturation, arterial pressure and pulse were determined. Patients took per os sildenafil dosed in 1,5 mg/kg. In an hour after drug intake check-up and Echo were made.

**Results:** After sildenafil intake decrease of systolic pressure in PA, increase of Qp/Qs, transintra-ventricular pressure gradient and oxygen saturation were diagnosed in 47 (90,4%) patients. On this occasion test is positive. In 5 (9,6%) patients systolic pressure in PA, Qp/Qs, intra-ventricular gradient and oxygen saturation didn't change after drug intake. Test was negative. Serious side effects of sildenafil were not observed during acute medical test. Three patients had redness of face, one patient had tachycardia.

Positive sildenafil reaction pointed at presence of functional vessels reserves of lesser circuit. These patients also had positive test with oxygen during heart catheterization. They had successful operation. There were not fatal cases. Patients with negative sildenafil test had negative test with oxygen during heart catheterization. This fact speaks for irreversible changes in pulmonary vessels. They were not performed an operation. Conservative therapy was put on.

**Conclusion:** Acute medical test with sildenafil is noninvasive method of examination, which allows to assess PH reversibility. This is important for choice of CHDs treatment.

#### P-42

##### **Immune Globulin Therapy is Associated with Better Outcome of Infants and Children with Cardiomyopathy Induced Heart Failure Results from centers with no pediatric heart transplantations**

*Kampel L. (1), Perles Z. (2), El-Lahham N. (2) Braun J. (2), Rein AJJT (2), Nir A (2)  
The Hebrew University Medical School (1), Jerusalem, Israel, Hadassah University Hospital (2), Jerusalem, Israel*

**Introduction:** Acute heart failure due to heart muscle diseases in infants and children may be mediated by auto-antibodies. The role of immune globulin (IVIG) therapy for this disease is unclear. The prognosis of patients treated medically only, with no heart transplantation (Tx) option is reported.

**Methods:** Retrospective review of infants and children (<18 years) who presented with cardiomyopathy (left ventricular shortening fraction, LVFS  $\leq 25\%$ ), between 1992 and 2007 was performed. Recovery was defined as normalization of left ventricular shortening fraction (LVFS).

**Results:** There were 62 patients, mean age 2.5 years, mean LVFS 16.3% (LVFS < 20% in 47 patients). Overall mortality of was 27%. Of the 38 patients (61%) who were eligible for Tx according to AHA guidelines, 12 (32%) recovered. Nineteen patients (31%) received IVIG. These patients tended to have lower LVFS % (14.7 Vs 17,  $p = 0.071$ ), were more likely to receive inotropic support and steroid treatment ( $p < 0.05$ ) and to be in stage D ( $p < 0.002$ ). Death occurred in 11% of IVIG and 35% of no IVIG group ( $p = 0.006$ ) yielding hazard ratio 0.129 (CI 0.23–0.7). Recovery occurred in 50% of IVIG and 18% of no IVIG ( $p < 0.05$ ). The groups did not differ in age at presentation or gender.

**Conclusions:** Survival and recovery of medically treated pediatric cardiomyopathy patients is a significant possibility and should be kept in mind when planning early heart transplantation. This study, although retrospective, suggests that IVIG therapy should be considered in any acute pediatric cardiomyopathy patient with unknown etiology.

#### P-43

##### **Executive function and social cognition in school-aged children after neonatal corrective cardiac surgery for transposition of the great arteries**

*Calderon J. (1,2), Bonnet D. (2), Courtin C. (3), Concordet S. (2), Plumet M.H. (1), Angeard N. (1)  
Groupe Neuropsychologie du Développement, UMR 663. Université Paris Descartes, Paris, France (1); 1Centre de Référence Malformations Cardiaques Congénitales Complexes – M3C, AP-HP, Université Paris Descartes, Paris, France (2); 3CI-NAPS UMR6232 CNRS, CEA, Université de Caen & Université Paris Descartes. Groupe d'Imagerie Neurofonctionnelle du Développement (GINDEV) Paris, France (3)*

**Aim:** Cyanotic cardiac malformations such as the transposition of the great arteries (TGA) have been associated with neurodevelopmental dysfunctions. Neuropsychological assessment has become an important focus of research in this population. The purpose of this study was to specifically evaluate, for the first time, executive function (EF) and social cognition development (SC) which are key components of cognitive development in TGA children.

**Methods:** Twenty-one patients who underwent neonatal open heart surgery for TGA ranging in age from 7 to 7, 11 years using full-flow cardiopulmonary bypass were compared to 21 normally developing age-matched children using different standardized neuropsychological measures specifically designed to assess EF (cognitive and response inhibition, verbal and spatial working memory and planning). They also were presented two SC tasks (first and second order false belief understanding).

**Results:** General intellectual capacity (IQ) was within the normal range for TGA children. Performances on all executive functions and social cognition were significantly lower for TGA group. A discriminant multivariate analysis provided evidence for cognitive and behavioral inhibition as well as false belief tasks as being the most important contributors to the differentiation between the groups. No significant differences were found between TGA children who had a prenatal or a postnatal diagnosis for all the cognitive measures.

**Conclusions:** TGA children demonstrate great difficulties in exerting cognitive and response inhibition. They also present specific deficits in false belief understanding which were inter-related with immature executive abilities suggesting a possible comorbidity of neurocognitive impairments. Further research is needed to determine how these deficits can be related to the higher frequency of academic and behavioral difficulties reported for these children. There is increasing need for longitudinal neuropsychological follow-up in this population.

#### P-44

##### **Quality of Life in Patients with Congenital Heart Disease**

Silva A. M. (1), Vaz C. (1), Areias M. E. G. (1), Vieira D. (1), Proença C. (1), Viana V. (1, 2), Moura C. (2), Areias J. C. (2) Instituto Superior de Ciências da Saúde – Norte, Gandra – Paredes, Portugal (1); Hospital S. João, Faculdade de Medicina da Universidade do Porto, Portugal (2)

**Objectives:** To assess the perception on quality of life (PQL) of Adolescents and Young Adults with Congenital Heart Disease (CHD) and to examine which variables play a negative impact on it and which add a resilience effect, improving the acceptance of and adjustment to illness.

**Methods:** Unbiased patients, 22 male and 18 female, aged 12 to 26 years old, 27 submitted to surgical procedure and 13 without, participated in this study. All subjects had complete medical records, and were interviewed once; demographic and clinical data were collected, and patients filled a self-report questionnaire on Quality of Life, the WHOQOL-BREF, and completed a semi-structured interview on several topics, such as, social support, family educational style, self-image, functional limitations and emotional adjustment.

**Results:** Our patients showed a better PQL than the general population, especially on the Psychological, Social Relationships and Environment ( $t = 2,502$ ,  $p = 0,017$ ;  $t = 3,544$ ,  $p = 0,001$ ;  $t = 2,925$ ,  $p = 0,006$ ) Scales, showing statistically meaningful differences. Older subjects hold better PQL than the younger ones on the Psychological Scale ( $t = 2,502$ ,  $p = 0,017$ ). Cyanosis didn't show any significant impact over PQL decay but,

consistently with other investigations, we found that the number of surgical procedures ( $t = 2,292$ ,  $p = 0,028$ ), and the persistence of moderate-to-severe residual injuries ( $z = 2,368$ ,  $p = 0,018$ ) had a great detrimental effect on PQL. Social support had an important impact on incrementing resilience, promoting adjustment and acceptance of illness ( $z = -3,152$ ,  $p = 0,002$ ).

**Conclusions:** Several factors may play a role on adjustment to CHD, either improving the adaptation to different life cycle's challenges, or worsening it. PQL is an important parameter that gives impressions of subjects on their quality of life, on several different matters. In this study, we may conclude, consistently, that some buffer variables on CHD may play very crucial roles on incrementing the PQL of patients along life. That is the case of social support that can probably explain why in our population, as in others referred by different authors, the PQL is better than in the normal population. However, in our patients, the number of surgeries and the moderate-to-severe residual injuries reverted that effect.

#### P-45

##### **Six-minute walk distance in obese children and adolescents – effect of a weight reducing program**

Geiger R. (1), Rauchenzauner M. (2), Willeit J. (1), Rummel M. (3), Högl W. (4), Stübing K. (3), Stein J. (1) Department of Pediatrics, Clinical Divisions of Cardiology (1); Neurology, Innsbruck Medical University, Innsbruck, Austria (2); Fachklinik Prinzregent Luitpold, Scheidegg, Allgäu, Germany (3); Department of Endocrinology and Diabetes, Birmingham Children's Hospital, Birmingham, United Kingdom (4)

**Background:** Overweight children tend to perform worse in the six-minute walk test (6MWT) than non-overweight children but prospective studies on 6MWTs in overweight children are sparse. This study aimed to assess the validity of the 6MWT in a weight reduction program (BE programme).

**Methods:** One hundred thirteen obese children and adolescents (BMI > 95th percentile; mean  $\pm$  SD age  $12.9 \pm 2.0$  years) underwent a standardized 6MWT at the beginning and end of an in-patient weight reduction program. Results of the 6-minute walk distance (6MWD) were compared to those of age and sex matched non-overweight children ( $n = 353$ ).

**Results:** In obese children, pre-intervention 6MWD averaged 93% of controls ( $631 \pm 88.0$  m vs.  $675 \pm 70.0$  m,  $p \leq 0.001$ ) with similar values post-intervention ( $667 \pm 90.1$  m vs.  $675 \pm 70.0$  m,  $p = 0.260$ ). After  $27 \pm 6.8$  days of intervention the 6MWD increased from  $631 \pm 88.0$  m to  $667 \pm 90.1$  m ( $p \leq 0.001$ ). Participants reduced their BMI percentile from  $98.2 \pm 2.1\%$  to  $96.8 \pm 3.8\%$  ( $p \leq 0.001$ ), and BMI-SDS from  $2.37 \pm 0.6$  to  $2.13 \pm 0.6$  ( $p \leq 0.001$ ). BMI-SDS ( $\beta = -0.391/-0.366$ ), height ( $\beta = 0.349/0.404$ ), and  $\Delta$ HRR ( $\beta = 0.311/0.272$ ;  $p \leq 0.001$  for each) were independent predictors of the 6MWD at pre- and post-intervention time-points ( $r^2 = 0.338$  and  $r^2 = 0.349$ , respectively).

**Conclusion:** The 6MWD improves on a weight reduction program and seems to indicate improvement of physical fitness and decreased metabolic demand during daily activities. The 6MWT represents an easy to administer and reliable assessment tool of exercise performance in obese children and adolescents.

#### P-46

##### **Anomalous Origin of the Left or Right Coronary Artery from the Pulmonary Artery (ALCAPA/ARCAPA): A 10-year single center experience**

Flosdorff P. (1), Riede F.T. (1), Suchowerskyj P. (1), Cho M. (2), Kostelka M. (2), Dähnert I. (1)

Heart Center, University of Leipzig, Department of Pediatric Cardiology, Leipzig, Germany (1); Heart Center, University of Leipzig, Department of Pediatric Cardiac Surgery, Leipzig, Germany (2)

**Introduction:** Patients with ALCAPA/ARCAPA often present with myocardial ischemia, severely impaired ventricular function and a-v valve regurgitation. Currently coronary artery transfer is regarded as first choice surgical treatment. Aim of the study was to evaluate the ventricular function as well as a-v valve competence during mid-term follow-up.

**Methods:** All patients with a diagnosis of ALCAPA (n = 18)/ARCAPA (n = 3) admitted to our hospital from 1999–2009 were included and retrospectively analyzed.

**Results:** Median patients age was 6 months (range 13 days to 17, 5 years). Left ventricular function (LVF<sub>x</sub>) at time of admission was severely impaired in 9 (43%), moderately impaired in 4 (19%), and normal or mildly reduced in 8 patients (38%). In 10 patients (48%) mitral regurgitation was grade 2 or greater. All patients underwent surgical treatment (direct reimplantation, n = 20, 95%, bypass procedure n = 1, 5% due to impossibility to use the CPB). One patient had simultaneous mitral annuloplasty.

Mean follow up was 3,5 years (range 4 month to 9,7 years) No early or late mortality was seen. Left ventricular assist devices (LVAD) were used in five patients (24%). At last follow up all patients were in NYHA class I. 20 patients (95%) had a normalized LVF<sub>x</sub>. In 4 patients (19%) a mitral regurgitation grade 2 or 3 was diagnosed. Mild coronary stenosis occurred in two patients, one with severe left coronary artery stenosis was treated with coronary stent implantation.

**Conclusions:** Coronary artery reimplantation in patients with ALCAPA or ARCAPA is a surgical procedure with low mortality and very good mid-term results. In patients with severe impaired ventricular function implantation of LVAD is a useful post-operative cardiac support technique. Left ventricular function normalizes over time. Mitral valve regurgitation often improves along with ventricular function. Coronary artery stenosis may occur as early or late postoperative complication.

#### P-47

##### **Volatile substance abuse: Evaluation of QT Dispersion and sudden death risk in children with volatile substance addiction**

Levent E. (1), Can S. (2), Demir E (2), Yüncü Z (3), Özyürek A.R. (1) Ege University, Faculty of Medicine, Pediatric Cardiology Department (1), Pediatric Pulmonology Department (2), izmir, TURKEY; Child and Adult Alcohol and Substances Addiction Research and Treatment Center of Ege University, izmir, TURKEY (3)

Abuse of organic volatile substances in children has become a social health problem that is increasing in the recent years. Volatile substances sensitize the heart to catecholamines, such that sudden alarm or exercise may precipitate sudden death. The aim of this study was to evaluate the electrocardiographic (ECG) and echocardiographic findings and sudden death risk in substance abuser children.

We examined 31 substance abuser children who had admitted to Child and Adult Alcohol and Substances Addiction Research and Treatment Center of Ege University (EGBAM). The study encompasses 31 male chronic volatile substance abusers with a 1 year or longer duration abuse aged between 15–19 years.

ECG and echocardiography was performed on these children and control group. QT, QTc and QTc dispersion were measured in all groups. The study group consisted of 31 chronic volatile

substance abusers aged between 15 and 19 ( $17.94 \pm 1.52$ ) years. Nineteen healthy subjects aged between 15 and 20 ( $18.37 \pm 1.16$ ) years were considered as control group. There was a statistically difference in the measurements of LVESD, LVEF, LVFS, mitral E, tricuspid E and A velocities and LVMPI between two groups. In study group, the mean values of QT ( $362.00 \pm 34.78$ ,  $329.00 \pm 21.3$  ms) and corrected QT interval ( $428.56 \pm 31.2$ ,  $356.5 \pm 28.71$  ms), measuring the duration of ventricular depolarization and repolarization, were higher compared to the values in the control group. As the markers of variability of ventricular repolarization, QTd and QTcd were measured and volatile substance abusers showed significantly higher mean QTd ( $70.19 \pm 22.24$ ,  $36.68 \pm 14.31$  ms) and QTcd values ( $89.02 \pm 19.3$ ,  $42.31 \pm 19.21$  ms) compared to the control group. Volatile substances sensitize the heart to circulating catecholamines, such that sudden alarm or exercise may precipitate sudden death. In our study, we found that volatile substance abuser children have electrocardiographic and echocardiographic pathological changes. In addition, the important effects of these agents have been associated with QT, QTc interval prolongation and increasing QTc dispersion. These findings may be associated with the sudden cardiac death in these children.

#### P-48

##### **Croatian Study (2002–2007): Distribution of congenital heart defects and outcome analysis**

Malčić I., Dilber D

University Hospital Zagreb, Departement of pediatrics, Division of pediatric cardiology, Zagreb, Croatia

**Objectives:** The aim of our study was to investigate the prevalence of congenital defects in children born in Croatia during a period of 5 years, extending from 2002 through 2007, association with extracardiac malformations, treatment and outcome analysis.

**Methods:** The study was set up according to the EUROCAT registries principles. Children with diagnoses of congenital heart defects from 01.01.01 through 09.29.31 according to the European Paediatric Cardiac Code, obtained from medical records from 14 paediatric cardiology centres that cover the whole country were included. Outcome analysis was performed using Risk Adjustment in Congenital Heart Surgery-1 method and Aristotle Basic Complexity Score.

**Results:** Between October 1st, 2002. and October 1st 2007., there were 205 051 live births in Croatia, of which 1480 patients were diagnosed with congenital heart disease, accounting for 0.72% of the live-born children. The distribution was made up of 34.6% children with ventricular septal defect, 15.9% with atrial septal defect, 9.8% with patency of arterial duct, 4.9% pulmonary valvar stenosis, 3.3% tetralogy of Fallot, 3.3% transposed great arteries, 3.3% aortic stenosis, 3.2% aortic coarctation, 4.3% atrioventricular septal defect and common atrioventricular orifice, 2.3% hypoplastic left heart syndrome and 8.3% other severe defects. The average age in the time of diagnoses is 70.41 days (SD 188.13), with statistically low average time of diagnoses of severe heart defects, 9.6 days, SD 32.52. Among patients, 14.5% had chromosomal defects, syndromes and/or other congenital major anomalies. Of the 556 operations performed, ABC score could be assigned to 553 and RACHS-1 score to 536 operations. Procedures were performed in 2 institutions in Croatia and 7 abroad. The average complexity for cardiac procedures done in Croatia was significantly lower ( $p < 0.001$ ). Only after adjustment for complexity are there marked differences in mortality and occurrence of postoperative complications.

**Conclusion:** The rates of specific cardiac defects and association with extracardiac malformations are generally comparable with those reported in similar studies. Both ABC and RACHS-1 were predictive of in-hospital mortality as well as prolonged post-operative length to stay, and can be used as a tool in our country to evaluate a cardiosurgical model and recognise potential problems.

#### P-49

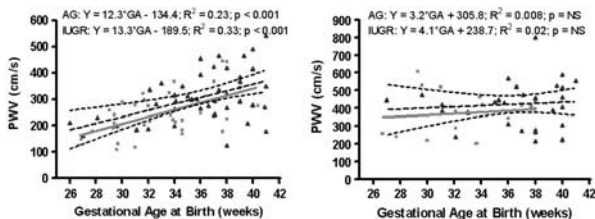
##### **Intrauterine Growth Restriction does not impair Cardiac Function and Vascular Compliance in Infants**

Jaeggi E. (1), Shah J. (2), Keating S. (2), Kingdom J. (2), Bradley T. (1), Keeley F. (1), Redington A. (1)  
The Hospital for Sick Children, Toronto, Canada (1); Mount Sinai Hospital, Toronto, Canada (2)

**Background:** The association between low birth weight and premature cardiovascular events has been demonstrated and led to “fetal programming” hypothesis. We postulated that chronic placental insufficiency leads to the antenatal onset of persistent changes in vascular morphology and function.

**Methods:** Since 2006, we prospectively enrolled 55 appropriately grown (AG) and 38 fetuses with intrauterine growth restriction (IUGR; defined as birth weight <10th percentile for age and gender). We studied the impact of gestational age/growth on myocardial mass and function (echocardiography); umbilical arteries (stress-strain; elastin/collagen content); and, at 1 week and 6 months of life, on vascular stiffness (echocardiography; pulse-wave velocity (PWV); blood pressure).

**Results:** Compared to AG, IUGR fetuses were more likely (<0.001) to have hypertensive mothers (32% vs 4%); higher placental (76% vs 8%) but lower cerebral arterial resistances (mean z-score: -1.5 vs. 0.3); earlier deliveries (32.4 (26.7–40.4) vs. 37.3 (26–41) weeks); placental weights <10% (78% vs. 9%); thinner umbilical artery walls ( $0.58 \pm 0.17$  vs.  $0.7 \pm 0.19$  mm) with lower elastin content ( $0.9 \pm 0.4$  vs.  $1.2 \pm 0.5\%$ ;  $p < 0.05$ ); and lower biometric z-scores at birth (weight: -2 vs. -0.2; length: -2.2 vs. 0.4; head circumference: -1.7 vs. -0.1). However, postnatally, indices of cardiovascular function (adjusted for gestational age) and dimensions (adjusted for body surface area) were comparable during early infancy with physiological, age-dependent changes between birth and 6 months. The figure shows brachioradial PWV at 1 weeks (left) and 6 months (right panel); blue  $\Delta$  = AG; red  $\square$  = IUGR).



**Conclusion:** Despite marked abnormalities in utero, we found no evidence of an early impact of IUGR on cardiac or vascular function. The manifestations of “fetal programming” appear to result from maturational changes later in life.

#### P-50

##### **Physical Examination: The Oldest but Still an Up-to-Date Diagnostic Method**

Kula S., Pektaş A., M; Cevik A., Taskin B. D., Oğuz A. D., Timaoglu F. S., Olgunturk R.  
Gazi University Medical Faculty Hospital, Ankara, Turkey

**Introduction:** The present study aims to emphasize the significance of cardiovascular system examination by investigating when the related cardiac murmur was auscultated and the congenital cardiac malformation was diagnosed.

**Methods:** The present study retrospectively surveys 284 children who were referred between January 2008 and December 2009 and in whom the diagnosis of cardiac anomaly was delayed.

**Results:** The study group consisted of 134 boys (47.2%) and 150 girls (52.8%). The mean height of the reviewed children was  $120.3 \pm 30.9$  cm while their average body weight was  $26.4 \pm 16.5$  kg. The most frequent cardiac anomaly was atrial septal defect (ASD, 33.1%) which was followed by patent ductus arteriosus (PDA, 19.0%), coarctation of aorta (10.6%) and ventricular septal defect (VSD, 9.2%) respectively. The majority of the reviewed patients were from the Internal Anatolia (72.5%). Cardiac murmurs were detected at an average age of  $8.0 \pm 5.2$  years and the congenital cardiac anomaly was specified after  $6.6 \pm 4.9$  years passed in average. Subjects with coarctation of aorta, ASD, PDA and VSD were diagnosed at an average age of  $9.2 \pm 4.7$  years,  $8.0 \pm 5.0$  years,  $7.7 \pm 5.2$  years and  $8.1 \pm 6.2$  years respectively. The diagnostic delay span was significantly longer in the reviewed children who had male gender and coarctation of aorta and who came from the Mediterranean region ( $p = 0.003$ ,  $p = 0.019$  and  $p = 0.040$  respectively). Patients with different social insurances were statistically similar in aspect of diagnostic delay span. As expected, non-cyanotic anomalies were detected after a significantly longer time interval when compared with cyanotic malformations ( $6.9 \pm 4.8$  vs  $1.2 \pm 1.0$  years,  $p = 0.001$ ). Thus the present study aims to emphasize on the negligence of physicians during the determination of non-cyanotic anomalies which are less symptomatic.

**Conclusions:** As the technological advances have been largely adopted by the health sector during the last decade, traditional methods of clinical evaluation are much ignored. However physical examination still preserves its importance as the simplest and most efficient diagnostic procedure. Therefore pediatricians always should remember that they can help to avoid considerable health and finance burdens just by auscultating the cardiac sounds, palpating the arterial pulses and measuring the blood pressure and thus specifying the cardiac anomalies as soon as possible.

#### P-51

##### **Catheter Haemodynamic Assessment with Pharmacological Stress to unmask Obstruction of the Fontan Circulation**

Manne P., Bhole V., Stumper O.

Birmingham Children's Hospital, Birmingham, United Kingdom

**Introduction:** The functional outcome of the Fontan palliation for hypoplastic left heart syndrome (HLHS) is dependent upon unobstructed pulmonary arterial and pulmonary venous pathways. In some symptomatic patients with Fontan circulation any obstruction may not be evident at rest or during cardiac catheterization which is performed under non physiologic conditions. We reassessed haemodynamics using pharmacological stress to mimic exercise/physiologic conditions.

**Materials and methods:** Three patients (mean age - 10 years) with HLHS with Fontan palliation underwent cardiac catheterization under general anaesthetic. Indication for catheterization was protein-losing enteropathy (PLE), bronchial casts and decreased exercise tolerance in 1 patient each. Haemodynamics were assessed at rest and after Dobutamine (n - 2) or Isoprenaline infusion (n-1).

**Results:** In the patient with PLE at baseline PA mean pressure was 15 mmHg, LA pressure was 11 mmHg, transpulmonary gradient

(TPG) was 4 mmHg with 1 mm Hg gradient across the interatrial septum (IAS). During pharmacological stress there was an increase in the gradient from 1 to 4 mmHg across the IAS and PA pressure was 20 mmHg. TPG increased to 10 mmHg unmasking a significant gradient across the IAS. Atrial septectomy with conversion to extracardiac fenestrated Fontan led to complete resolution of PLE.

The patient with bronchial casts at rest revealed LA pressure of 12 mmHg and RA of 15 mmHg with no gradient between pulmonary artery and Fontan tunnel. With pharmacological stress there was no change in LA and RA pressure unmasking a gradient of 1 mm Hg between the LPA and lateral tunnel. Symptoms improved with LPA stenting.

The patient with decreased exercise tolerance at rest (nodal rhythm) showed LA and RA pressure of 13 and 16 mmHg respectively. During pharmacological stress TPG increased to 7 mm Hg suggesting increased pulmonary vascular reactivity. The patient was treated with Sildenafil with symptomatic improvement.

**Conclusion:** Catheter haemodynamic assessment with pharmacological stress testing can unmask Fontan pathway obstructions which may not be evident during resting conditions. Thus, it should be considered in the evaluation of patients with a failing Fontan circulation where resting haemodynamics are inconclusive.

#### P-52

Withdrawn

#### P-53

##### **Interest of beta-blockers in patients with right ventricular systemic dysfunction**

Bouallal R. (1), Godart F (1), Richard A. (1), Francart C. (1)  
(1) Hôpital Cardiologique. Centre Hospitalier Universitaire. Lille.  
France

**Introduction:** beta-blockers improve the prognosis of patients with heart failure due to left ventricular systolic dysfunction. The aim of this study is to assess the efficacy of beta-blockers in patients with dysfunctional systemic right ventricle.

**Methods:** fourteen patients with systemic right ventricle following Mustard or Senning operation for transposition of the great arteries, or congenitally corrected transposition of the great arteries were included in the study. All patients had a decreased systemic right ventricular ejection fraction despite standard heart failure therapy. Quality of life, NYHA-class, aerobic capacity and systemic right ventricular function were assessed before treatment with beta-blockers and at the end of the follow-up period (mean  $12.8 \pm 10.1$  months, range 3–36 months).

**Results:** change in NYHA-class was significant from a median NYHA – class II (range I-IV) to a median NYHA functional class II (range I-II) ( $p = 0.016$ ). Quality of life improved significantly throughout the study from a median grade 2 (range 1–3) to a median grade 1 (range 1–2) ( $p = 0.008$ ). Systemic right ventricular ejection fraction assessed by radionuclide ventriculography improved significantly from a median of 41% (range 29–53%) to 49% (range 29–62%) ( $p = 0.031$ ). However, change in ejection fraction assessed by magnetic resonance imaging was not significant from a median of 29% (range 12–47%) to 32% (range 22–63%) ( $p = 0.063$ ).

**Conclusions:** In patients with heart failure due to systemic right ventricular dysfunction, beta-blockers improve NYHA-class, quality of life and systemic right ventricular ejection fraction assessed by radionuclide ventriculography.

#### P-54

##### **Beta-blocker therapy improves diastolic function and reduces hypertrophy in asymptomatic childhood familial hypertrophic cardiomyopathy.**

Östman-Smith I., de-Wahl Granelli A, Allahyari P

Institute of Clinical Sciences, Division of Paediatric Cardiology at Queen Silvia Children's Hospital, The Sahlgrenska Academy, Gothenburg University, Gothenburg, Sweden

**Introduction:** Currently asymptomatic children with hypertrophic cardiomyopathy (HCM) are not recommended active treatment unless they have risk factors for sudden death present.

**Methods:** Asymptomatic children with familial HCM were identified by family-screening. After a risk-stratification low-risk individuals were randomized between life-style modification only (controls), and metoprolol plus life-style modifications; high-risk children were started on propranolol; in both instances a target dose of 6 mg/kg/day was aimed for. Echocardiographic measures of cardiac hypertrophy and diastolic function were compared initially and after one year.

**Results:** The group treated with beta-blocker therapy (average dose  $6.1 \pm 0.9$  mg/kg/day,  $n = 12$ ) had the same age,  $14.5 \pm 5.7$  yrs (mean  $\pm$  SD), as controls ( $n = 6$ ;  $14.9 \pm 7.1$ ) but, because the beta-blocker group contained the high-risk patients, this group had more severe cardiac hypertrophy at the beginning (septum-to-cavity ratio  $0.36 \pm 0.10$  versus  $0.26 \pm 0.02$ ,  $p = 0.005$ , signed rank, and left ventricular wall-to-cavity ratio  $0.24 \pm 0.04$  versus  $0.19 \pm 0.02$ ,  $p = 0.017$ ). There was also more severe initial diastolic dysfunction in the beta-blocker group affecting E:A ratio, E:e ratio and e:a ratio. After one year there were no significant changes in the control group, however in the beta-blocker group there had been a significant reduction in degree of hypertrophy, in septum-to-cavity ratio to  $0.31 \pm 0.10$ ,  $p = 0.006$ , and in left-ventricular wall-to-cavity ratio to  $0.19 \pm 0.03$ ,  $p = 0.008$ . Furthermore there were significant improvements in several measures of diastolic function, in pulmonary vein S:D ratio from  $1.06 \pm 0.22$  to  $0.84 \pm 0.15$ ,  $p = 0.013$ ; E:e ratio from  $10.2 \pm 1.8$  to  $9.2 \pm 2.1$ ,  $p = 0.006$ ; and E:A ratio from  $1.50 \pm 0.42$  to  $1.88 \pm 0.39$ ,  $p = 0.045$ . Relaxation was faster as evidenced by a reduction in isovolumic relaxation time from  $74 \pm 21$  ms to  $66 \pm 21$  ms ( $p = 0.04$ ) and a reduction in mitral valve deceleration time from  $185 \pm 13$  ms to  $174 \pm 50$  ms ( $p = 0.013$ ).

**Conclusions:** Beta-blocker therapy improves diastolic function and reduces cardiac hypertrophy in asymptomatic familial childhood HCM which explains why they maintain an unchanged exercise capacity in spite of a significant reduction in maximal heart rate on exercise.

#### P-55

##### **Quality of life in children, adolescents, and young adults with congenital heart disease**

Henzinger H., Stein J.-I.

Medical University Innsbruck: Department of Paediatrics III, Cardiology, Pulmology, Allergology, Cystic Fibrosis; Austria

**Objectives:** The aim of this study was to explore the association between severity of congenital heart disease (CHD) and both self- and parent-reported health related quality of life (HRQOL).

**Methods:** A parent (370 parent-reports for patients from 2 to 18 years) and/or the patient (259 self-reports for patients 8 years of age and older) completed HRQOL questionnaires during a visit to the outpatient clinic of our paediatric cardiology department. HRQOL was assessed using the German version of the



PedsQLTM 4.0, a questionnaire with self- and parent-report modules for different age groups. Disease severity was categorized by a cardiologist into three classes: mild, moderate, and severe heart defects, and a control group of healthy participants who presented at the clinic for suspicion of CHD.

**Results:** Patients with severe CHD reported significantly lower physical HRQOL than patients with mild CHD and the reference group; however, they only reported significantly lower psychosocial HRQOL than patients with mild CHD but not the reference group. These differences were also observed in the parent-reports. Patients between 8 and 12 years of age reported lower psychosocial HRQOL than patients aged 13 and older. Parents of patients between 8 and 12 years reported lower levels of psychosocial HRQOL than parents of 2 to 4 year olds did. Overall, parents reported lower psychosocial HRQOL than their children did. Compared to the healthy control sample of similar published studies the participants of this study reported higher psychosocial HRQOL, however their parents reported lower psychosocial HRQOL than healthy controls.

**Conclusion:** Overall, CHD patients reported high levels of HRQOL, comparable to healthy controls. However, HRQOL decreases with increasing severity of CHD. Although the reference group and their parents reported higher physical HRQOL than patients with complex CHD, their psychosocial HRQOL was not higher. This highlights the subjective component of HRQOL and the importance of the context in which data are collected. The results further suggest that the psychosocial effects of CHD are not constant across the ages, with 8 to 12 year olds experiencing the most difficulties. Parents judge the psychosocial impairments of their children to be more severe than they themselves.

#### P-56

##### **Analytical comparison for the case definition of elevated type-b amino-terminal pro-natriuretic peptide in acute Kawasaki disease**

Dahdah N. (1), Fournier A. (1), Cousineau J. (1), Delvin E. (1), Mir T. (2), Nir A. (3)

Centre Hospitalier Universitaire Sainte-Justine, Montréal, Canada (1); University Heart Center, Hamburg, Germany (2); Shaare Zedek Medical Center, Jerusalem, Israel (3)

**Introduction:** The clinical criteria and the usual biological markers used for the diagnosis of Kawasaki disease (KD) lack the necessary specificity for an accurate diagnosis. Furthermore, with the increased awareness of cases presenting with incomplete diagnostic criteria the need for a biochemical diagnostic marker became even a greater necessity. We have recently identified that the N-terminal moiety of the B-type natriuretic peptide (NT-proBNP) is elevated in acute KD with a cut-off value of 170pg/ml based on ROC analysis.

**Objectives:** to study the diagnostic value of NT-proBNP.

**Methods:** Serum levels of NT-proBNP obtained from KD patients upon onset were compared to those obtained from febrile controls, to the cut-off value, as well as to the published upper limits for age and to a Z-score derived from a large series of healthy children.

**Results:** There were 58 KD patients and 37 controls aged  $3.74 \pm 2.69$  [median 2.9; range 1.9–5.1] v.s.  $5.75 \pm 3.70$  [6.0; 2.8–7.9] years, respectively;  $p = 0.014$ . NT-proBNP was significantly increased in KD patients compared to controls ( $956.5 \pm 1243.9$  [568.0; 251.0–1258.1] v.s.  $186.3 \pm 193.9$  [134.0; 73.7–235.0] pg/ml, respectively;  $p < 0.001$ ). The sensitivity, specificity, positive and negative predictive values

varied between 70–86%, 73–92%, 83–96%, and 67–77%, respectively. The highest positive predictive value and specificity were obtained with the Z value  $> 2.5$ . The upper limit for age definition were also highly reliable, adjusting for the age difference between cases and controls. Finally, the odds ratio for the recognition of KD versus no KD subjects varies between 12.1 (95%CI 4.4–33.4), 14.4 (95%CI 5.2–39.9) and 23.8 (95%CI 6.4–88.0), for the upper limit for age definition, the 170 pg/ml cut-off value, and the Z value  $> 2.5$ , respectively ( $p < 0.0001$ ). **Conclusion:** Our data indicate that NT-proBNP is a good and reliable biochemical marker with an excellent positive predictive value for the diagnosis of KD regardless which case definition is used. Future studies are needed to specifically determine its utility in cases with incomplete clinical criteria.

#### P-57

##### **Dysphagia lusoria caused by Aberrant right subclavian artery in severity is as same as complete vascular ring in schoolchildren: validated by Esophageal Manometry**

Chen F.L. (1, 2), Kuo S.M. (3), Li Y.C. (4)

Division of Pediatric Cardiology, Department of Pediatrics, Chung Shan Medical University Hospital, Taichung, Taiwan (1); Institute of Medicine, Chung Shan Medical University, Taichung, Taiwan (2); Division of Cardiovascular Surgery, Chung Shan Medical University Hospital, Taichung, Taiwan (3); Institute of Medicine, Department of Pharmacology, Chung Shan Medical University, Taichung, Taiwan (4)

**Background:** Dysphagia lusoria (DL) was a term to describe dysphagia caused by external vascular compressions. Among them, congenital vascular ring was the most common cause. According to the classifications of CVR, aberrant right subclavian artery (ARSCA), a form of partial CVR, is believed to have a benign natural history with no significant compression of the trachea and esophagus, in contrast to complete vascular ring (CVR). However, our clinical observation is that ARSCA can cause significant swallowing difficulties. In this study, we prospectively performed esophageal manometry to evaluate the severity of esophageal function in subjects with ARSCA and CVR to determine the difference in the severity of DL and efficacy of surgical intervention.

**Methods:** From January, 2002 to December, 2007, 46 children of ARSCA ( $8.5 \pm 2.9$  years) and 40 with CRV ( $6.9 \pm 4.1$  years,  $p > 0.05$ ) were enrolled after confirmation of diagnosis by a history of dysphagia, barium swallow, echocardiography, and computed tomography. Patients with developmental delay or mental retardation were excluded. Esophageal manometry was performed preoperatively and postoperatively to measure the severity of esophageal compression.

**Results:** Esophageal manometric measurement was successfully obtained for all subjects. All had symptomatic relief of dysphagia. All subjects had a high-pressure zone at the site of vascular compression. The maximal and mean preoperative esophageal pressures were not statistically different between ARSCA ( $77.4 \pm 36.9$  mmHg and  $45.8 \pm 25.8$  mmHg) and CVR ( $98.6 \pm 37.1$  mmHg,  $p > 0.05$  and  $53.1 \pm 28.9$  mmHg,  $p > 0.05$ ), respectively. Postoperatively, both maximal and mean pressures were significantly reduced for ARSCA ( $41.8 \pm 25.8$  mmHg and  $23.8 \pm 15.3$  mmHg) and for CVR ( $53.1 \pm 37.1$  mmHg and  $28.9 \pm 15.1$  mmHg), respectively.

**Conclusion:** This is the first study to demonstrate that CVR is a major cause of DL in schoolchildren. Characteristics of dysphagia in patients with ARSCA were similar to those observed in complete VR. They can have a significant severity of dysphagia in schoolchildren, and should be pay attention to their impact on schoolchildren.

## P-58

**Effects of Endothelin-Receptor-Antagonism on Left Ventricle and Arterial Blood Pressure in Children with Pulmonary Arterial Hypertension**

Baumgartner S., Salzer-Muhar U.

Division of Paediatric Cardiology, Department of Paediatric and Adolescent Medicine, Medical University of Vienna, Austria

**Objectives:** The aims of this retrospective study were to analyze and to describe time courses of echocardiographic left ventricular diameters and systemic arterial blood pressure in children with pulmonary arterial hypertension who were treated with the endothelin-receptor-antagonist Bosentan.

**Methods:** The following data from 13 children aged from 3 to 18 years suffering either from idiopathic pulmonary arterial hypertension (n = 2), or from pulmonary arterial hypertension associated with congenital heart disease (Eisenmenger physiology with post-tricuspid shunt n = 6, pulmonary atresia with ventricular septal defect n = 5) were analyzed: enddiastolic and endsystolic diameter of the left ventricle (M-mode), stroke volume, blood pressure (systolic, diastolic, mean arterial pressure, pulse pressure) and transcutaneous oxygen-saturation. Time points were prior to initiation of therapy with Bosentan and at 3- and 6- month follow-up. Left ventricular enddiastolic and endsystolic diameters were expressed in z-scores, stroke volume in ml/kg body weight and blood pressure in mmHg.

**Results:** Left ventricular enddiastolic and endsystolic diameter increased in children with idiopathic pulmonary hypertension, only left ventricular endsystolic diameter increased in patients with Eisenmenger physiology. Systemic arterial blood pressure: Pulse pressure increased in children with idiopathic pulmonary hypertension and Eisenmenger-physiology but not in children with pulmonary atresia and ventricular septal defect. In the latter group of patients both systolic blood pressure and pulse pressure decreased.

**Conclusions:** The effect of Bosentan on left ventricular systolic and diastolic diameters in children with idiopathic pulmonary hypertension can be explained by both the direct and serial right-left ventricular interaction. Whether the decrease of systolic blood pressure and pulse pressure in children with pulmonary atresia and ventricular septal defect may be explained by a specific effect of Bosentan on the media of the dilated aorta needs further research.

## P-59

**Coronary artery ostial anomalies in common arterial trunk**

Houyel L. (1), Bajolle F. (2), Bonnet D. (2)

Centre Chirurgicale Marie Lannelongue M3C, Université Paris Sud, Paris, France (1); Necker-M3C, Université Paris Descartes, Paris, France (2).

Coronary artery ostial anomalies in common arterial trunk.

**Background:** It has been recently shown that *tbx1* controls outflow tract identity and coronary patterning in mouse mutants. The subaortic or subpulmonary myocardial identity of the ventricular outlet in the *tbx1*-null mouse is related to the positioning of the coronary ostia (CO) in this model of common arterial trunk (CAT).

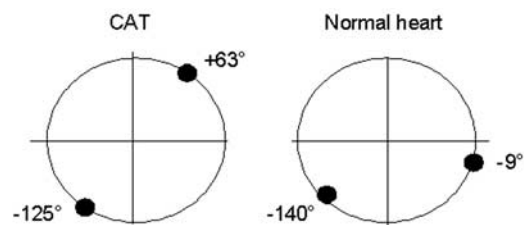
**Aim - hypothesis:** To review our specimens of human CAT to describe CO position and anomalies to attempt to show that the potential "pulmonary identity" of the CAT influences coronary patterning.

**Methods:** We reviewed 46 heart specimens with CAT and 10 with normal anatomy. Anomalies of CO ostia were described. Position of

the CO over the truncal circumference was measured in degrees as the direction from the middle of the truncal valve orifice (figure).

**Results:** CAT types were: 28 type I, 8 type II, 3 type III, 7 type IV. The truncal valve bicuspid in 2, tricuspid in 36, and quadricuspid in 7. Overall, 87% cases had malformations of the CO size and shape, location relative to the commissures or to the sinotubular junction. Left CO was abnormal in 33/46 (slit-like in 16, pinpoint in 10, tangential in 7) while right CO was abnormal in 20/46 (p < 0.01): slit-like in 10, pinpoint in 8, tangential in 2. Left CO was located above a commissure in 17% vs 2% for right CO (p < 0.05) and above the sinotubular junction in 28% vs 10% (p = 0.053). Left CO was always located on the dorsal-left part of the truncal circumference (median direction 63°, range -14° to +131°, p < 0.002) and right CO was always located on the ventral-right part of the truncal circumference (median direction -125°, range -179° to +179°, p < 0.05). The median angle between the 2 CO was 147° in CAT specimens vs 236° in normal hearts (figure).

**Conclusion:** Abnormal position and ostial malformations of the left CO in CAT is more frequent than for the right CO. This might be due to the obligatory dorsal connection of left CO because of the potential presence of coronary-refractory subpulmonary myocardium in the ventral left part of the CAT.



## P-60

**Home INR monitoring of oral anticoagulation in children using a point-of-care INR monitor**

Bajolle F., Lasne D., Elie C., Souillard P., Bonnet D. M3C-Necker, Université Paris Descartes, Paris, France.

**Background:** Children receiving Vitamin K Antagonist require more frequent monitoring than their adult counterparts. Point-of-care International Normalized ratio (POC INR) monitor may help to optimize anticoagulation control and to improve comfort of surveillance in children.

**Aim:** To evaluate outcomes of home INR monitoring in children through a standardized education/training program.

**Methods:** Since September 2008, POC INR monitoring has been proposed in children receiving Vitamin K Antagonist. Parents had to complete a one-day intensive education/training program. After demonstrating theoretical and practical competences, home monitoring was started. All INR results were collected by one of us for dose adaptation (self control but not self management). The proportion of INR within the target range, incidence of anticoagulation adverse events and quality of education program were analyzed.

**Results:** Fifty children (mean age 6.8 ± 4.7 years - range 2 months, 17 years) were included. Indications for anticoagulation were: 25 mechanical valves (mitral 18, aortic 7); 15 total cavopulmonary connections; 7 dilated cardiomyopathies; 2 giant aneurysms after Kawasaki's disease; 1 antiphospholipids syndrome. Thirty patients were naïve for Vitamin K Antagonist therapy and 20 patients were switched from venipuncture to POC INR monitoring. The results of theoretical test of parents were excellent (mean 17,3/20). Three families had transitory practical difficulties at home. During the observation period (279 ± 102 days),

INR was in the target range in  $79 \pm 17\%$  of cases. The number of INR/months was  $3.3 \pm 1.3$  (2 measurements systematically prescribed). There were no haemorrhagic and thrombosis complication. The education program was efficient as the number of home controls was always performed when requested and parents did not perform not requested controls. After 6 months, all families wished to pursue anticoagulation surveillance using home INR monitoring. **Conclusion:** Use of POC INR monitor was safe in our series. It did not lead to unnecessary INR controls. Families comfort was improved. A strict education/training program with recurrent controls of the quality of education is mandatory.

### P-61

#### Prediction of tilt test results based on patient's history in children with neurocardiogenic syncope.

Wieniawski P., Janiec I., Werner B.

Department of Pediatric Cardiology and General Pediatrics Medical University of Warsaw

**Objectives:** to analyse the relation of patient's history to tilt test results.

**Methods:** 29 children were involved in the prospective study: 20 girls, 9 boys aged 10–18 years (mean 15,8 years). 29 tilt tests were performed.

Tilt tests were performed according to Westminster protocol. Blood pressure was recorded by photoplethysmographic continuous automatic noninvasive method (using calf on finger) and automatic oscillometric method (using arm calf). Heart rate was continuously monitored and recorded by Holter ECG.

Neurocardiogenic response was classified according to VASIS classification.

Prodromal symptoms reported in history	Negative head-up tilt test n = 15	Positive head-up tilt test n = 14	p
blurred vision	5	7	ns
Dizziness	6	4	ns
Flushes	4	5	ns
Nausea	2	4	ns
breathlessness	2	2	ns

Syncope in anamnesis	Negative head-up tilt test	Positive head-up tilt test	p
>5	2	6	ns
4–5	3	3	ns
2–3	7	4	ns
1	3	1	ns
Presyncope in anamnesis			
many	3	3	ns
few	9	6	ns
1–2	3	5	ns

Number and frequency of syncope and type of prodromal symptoms reported in anamnesis were compared in patients with positive and negative tilt test results. Additionally symptoms preceding syncope during test and those reported in anamnesis were compared in the group of children with positive tilt test. In statistical analysis Fischer's exact test was used.

**Results:** 14 (48%) children had positive tilt test results. The main reported symptom was unspecified discomfort in 100% of patients, followed by blurred vision in 41%, dizziness in 35%, flushes in 31%, nausea in 21% and breathlessness in 14%. Chest discomfort, numbness of face and hands were reported less frequently. In 9 (31%) patients mixed type of vasovagal reaction was diagnosed, in 4 (14%) vasodepressive type, in 1 (3%) cardiodepressive type.

Symptoms preceding syncope during tilt test and those reported in history were consistent in 100% of children with positive tilt test result.

**Conclusions:** 1. Number of syncope and presyncope reported in anamnesis do not influence tilt test results. 2. No relation between prodromal symptoms reported in anamnesis and positive tilt test result was found.

### P-62

#### The reference values of aortic pulse wave velocity (PWVao) in a healthy population aged 3–18 years and the plausible physiological background of the observed changes

Hidvégi E. (1), Huberth J. (2), Jáklí Gy. (3), Kelemen I. (4), Klemencz H. (5), Illyés M. PhD (6)

Dr. Jakab Ltd. Outpatient's Dept. of Paediatric Cardiology, Szolnok, Hungary (1); 5th GP District, Tapolca, Hungary (2); Paediatric Praxis, Dabas, Hungary (3); Paediatric Praxis, Pilisvörösvár, Hungary (4); GP Praxis, Dabas, Hungary (5); Heart Institute, Faculty of Medicine, University of Pécs, Hungary (6)

**Objectives:** The assessment of the reference values of PWVao in a healthy population aged 3–18 years and to find the physiological background of the observed changes.

**Methods:** We performed arterial stiffness measurements on 4258 healthy children (2263 boys, 1995 girls) aged 3–18 years, using a validated, oscillometric, occlusive method (Arteriograph, TensioMed Ltd. Budapest, Hungary). This method is simple, fast and user-independent, which allowed us to perform measurements on large population in nursery, elementary and grammar schools. Written consent was given by the parents. The examinations were performed in calm, supine position. We used the sternal notch – pubic bone distance for assessing aortic PWV.

**Results:** The mean PWVao values are shown in Table 1, according to age in boys and girls.

Age (years)	Boys		Girls		Student's t-test
	n	PWVao (m/s) $\pm$ 2SD	n	PWVao (m/s) $\pm$ 2SD	
3	37	5.57 $\pm$ 0.59	31	5.59 $\pm$ 0.65	NS
4	44	5.60 $\pm$ 0.63	35	5.44 $\pm$ 0.51	NS
5	63	5.59 $\pm$ 0.63	36	5.37 $\pm$ 0.78	NS
6	102	5.49 $\pm$ 0.66	56	5.37 $\pm$ 0.69	NS
7	74	5.54 $\pm$ 0.51	59	5.62 $\pm$ 0.61	NS
8	57	5.58 $\pm$ 0.52	34	5.44 $\pm$ 0.62	NS
9	79	5.69 $\pm$ 0.56	45	5.53 $\pm$ 0.61	NS
10	65	5.67 $\pm$ 0.65	44	5.85 $\pm$ 0.60	NS
11	65	5.69 $\pm$ 0.59	40	5.70 $\pm$ 0.57	NS
12	83	5.82 $\pm$ 0.61	74	5.83 $\pm$ 0.53	NS
13	151	5.84 $\pm$ 0.59	159	5.97 $\pm$ 0.58	NS
14	166	6.04 $\pm$ 0.62	160	5.93 $\pm$ 0.55	NS
15	137	6.08 $\pm$ 0.58	154	5.99 $\pm$ 0.53	NS
16	139	6.24 $\pm$ 0.51	146	6.10 $\pm$ 0.48	NS
17	175	6.32 $\pm$ 0.53	157	6.22 $\pm$ 0.47	NS
18	106	6.45 $\pm$ 0.47	96	6.43 $\pm$ 0.48	NS

The mean PWVao did not show any significant differences in any age group between genders and increased from

5.6 ± 0.6 m/s to 6.4 ± 0.7 m/s in both genders between 3–18 years. The PWV<sub>ao</sub> did not show any significant correlation with heart rate. However a very significant correlation was found between PWV<sub>ao</sub> and SBP, MAP, BSA, BMI and body weight in both genders.

**Conclusions:** The more elastic the aortic wall, the lower the PWV<sub>ao</sub>. The observed increase in the PWV<sub>ao</sub> refers to the wall stiffening with age. Although atherosclerotic plaques may occur even in childhood, but their prevalence between 15–19 years is rather small (<13%), so its effect on the PWV<sub>ao</sub> in our materials is negligible. The observed relationship between PWV<sub>ao</sub> and MAP, BSA, BMI, and body weight sheds light on the physiological background of the increase of PWV<sub>ao</sub>. The aortic stiffness could be increased not only due to morphological changes of the vessel wall, but also due to functional changes, i.e. MAP, stroke volume, cardiac output. Thus the increasing MAP causes pressure elevation, the increasing BSA, BMI, body weight cause volume loading which leads to stiffening of the aortic wall. The question whether the stiffening of the aorta due to pathological increase of MAP, BSA, BMI, and body weight may play etiological role in the later developing atherosclerosis, remains to be elucidated by further prospective studies.

### P-63

#### **Incidence and risk factors of restenosis after surgery for subaortic stenosis**

*Varan B (1), Gokdemir M. (1), Tokel K. (1), Cindik N. (1), Ozkan S. (2), Ozkan M. (2), Aslamaci S. (2)*

*Baskent University Faculty of Medicine, Ankara, Turkey, Pediatric Cardiology Unit (1) and Department of Cardiovascular Surgery (2)*

**Introduction:** Restenosis is the most common cause of reoperation after surgery for subaortic stenosis (SAS). Residual stenosis, inadequate resection, abnormal healing and fibrosis, abnormal tissue proliferation may lead restenosis. Incidence of restenosis is still high despite successful surgery. This study aimed to determine the incidence and risk factors of restenosis after surgery for SAS.

**Patients and Methods:** 188 patients operated for SAS between January 1995 and December 2007 were retrospectively evaluated. Age and body weight at the time of surgery, previous surgery and catheterization, surgical technique, early and late echocardiographic examination results were noted. Patients were reevaluated for restenosis and risk factors were determined.

**Results:** 81% of the patients had an associated cardiac lesion and 19% had isolated SAS. 182 patients had a discrete SAS and 6 had tunnel type SAS. Operation age was 5.79 ± 4.18 (0.07–16.14) years and M/F: 2.83. In 17 patients SAS developed during follow-up of an other cardiac lesion. 56.5% of patients underwent membranectomy and septal myectomy, 41.5% had membranectomy and 2% had more invasive procedures. Left ventricular outflow tract (LVOT) echo gradient decreased from 48.5 ± 32.5 (7–150) mmHg to 10.4 ± 11.2 (1–88) mmHg ( $P < 0.001$ ). 31 of 188 patients (16.5%) had restenosis (gradient >30 mmHg) and 16 patients had surgery for restenosis 4.4 ± 3.6 (0.6–15.5) years after the first operation. The mean age of the patients at the last visit was 10.5 ± 5.6 (1.6–32.6) years after a follow-up of 5.51 ± 3.68 (0.61–18.7) years. Small aortic annulus, high preoperative echo gradient, high postoperative echo gradient, isolated SAS, discrete membranous type stenosis and early postoperative echo gradient ≥10 mmHg were risk factors for restenosis.

**Conclusion:** Patients with mild stenosis can be followed conservatively however progressive stenosis must be considered as an indication for surgery to decrease risk of restenosis.

### P-64

#### **Cardiovascular abnormalities in children with Alagille syndrome**

*Tirska-Kmiec A., Mirecka-Rola A., Zuk M., Biernatowicz M., Daszkowska J., Rewers B., Ziolkowska L., Kowalczyk M., Kawalec W., Jurkiewicz D, Zubrzycka M.*  
*Children's Memorial Health Institute, Warsaw, Poland*

**Introduction:** Alagille syndrome (AGS) is one of the most common autosomal dominant inherited disorder (JAG1 mutations) that cause chronic liver disease in children, characterized also by cardiovascular abnormalities (CVA).

**Objectives:** Aim of this study was to evaluate the spectrum and prognosis of congenital CVA in patients (pts) with AGS.

**Patients and methods:** Retrospective chart review of 30pts with AGS was performed. A transthoracic 2-D echo and radioisotope pulmonary angiography was performed in 30 pts. Mean age at AGS diagnosis was 6months, at last examination 11.6 yrs. Categorical variables were compared by chi-square test.

**Results:** AGS was diagnosed according to conventional criteria by the association of at least 3 of the following 5 abnormalities: chronic cholestasis (23/30 = 77%), pulmonary branch or peripheral stenosis (PS)(24/30 = 80%), “butterfly-like” vertebral defects (15/30 = 50%), embryotoxon (19/30 = 63%), peculiar facies (29/30 = 97%). Successful liver transplantation underwent 2/9pts with end stage liver disease (ESLD) and nonsignificant PS. Cardiological examination revealed heart murmur and CVA in 29pts (97%): isolated congenital heart defect (CHD) – 5, isolated PS – 14, CHD with PS – 10. All together CHD had 15 pts(30%) (complex cyanotic – 4, increased pulmonary flow – 6, other -5). Among all 24pts with PS: 7 – had peripheral PS, 4 – hypoplastic pulmonary arteries (HPA), 13 – right and/or left pulmonary branch stenosis. Jagged1 mutations were identified in 8/18 diagnosed pts (44%). Statistical correlations were found between the presence of: JAG1 mutation and PS (7/8pts,  $p < 0.05$ ), ESLD and PS (8/9pts,  $p < 0.05$ ). Cardiac catheterization was performed in 9/30 pts (30%) with significant CVA, 7/9pts had also 1–2 (total 10) percutaneous interventions (balloon atrioseptostomy-1, Amplatzer septal occluder – 1, pulmonary balloon valvuloplasty – 2, pulmonary balloon angioplasty – 4, pulmonary artery stenting – 2), 5/9pts underwent 1–3 (total 10) cardiosurgical procedures (palliative – 6, corrective – 4). Four pts (13%) died (3 had complex cyanotic CHD with HPA, 1 ESLD).

**Conclusions:** CVA had 97% of children with AGS patients and 30% of patients with severe forms of PS or complex cyanotic CHD needed percutaneous and/or cardiosurgical intervention. Children with AGS associated with complex CHD and hypoplastic pulmonary arteries had high mortality.

### P-65

#### **Non-invasive cardiac output determination in children with congenital AV block treated by ventricular pacing**

*Wiegand G., Kerst G., Baden W., Hofbeck M.*  
*Department of Pediatric Cardiology, University Children's Hospital, Tuebingen, Germany*

**Background:** The standard methods for determination of the cardiac output (CO) are either invasive or technically demanding. CO measurement by the inert gas re-breathing (IGR) method uses a low concentration mixture of an inert and a blood-soluble gas respectively. The aim of our study was to compare CO in resting and during exercise conditions in children with 3rd degree congenital AV block (AVB III) stimulated with a permanent VVI pace-maker.

**Patients/Controls:** 5 female AVB III patients (age 9.2–17.4 years) with VVI-R pacing were included in the study. Five healthy, age-matched boys and girls respectively, served as controls.

**Methods:** The Innocor® system is based on the IGR principle. Patients are breathing over a period of 30 seconds a low concentration mixture of an inert and a blood soluble gas from a closed system. The pulmonary blood flow (PBF) absorbs the blood soluble component, the rate of absorption being proportional to the PBF; the higher the PBF the higher the absorption rate will be, while the inert component is expired unaltered. To find out the CO during exercise the standard treadmill protocol of the German Society of Pediatric Cardiology was used. Cardiac index (CI), stroke volume (SV) and the heart rate (HR) were determined during rest and two standardized exercise levels.

**Results:** The patients and the female controls had a lower resting CI compared to male controls (CO = [l/min]; CI = [l/min/m<sup>2</sup>]). The CI increased in all the subjects during exercise. Patients reached only a mean CI of 5.9 l/min/m<sup>2</sup>, while the controls reached a CI of 7.3 (boys) and 7.2 l/min/m<sup>2</sup> (girls) respectively. The SV increase under exercise was also reduced in patients compared to controls.

**Discussion:** According to our experience the IGR method allows estimation of CO in resting conditions as well as under exercise. Under exercise AVB III patients paced in a VVI-R modus increase their CI less than healthy controls. The HR response to exercise can be optimized by pace-maker reprogramming and the subsequent improvement of the CI can be verified by the IGR method.

## P-66

### The CIVIC Study: Hospitalizations for acute respiratory infections in young children with significant heart disease. Four seasons (2004–2008) in Spain.

Medrano C., Garcia-Guereta L., Grueso J., Insa B., Ballesteros F., Casaldaliga J., Cuenca V., Escudero E., Garcia-de la Calzada D., Luis M., Mendoza A., Prada E., Rodriguez MM., Suarez P., Quero C., Rueda E., Gomez H.

On behalf the CIVIC Study group from the Spanish Society of Pediatric Cardiology and Congenital Heart Disease (SECPCC).

**Objectives:** To investigate the epidemiology of acute respiratory tract infections (ARI) in children younger than 24 months old with hemodynamically significant congenital heart diseases (HSCHD). Primary aim: incidence of hospital admission due to ARI. Secondary aims: risk factors, etiological agents, clinical outcomes and usefulness of preventive measures.

**Methods:** Four seasons (from October to April, from 2004 to 2008), epidemiological, multicentric (57 hospitals in Spain), observational and prospective study. 2613 patients were eligible for the study.

**Results:** 344 patients were hospitalized with a respiratory infection with a rate of 13,5% and a total 453 hospital admissions. The most frequent clinical diagnosis were bronchiolitis (54,1%), upper respiratory tract infection (21%) and pneumonia (19,9%). In two-thirds of the patients no etiological agent was identified. In the remaining patients either a single agent (26,8%) or polymicrobial infection (5%) was identified. Respiratory syncytial virus (RSV) was the agent most commonly found (in 96 patients with a 3,8% specific RSV hospitalization rate). Prophylaxis against RSV, recommended in all cases, was completed in 90,5%. Complete versus non-complete RSV prophylaxis reduced a 58,2% the relative risk for RSV hospitalization: 3,3% versus 7,9% rates. Significant risk factors for ARI admission included wheezing, chromosome alterations (22q11 deletion, Down syndrome), siblings <11 years of age and inadequate RSV prophylaxis fulfillment.

The protective factors included no malnourishment, female gender and older age (>12 month). See table:

CIVIC	OR	Lower Limit 95%CI	Upper Limit 95%CI
<b>Risk factors</b>			
Wheezing	3,09	2,05	4,66
22q11 deletion	2,84	1,38	5,84
Down syndrome	1,97	1,45	2,70
Siblings <11 y/o	1,85	1,35	2,54
Incomplete RSV prophylaxis	1,77	1,23	2,53
<b>Protective factors</b>			
Weight >3 percentile	0,75	0,58	0,97
Gender (female)	0,70	0,55	0,89
Age >12 m	0,41	0,27	0,62

Hospitalization median stay was 7 days. PICU care was required in 21.8% of the admissions and nine patients (0.34%) died for ARI.

**Conclusions:** Hospital admission rate and severity of ARI infection remains an important issue in HSCHD patients. Direct cardiac risk factors were not found. The strict fulfillment of prophylactic recommendations against RSV is the only protective factor that can be modulated in order to decrease ARI hospital admission rate.

## P-67

### Longitudinal data on ascending aortic dimension in Turner patients

Fournier A., Hugues N., Bruçada G., Mivelaz Y., Deal C., Thambo J.B., Dahdah N.  
CHU mère-enfant Ste-Justine, Montréal, Québec, Canada

**Introduction:** Dilatation of the ascending aorta is a well-known cardiac anomaly in patients with Turner syndrome (TS), and can lead to aortic dissection. The purpose of this study was to analyze prevalence, risk factors, evolution and effect of treatment for this anomaly in patients with TS.

**Methods:** Retrospective analysis for all patients with TS followed in our institution from 1992 to date was performed. Aortic dimensions were obtained by echocardiographic studies and converted to Z scores using data from normal control females of our laboratory. Data was collected concerning associated congenital heart defects and particularly bicuspid aortic valve, growth hormone treatment, caryotype (45, X0 vs. mosaic pattern) and use of beta-blockers or vasodilators.

**Results:** Among 120 individuals with TS, 27.5% had associated congenital heart defects (CHD) the most frequent one being: bicuspid aortic valve (18 patients, 15%), coarctation of the aorta (14 patients, 11.6%), aortic stenosis (7 patients, 5.9%) and left superior vein cava (3 patients, 2.7%). In total, 29.2% were under growth hormone treatment during the course of the evaluation; caryotype showed monosomy in 63% and mosaic forms in 37%. At first echo (mean age of 9.8 years (0.2–44)), 29.4% of patients had aortic dilatation, which increased to 48.6% at the end of a mean follow-up of 8.15 years (0.53–15.71). The progression of the dilatation was independent of the intake of growth hormone, caryotype or presence of CHD, even bicuspid aortic valve. Moreover, significant progression was noted in females with normal hearts (mean Z score at first echo  $1.31 \pm 1.69$  vs follow-up echo  $1.89 \pm 2.02$ ;  $p = 0.014$ ). Treatment with beta-blockers or angiotensin converting enzyme inhibitors reduce the Z score

of the ascending aorta without reaching statistical significance in our study (18 treated patients during 25.7 month (1–82)):  $3.3 \pm 1.97$  to  $2.97 \pm 1.55$ .

**Conclusions:** Risk of aortic dilatation is high in females with TS. This dilatation is independent to other cardiac anomalies, growth hormone intake or karyotype. Pharmacological treatment tends to prevent further dilatation. Due to critic consequences of progressing dilation, indefinite surveillance of these patients is highly recommended, even in the lack of other associated cardiac malformations.

#### P-68

##### **Steroids and Vascular Resistance Reducing Agents Improve Cardiac Outcome and Survival in Patients with Duchenne Muscular Dystrophy**

Fournier A., Thérien J., Schram G., Dahdah N.

CHU mère-enfant Ste-Justine, Montréal, Québec, Canada

**Introduction:** Cardiac involvement is a non equivocal complication of Duchenne muscular dystrophy (DMD), 59% will have developed dilated cardiomyopathy (DCM) at 10 years of age and 100% at 18 years. It is also suggested that treatment with corticosteroids (CS) and/or angiotensin converting enzyme inhibitors (ACEi) improves survival and delays the onset of DCM in DMD.

**Methods:** The cardiac condition of a consecutive longitudinal cohort of 98 boys with DMD was analyzed.

**Results:** Initial age of cardiac evaluation was  $8.1 \pm 3.2$  years [range 2.2–16.6]; age at follow-up was  $16.4 \pm 6.7$  years [2.0–34], with a follow-up duration of  $7.9 \pm 4.6$  years [0–25], 66 patients were treated with CS starting at  $9.1 \pm 3.7$  year-old [4.1–22.9], and 88 were treated with ACEi or angiotensin receptor antagonist (ARA), starting at  $12.2 \pm 4.1$  year-old [5.7–31.7]. The incidence of DCM was 46/98 patients (47%) defined as left ventricular end-diastolic diameter Z score (LVdZ)  $> 2$  and/or a shortening fraction (SF)  $< 28\%$ . The mean age at DCM diagnosis was  $14.3 \pm 3.8$  years [8.7–26.1]. Overall, the DCM was mild (SF 25–28% and/or LVdZ  $> 2$ ) in 28 pts (61%), moderate (SF 20–24%) in 10 (22%) and severe (SF  $< 20\%$ ) in 8 (17%). The incidence of moderate-to-severe DCM among those with or without CS therapy was 12% vs 31% respectively ( $p < 0.05$ ). In comparison with historic data, there were no DCM observed at 10 years of age (compared to 59%), only 16% at 14 years and 23% (compared to 100%) at 18 years. Overall survival was also remarkable in our cohort, with 11 (11.2%) cardio-respiratory and 2 accidental deaths, all with DCM except the youngest one who died in a car accident. Age at death was  $20.3 \pm 4.8$  years [8.7–26.1], compared to death in mid-adolescence in historic data.

**Conclusion:** Our data supports the beneficial effect of CS therapy for DMD in terms of survival rate and onset of DCM. This is probably enhanced with ACEi or ARA therapies, although our data do not allow precise determination due to combined therapy in most cases.

#### P-69

##### **Doppler tissue imaging analysis of right ventricular myocardial velocities in the third trimester fetuses and normal neonates – preliminary study**

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

(1) Perinatal Cardiology Dept., 2nd Dept. of Obstetrics and Gynecology, Medical University of Warsaw; (2) Neonatology and neonatal Intensive Care Dept., Medical University of Warsaw

**Objectives:** The aim of this study was to compare myocardial velocities measured by Doppler tissue imaging (DTI) of the right

ventricular (RV) wall in the third trimester fetuses and normal newborns in the first day of life.

**Methods:** The pulsed Doppler DTI was measured using Sequoia 512 in fetuses and Philips HDI11 in neonates. Peak myocardial diastolic and systolic velocities were measured by Doppler tissue imaging of the RV wall in 30 normal singleton fetuses at 35 to 40 weeks of gestation and 10 normal neonates in the first day of life after term delivery (37–41 weeks). The sample volumes were placed in the basal part of RV in the apical 4-chamber view.

**Results:** Mean fetal heart rate was  $140 \pm 10$  bpm. Mean neonatal heart rate was  $128 \pm 11$  bpm.

The table shows comparison of the right ventricular peak myocardial velocities in fetuses and newborns.

DTI	Fetuses		Newborns		p
	mean	SD	mean	SD	
Em	8,3	1,8	9,7	2,3	0,077
Am	10	1,8	10	3,8	0,362
Em/Am	0,82	0,13	1,03	0,27	0,026
Sm	6,7	1,9	6,3	1,1	0,833

**Conclusions:** Pulsed Doppler DTI is feasible method for evaluation of the fetal and neonatal heart function. Fetal right ventricular peak myocardial velocities during atrial contraction (Am) and systole (Sm) were similar to the newborns in the first day of life. Peak myocardial velocities during early diastole (Em) and Em/Am ratios were slightly higher after delivery what is most likely determined by improvement of diastolic function, but further studies are necessary to prove this findings.

**Acknowledgments:** This study was partially funded by grant: MZiNSW No NN 407414336.

#### P-70

##### **Acute rheumatic fever outcomes in 602 children; Kayseri/Türkiye experience**

Narin N, Mutlu FT, Baykan A, Hanedan Onan S, Sezer S, Uzum K, Baykan Z

Erciyes University Medical Faculty, Department of Pediatric Cardiology Kayseri/Turkey

**Introduction and aim:** Acute rheumatic fever (ARF) incidence decreased in developed countries, but it is still an important public health problem in developing countries including our country. The aim of this study is to share our 12 years experience about ARF.

**Materials and methods:** 602 patients met Modified Jones Criteria from January 1997 to December 2008 were evaluated retrospectively.

**Results:** The mean age was  $11.1 \pm 2.7$  (2.5–18 years) and female/male ratio was 1.3. Family history was 9% positive. Most common presentation complaint was joint pain, other complaints were involuntary movement, joint swollen, chest pain, hearing cardiac murmur, palpitation. Out of 602 patients 443 patients were diagnosed on acute phase and 159 had sequel of ARF during diagnosis. Carditis incidence was 58% (8.3% subclinical carditis), arthritis incidence was 44.5%, chorea incidence was 25%, subcutaneous nodules incidence was 0.5%, and erythema marginatum was not present in any patient. The most two common major criteria were carditis and chorea accompaniment (32%). Chorea was seen especially in prepubertal girls. Mean follow-up time was  $35.3 \pm 30.0$  months (3–120 month). Mitral regurgitation decreased at the rate of 26.9% and aortic regurgitation decreased at the rate of

17.4% at the end of first year. Mitral and aortic regurgitation decreased at the rate of 32% and 20.6%, respectively at the end of five years follow up. In 12 years period 17 patients had recurrent attack and 13 patients operated for valve replacement. No patient died during this period.

**Conclusion:** Increased levels (incidence) of monoarthritis and chorea should be taken into consideration. In our opinion, because of decreased incidence of subcutaneous nodules and erythema marginatum they should be reevaluated as a major criteria, and give more importance to the changing face of ARF. Even acute rheumatic fever is not a common cause of mortality but it continues to cause significant morbidity in children in Kayseri.

#### P-71

##### **Surgical treatment of aortic coarctation within the first year of life: a 10 year review (clinical, surgical, biomechanical aspects)**

*Teivane E., Lais A., Ozolins V., Smits L., Sikora N., Bergmane I Children's Clinical University Hospital, Department for Pediatric Cardiology and Cardiac Surgery, Riga, Latvia*

**Introduction:** We analyzed a single institution 10-year experience with coarctation repair within the first year of life to define risk factors for recoarctation, evaluate morbidity, mortality, short and long term results.

**Materials and methods:** Retrospective and prospective study/follow-up of all (71) patients younger than 12 months undergone aortic coarctation repair in our institution between January 1, 2000 and December 31, 2009.

**Results:** The patients were 42 boys and 29 girls divided into two groups according the age at the time of repair: neonates < 30 days old n = 38 (53%) (mean age 11,6 (2–30 days)), infants > 30days old n = 33 (47%) (mean age 2,9 months +/-1,8). The mean weight at surgery for neonates was 3,3kg (+/-0, 5). Patients were dividend into three groups: group I- simple coarctation (neonates n = 16, infants n = 24), group II - coarctation and VSD (neonates n = 11,infants n = 3), group III -different complex intracardiac lesions(neonates n = 11 infants n = 6). Infantile coarctation was present in 46 cases (65%), hypoplastic arch in 11 cases.

Techniques for primary repair included resection with extended end-to-end anastomosis in neonatal group n = 4/older babies n = 3, subclavian flap aortoplasty (SFA) n = 29 neonates/n = 18 babies, resection with simple end-to-end anastomosis (ETE) n = 4 neonates/ n = 12 infants.

Early mortality n = 4 (5,6%) (neonates), late mortality n = 9 (12, 7%) (neonates).

Slight shortening of the left arm found in 3 patients, asymmetry of palms in one patient in the group of Waldhausen's operation (8,5%). No paraplegia occurred. Study includes follow-up data from 63 patients (mean follow up 4, 3 years (range 0–9 years)). Recoarctation occurred at the age 2 months–7 years in 12 from 67 patients (17, 9%) (Early death patients excluded), most frequent with ETE and SFA repairs (4/8 cases respectively). Patients required balloon angioplasty and all except one have no significant residual gradient. There was no mortality or complications after reinterventions.

**Conclusions:** Infantile aortic coarctation in neonatal age is a significant problem and carries high mortality and recoarctation rate. Baloon angioplasty of recoarctation is safe and effective.

Mortality is most importantly influenced by preoperative status, the severity of the associated anomalies, perioperative intensive treatment. There were no severe ischemic complications of the left arm connected with subclavian flap aortoplasty.

#### P-72

##### **Williams Beuren syndrome may present as neonatal coarctation**

*Kammache I. (1), Acar P. (2), Amedro P. (3), Fouilloux V. (1), Ou P. (4), Kreitmann B. (1), Fraisse A. (1).*

*La Timone Children's Hospital, Marseille, France(1); Purpan Children's Hospital, Toulouse, France (2); Arnaud de Villeneuve Children's Hospital, Montpellier, France(3); Necker Children's Hospital, Paris, France*

**Introduction:** Williams Beuren syndrome in neonates is most frequently asymptomatic. However, there are few reports on neonatal presentation with aortic coarctation. In this study we aim to determine the frequency, the clinical profile as well as the outcome of patients with Williams Beuren syndrome presented with neonatal coarctation.

**Methods:** Between January 1996 and June 2007, 135 infants less than 3 months of age were operated for aortic coarctation in our institution. Among them, 3(2.2%) were diagnosed with Williams-Beuren syndrome.

**Results:** In all 3 cases, the diagnosis of Williams syndrome was confirmed by the FISH analysis showing deletion of 7q11.23. The 3 infants with Williams-Beuren syndrome presented with congestive heart failure and absent femoral pulses at a mean age and weight of 30( $\pm$ 15) days and 3.4( $\pm$ 0.6) kg, respectively. The diagnosis of coarctation was confirmed by echocardiography. Associated cardiac anomalies were a ventricular septal defect and a small ostium secundum atrial septal defect in one case, and a bicuspid aortic valve in another patient. The 3 patients were operated at a mean age and weight of 51.6( $\pm$ 32.7) days and 3.5 ( $\pm$ 0.6)kg, respectively. Resection of the coarctation segment with end-to-end anastomosis was performed through left thoracotomy in 2 cases. The last patient was operated though median sternotomy and under cardiopulmonary bypass with large enlargement of aortic arch, associated with atrial and ventricular septal defect closure. The 3 patients remained hypertensive and presented with recoarctation 2, 2.3, and 14.3 months after initial repair, respectively. Transcatheter balloon dilation was unsuccessful in all cases. A surgical reoperation was necessary 0.6, 1.2 and 10.9 years after initial repair, respectively. Patch enlargement of the aorta was performed in two cases, whereas the third patient necessitated an extra-anatomic conduit because of long and severe aortic stenosis. After a mean follow-up of 3.4(+/-07) years, the patients are asymptomatic without recurrent aortic obstruction and discontinuation of antihypertensive treatment.

**Conclusion:** Williams-Beuren syndrome presenting with neonatal coarctation is characterized by recurrent aortic obstruction after initial surgery, non responsive to balloon dilation. Reoperation is successful but often complex with patch enlargement and/or exclusion of the stenotic aortic segment.

#### P-73

##### **Severe pulmonary hypertension in bronchopulmonary dysplasia: high incidence of associated cardiovascular abnormalities**

*Cerro MJ., Deiros L., Labrandero C., Guereta L., Rubio D., Fernandez A., Burgueros M., Bret M., Barrio M., Martinez MC., Diaz C., Menendez J.*

*Hospital Universitario La Paz. Madrid.Spain*

**Background:** Pulmonary bronchodysplasia can be complicated with pulmonary hypertension (PH). Incidence, clinical characterization, and outcome of PH in this setting haven't yet been fully established.

**Patients and methods:** Retrospective study of clinical records, MRI/multislice TC imaging, and Catheterization data of 18 patients referred to our Pulmonary Hypertension Unit with the diagnosis of pulmonary bronchodysplasia and PH (over 50% of systemic pressure in two consecutive echocardiographies), from 3/2006 to 12/2009. Age of the patients at the time of referral was 8,1 + 9 months (range 2–33 months).

**Results:** We performed multislice TC in 11 patients and cardiac catheterization in 10 patients. We found a high incidence of associated cardiovascular anomalies: Pulmonary vein stenosis in 6/18 (33%) patients (2 mild, 4 severe), variable degrees of hypoplasia of intrapulmonary arteries in 7 (38%), moderate/large Ductus arteriosus in 4 (22%, all diagnosed after 4 months of postnatal age), ASD in 5 patients (27%), VSD (2 patients), and tetralogy of fallot in one. We also found non vascular abnormalities: tracheal stenosis (n = 1), and partial left diaphragmatic hernia (n = 1). Due to late referral, diagnosis was delayed in some patients with moderate or large septal defects; some of whom had been initially misdiagnosed of small defects, due to bidirectional or predominant right to left shunting.

Mean follow-up was 11,8 + 7,8 months (range 1–24 months). Three patients died (16%) due to PH or complications of their lung disease. Seven patients underwent surgical closure of their septal defects; 6/7 had persistent postoperative severe pulmonary hypertension and needed vasodilator treatment. Two patients were considered unoperable due to cardiac catheterization findings. In the whole group, 11 patients received treatment with sildenafil, and three of them combined treatment with sildenafil+bosentan (n = 1), inhaled iloprost+ sildenafil (n = 1) or sildenafil+bosentan+subcutaneous treprostinil (n = 1).

**Conclusions:** Pulmonary bronchodysplasia (PBD) is a complex disease affecting not only lung parenchyma and airway, but also pulmonary vessels. In patients with pulmonary bronchodysplasia and pulmonary hypertension, the incidence of associated cardiovascular abnormalities is high. This patients should undergo complete and early evaluation in pulmonary hypertension units, in order to detect and treat cardiovascular abnormalities before irreversible pulmonary vascular disease develops.

#### P-74

##### **Intramyocardial Injection of Autologous Bone Marrow derived progenitor Cells in Case of Dilated Cardiomyopathy in Four Month Old Child**

Bergmane I. (1), Lubaua I. (1), Ozolins V. (1), Lacis A. (1), Narbutė I. (2), Jegere S. (2), Ramata-Stunda A. (2), Jakobsons E. (2), Erglis A. (2)

*Clinics for Pediatric Cardiology and Cardiac Surgery, Children's Clinical University Hospital, Riga, Latvia (1); Cell Laboratory, Latvian Institute of Cardiology, Pauls Stradins Clinical University Hospital, Riga, Latvia (2)*

**Introduction:** Dilated cardiomyopathy is a serious disorder of the myocardium in pediatric age. Conservative therapy is limited and lethal outcome observed in one third of patients within a year. Bone marrow derived progenitor cell transplantation is becoming a promising method of treatment in adult population and there is ground to believe there are perspectives in pediatric cardiology, especially in early age group where transplantation is limited due to lack of donors and technical difficulties.

**Case:** We demonstrate first case of documented autologous bone marrow progenitor cell intramyocardial implantation done in 21.05.2009. in Children's Clinical University Hospital in collaboration with Latvian Center of Cardiology, Pauls Stradins Clinical University Hospital and Cell laboratory. There were

signs of cardio-vascular insufficiency observed already from the first weeks of life from the data of case-record. At the age of 4 months the child was admitted to intensive care unit due to severe congestive heart failure, severe respiratory insufficiency and marked hepatomegaly. Laboratory investigations revealed no inflammatory or autoimmune process, also genetically determined metabolic disorders were excluded. Despite massive therapy of cardio-vascular insufficiency cardiac dilatation, low cardiac output state remained stable and the child was in need of mechanical ventilation. The decision to perform bone marrow progenitor cell intramyocardial implantation was made due to critical condition of the patient and no possibilities for heart transplantation. The procedure was technically performed without side effects or deterioration of the patient. The stabilization of the child was observed following the procedure, 1 week following transplantation the child was extubated and discharged from the hospital three weeks later. Within the year of follow-up dilatation of the left ventricle has decreased LVDd 47mm to 41mm, contractility has improved from 5% to 19%, ejection fraction improved from 11% to 42%. Thoracic X-ray showed decrease of cardiomegaly: cardiothoracic ratio decreased from 0,75 to 0,56. Psychomotor development of the child is normal for patient's age, positive weight gain observed. There were no signs of cardio-vascular deterioration due to intercurrent illnesses within the year. **Conclusion:** This first case let as hope that stem cell therapy might become approved method of therapy of dilated cardiomyopathy.

#### P-75

##### **Congenital Heart defects in Kabuki Syndrome at North-Eastern Region of Romania**

Iordache C. (1), Luca A.C. (1), Panzaru M.C. (2), Volosciuc M. (2), Braha E. (2), Moisa S.M. (1)

*1st Pediatric Cardiology Clinic, "Sfanta Maria" Children Emergency Hospital, Iasi, Romania (1); Medical Genetic Center, "Sfanta Maria" Children Emergency Hospital, Iasi, Romania (2)*

**Introduction:** Kabuki syndrome (KS) is a congenital mental retardation syndrome with additional features, including distinctive facial features, dermatoglyphic abnormalities and short stature. Some cases associate heart defects. The underlying genetic mechanism remains unknown.

**Methods:** Prospective clinical and echocardiographic evaluation of consecutively recruited children at North-East Region of Romania with Kabuki syndrome.

**Results:** We have analysed the prevalence and types of congenital cardiac defects in 20 children with Kabuki syndrome recorded in the files of Iasi Medical Genetics Center. There were 7 girls and 13 boys, diagnosed at a median age of 4,9 years (range from 0,2 to 13 years). The diagnosis was based on the presence of the characteristic feature of the disorder.

Cardiac defects were present in 9 children (45%) – atrial septal defect (35%), coarctation of the aorta, aortic stenosis, bicuspid aortic valve (5%) and dextroposition (5%). The prevalence of heart defects was higher in boys (53,8%) than girls (28,5%). Complex heart defects were present only on males. In the literature aortic coarctation, atrial septal defect and ventricular septal defect are the most frequent congenital defect associated with Kabuki syndrome. The comparison of our data and the literature data will be presented in detail.

**Conclusions:** In conclusion we present a study of 20 cases with Kabuki make-up syndrome, 45% of them having heart defects. Cardiac defects are commonly associated to KS and males are more frequently and severely affected than females. Patient's



prognosis depends on the presence/absence of cardiac defects, renal failure and severity of mental retardation.

**Keywords:** Congenital heart malformations, Kabuki syndrome.

#### P-76

##### Romanian Study on Arterial Hypertension Prevalence and Cardiovascular Risk Factors on 5290 Children and Adolescents

Cinteza E, Gherghina I, Anca I, Iagaru N, Matei D  
Institute for Mother and Child Health, Bucharest, Romania; University of Medicine and Pharmacy, Bucharest, Romania

**Background:** Romania has the most important cardiovascular morbidity and mortality in European Union. Studies on arterial hypertension (AHT) and cardiovascular risk factors (CVRF) in pediatric population date 20 years ago. AHT prevalence in children is difficult to estimate, on one hand due to the lack of homogeneity in all pediatric ages and on the other hand it is not a common measurement, although it should.

**Purpose:** Evaluation of the new data regarding AHT and CVRF in children and adolescent in Bucharest and surrounding area.

**Methods:** A transversal study was performed on 5290 children and adolescents, aged 3–17 years, from 24 schools, kindergartens and high schools from Bucharest and Ilfov County. We measured weight, height, BP, calculated the body mass index (BMI). BP over the 95th percentile at three measurements was considered AHT. We also investigated associated possible risk factors: family history of cardiovascular diseases (FHCD), birth weight, smoking, energizing drinks consumption (coca/pepsi cola) (EDC), missing extra school sport activity (MESSA), length of the nocturnal sleep. We used VassarStats calculator and Student  $t$  test for statistics.

**Results:** AHT prevalence in the studied group was 7.36%. Overweight and obesity were present in 10.07%, and 6.39% comparing with 17.03%, and 21.22% in AHT group ( $p < 0.0001$ ). Underweight was present in 14.20% comparing with 7.26% in AHT group ( $p = 0.0002$ ). EDC was 11.73%. AHT was present in 9.72% of the more than 2 l/day consumers of EDC ( $p = 0.44$ , OR 1.355). Small birth weight was present in 1.39% comparing with 1.87% in AHT group ( $p = 0.51$ ). FHCD were present in 9.96% comparing with 13.12% in AHT group ( $p = 0.0563$ , OR 1.365). Smoking was present in 3.43% comparing with 3.35% in AHT group ( $p = 0.9362$ ), but in adolescents, 14–17 years, smoking was present in 10.49%. MESSA was present in 15.61% comparing with 15.64% in AHT group ( $p$  non significant). Nocturnal sleep shorter than 7 hours was present in 10.52% comparing with 10.05% in AHT group ( $p$  non significant).

**Conclusion:** Significant risk factors associated to AHT were overweight and obesity. The other evaluated factors were not proved to be risk factors although they were more present in the AHT group.

#### P-77

##### Morphometric analysis of coronary arteries after arterial switch procedure for transposed great arteries.

Moll M. (1), Michalak K.W. (2), Moll J.A. (2), Moszura T. (2), Kopala M. (1), Młodzik K. (2), Moll J.J. (1)  
Cardiosurgery Department, Polish Mother's Memorial Hospital, Lodz, Poland (1); Cardiology Department, Polish Mother's Memorial Hospital, Lodz, Poland (2)

**Background:** During arterial switch procedure for TGA coronary arteries are relocated from primary aortic sinus to neo-aortic sinus. The patency and proper route of transplanted arteries is important for myocardial perfusion and good postoperative effect.

The aim of this study was to assess coronary artery pathway variants and patency in children with TGA after switch procedure, estimate anatomical and demographic factors correlated with coronary anomalies and describe the norms of coronary arteries in children after ASO.

**Material and methods:** Between years 1992 and 2008 519 children had undergone switch procedure for ASO. From this group 136 patients were selected for this study. Inclusion criteria was over 5 years follow up with postoperative echocardiographic and angiographic examinations. Coronary route and patency as well as size of coronary arteries was assessed during routine coronarography. Diameter of coronary arteries to BSA ratio was evaluated in case of each artery. Anatomical details i.e. arterial discrepancy, non facing commissures, coronary anomalies, associated heart defects were analyzed using retrospective data.

**Results and conclusions:** Coronary artery anomalies are associated frequently with transposition of the great arteries (36%). In a majority of abnormal coronary routes Circumflex (Cx) coronary artery is a branch of the right coronary artery (16,7% of all anomalies). Coronary arteries to BSA ratio in whole group was shown in table. In our material this ratio was statistically significant different in case of coronary abnormal route as well as in case of VSD associated with TGA ( $p < 0,05$ ).

Coronary anomalies occurs more frequently in children with non facing arterial commissures ( $p < 0,05$ ), no other significant relations with analyzed risk factors were noted.

In 5 patients significant stenosis of a coronary artery was detected in a postoperative period. In 3 cases a coronary artery was narrow in its distal part which was considered a congenital anomaly.

	N	Mean	Norm range	Standard Deviation	
LAD/BSA	62	2,411	2,096	2,898	0,479
CX/BSA	62	2,058	1,868	2,525	0,400
RCA/BSA	62	2,800	2,438	3,374	0,659

#### P-78

##### Incidence of congenital heart disease in a series of out-patients referred to a tertiary level pediatric cardiac center

Kristo I. (1),(2), Lunardini A. (1), De Lucia V. (1), Recla S. (1), Ait Ali L. (1), Spadoni I. (1), Giusti S. (1), Biagini A. (1)  
(1) Pediatric Cardiology and GUCH Unit, "G. Monasterio" Tuscan Foundation, "G. Pasquinucci" Heart Hospital, Massa, Italy; (2) The Sant'Anna School of Advanced Studies, Pisa, Italy

**Objective:** To assess the incidence of congenital heart disease (CHD) in patients referred for the first time to the outpatient clinic of a tertiary care pediatric cardiac center.

**Methods:** The data-base of the outpatient clinic of our Department was retrospectively analysed, between November 2008 and November 2009, selecting 829 cases (439 males and 390 females) who were referred for the first time for evaluation by the Department of General Pediatrics and by community Pediatricians. All patients were evaluated with physical examination, electrocardiogram (ECG), and echocardiography.

**Results:** The mean age and weight at presentation were 4,4 years  $\pm$  4,7 (range 1 day–18 years) and 18,9  $\pm$  16,9 kg (range 1,5–80 kg), respectively. In 214 cases (25,8%), the reason for referral was a heart murmur. In 138 cases (16,6%), the referral was for suspected familiar recurrence or genetic syndrome, in 59 cases (7,1%) for symptoms of arrhythmias and in 58 cases (6,9%) for prematurity and/or problems during pregnancy. The other

360 patients (43,5%) were referred for signs and symptoms of suspected CHD. Among the 829 patients examined, 616 (74,3%) had a normal heart, 25 (3%) had a nonspecific cardiac anomalies (mild valvular regurgitation, Kawasaki disease, mild pericardial effusion, etc) and 7 patients (0,8%) had anatomic variants (dextrocardia, right aortic arch, persistent left superior vena cava and azygous continuation). In 181 patients (21,9%) a CHD was diagnosed. Simple CHD was detected in 165 of 829 cases (19,8%) and complex CHD in 16 of 829 (1,9%).

**Conclusion:** The diagnosis of CHD was limited to less than one quarter of this selected population, showing that most of the requested cardiac examinations with ECG and echocardiography were not appropriate. In 75% of cases, if pediatric clinical examination had been done correctly, it could have been possible to avoid ECG and Echocardiography examination. An accurate pediatric screening is mandatory to reduce the number of unnecessary sophisticated examinations, in order to limit unjustified waste of resources.

### P-79

#### Health- related quality of life in patients with congenital heart disease – comparison with healthy children and adolescents

*Toussaint-Goetz N., Vahsen N, Kaestner M., Hombach A., Kallenberg R., Zartner P., Wiebe W., Schneider M.*

*Department of Paediatric Cardiology, DKHZ Asklepios Klinik, Sankt Augustin, Germany*

**Objective:** The aim of the surgical, interventional, and medical treatment in patients with congenital heart disease is an improvement of cardiac function and quality of life. In this study the health- related quality of life in children and adolescents with congenital heart defects by self-assessment and external assessment has been recorded.

**Methods:** 61 patients aged from 8–16 years and their parents completed a health related quality of life questionnaire KINDL-Questionnaire (children –/youth version, parents version). The KINDL- Questionnaire replies the following six dimensions of quality of life: Physical well- being, emotional well- being, self-esteem, family, friends and everyday functioning. Furthermore a classification of the patients in groups (complex and non-complex heart disease) was done. The results were compared with reference values of healthy children and adolescents.

**Results:** Compared to the reference values of the interviewed children showed a significantly higher total score ( $p = 0.02$ ), which is due to significantly higher levels of physical well-being ( $p = 0.02$ ) and of self-esteem ( $p = 0.03$ ). Patients with complex congenital heart defects showed a descriptive, but not significantly lower total score and lower values for the dimensions of self-esteem, emotional well-being, family and friends. Parents reported significantly higher scores for their children than the children for themselves in the domain self-esteem and lower scores in the domain family.

**Conclusions:** Children and adolescents with congenital heart defects showed a normal to in some domains above- average health- related quality of life. But patients with complex congenital heart defects showed descriptively lower values

### P-80

#### Determining the Approaches to Facilitate the Patent Ductus Arteriosus Closure procedure with Amplatzer Duct Occluder Device in Adult Patients

*Pac F.A., Polat T.B., Oflaz M.B., Balli S., Kibar E., Ece I.*

*Clinic of Pediatric Cardiology, Turkiye Yuksek Ihtisas Education and Research Hospital, Ankara, Turkey*

**Objective:** The anatomic changes of the patent ductus arteriosus (PDA) in adult patients can complicate its transcatheter occlusion. The aim of the study was to determine procedural approaches to facilitate transcatheter closure of PDA in adult patients using the Amplatzer duct occluder (ADO).

**Methods:** Thirty-three adult patients with a moderate to large-sized PDA underwent transcatheter closure via the antegrade venous approach. Conventional antegrade approach was used in 14 patients, while retrograde wire-guided assisted approach in remaining 18 patients to access the PDA antegrade. If the initial procedure failed, snare-assisted technique was used to pass through the ductus antegradely. The ADO device used was at least 2 mm larger than the narrowest diameter of the PDA. Follow-up evaluations were performed at 24 h, 1, 3, 6 and 12 months after implantation.

**Results:** Thirty-two patients had successful PDA occlusion. The PDA measured from 2.6 to 5.2 mm (mean 3.4) at the narrowest diameter. PDA ostium could be engaged in 10 of 14 patients by conventional approach, while 17 of remaining 18 patients by retrograde wire-guided assisted approach ( $p = 0.07$ ). PDAs could be passed antegradely using the snare-assisted technique in those five patients in whom the initial procedure failed. The mean size of implanted devices was 3.76 mm larger than the narrowest size of the PDA. Spontaneous embolization occurred in one patient in whom smaller device implanted due to his short ductus. All patients attained complete occlusion and there were no complications encountered during follow-up.

**Conclusions:** The retrograde wire-guided technique is an effective approach and offers an alternative to access the PDA antegrade in adult patients. Bigger than recommended size ADO device can be used safely in all adult patients with feasible ductal ampulla and may prevent device migration.

### P-81

#### When is the right time for MRI in patients after TOF correction?

*Kantorova A. (1), Kaldararova M. (1), Neuschl V. (1), Nosal M. (1), Masura J. (1), Hraska V. (2)*

*National Institute of Cardiovascular Diseases – Children's Cardiac Centre, Bratislava, Slovakia (1); German Children's Cardiac Centre, Sankt Augustin, Germany (2)*

**Introduction:** Magnetic resonance imaging (MRI) is an objective and exact possibility for right ventricle (RV) volume measurement. On the other hand MRI is a relatively complicated, time-consuming and not always accessible method; so to estimate the reasonable timing for MRI may be very helpful. We analyzed the correlation between various echocardiographic (echo) measurements and MRI, with the aim to define the most optimal echo parameter indicating the necessity for further investigations.

**Methods:** During a long-term follow up in 50 patients after a complete correction Tetralogy of Fallot (TOF) detailed echo and MRI measurements were performed and correlation between both diagnostic methods was analyzed.

**Results:** All patients were  $>10$  years after surgical correction ( $13 \pm 3.1$  years). In echo 27 patients (54%) had severe pulmonary regurgitation and 7 (14%) had pulmonary stenosis above 30 mmHg. End-diastolic diameter (EDD)  $>30$  mm was measured in 13 patients and in 11 patients EDD was above 90% their normal value. Ejection fraction (EF)  $<40\%$  was present in 4 patients. MRI finding of end-diastolic volume (EDV)  $>160$  ml/m<sup>2</sup> was present in 10 patients and end-systolic volume (ESV)  $>85$  ml/m<sup>2</sup> in 15 patients.

A correlation between previously mentioned echo parameters and increased RV volumes measured by MRI was found.

**Conclusions:** MRI is a golden standard for indicating pulmonary valve implantation in TOF patients after correction. MRI though cannot always be carried out. We found correlation between echo and MRI parameters. Although echo is not optimal for establishing the exact RV volume, EDD >30 mm or >90% their normal values meet most probably the MRI criteria for reintervention; and so this may be the optimal timing for further examinations and exact RV volume estimation by MRI.

## P-82

### Psychosocial Adjustment and Proneness to Psychopathology in Adolescents and Young Adults with Congenital Heart Diseases (CHD)

Vieira D. (1), Proença C. (1), Areias M. E. G. (1), Silva A. M. (1), Vaz C. (1), Viana V. (1, 2), Moura C. (2), Areias J. C. (2)  
 Instituto Superior de Ciências da Saúde – Norte, Gandra – Paredes, Portugal (1); Hospital S. João, Faculdade de Medicina do Porto, Porto, Portugal (2)

**Objectives:** Several previous studies suggest that adolescents and young adults with CHD are more likely to show psychopathology and to have worse psychosocial adjustment. Therefore, in this study our aim was to assess psychosocial adjustment and proneness to psychopathology in our population of adolescents and young adults with CHD.

**Methods:** Forty patients with CHD, 22 male and 18 female, 23 with surgical repair and 17 without, aged 12-to-26 years old, were interviewed once with the Standardized Psychiatric Interview SADS-L (Schedule for Affective Disorders and Schizophrenia – Lifetime Version), and with a semi-standardized interview on different topics of self-image, social support, parental educational style, school performance, and functional limitations. They were also submitted to a self-report questionnaire on psychosocial adjustment (YSR, Young Self-Report and ASR, Adult Self-Report) and their caretakers filled an observational version of the same questionnaires.  
**Results:** The subjects of our study showed a slightly increased proneness to psychopathology (frequency of 15%) compared to the general population; cyanosis hasn't a relevant impact increasing the likelihood to have a psychiatric disorder or worsening psychosocial adjustment. Consistently with other studies, the severity of CHD ( $u = 109$ ;  $p = 0,02$ ) and the serious physical limitations ( $u = 110,5$ ;  $p = 0,05$ ) predicted a worse adjustment, and internalization problems, withdrawal, isolation. Patients submitted to surgical repair showed more internalization (isolation) problems ( $u = 85$ ;  $p = 0,008$ ), but the number of surgeries hadn't any additional impact over psychosocial adjustment. No other sign normally observed in patients with CHD was relevant for our results.

**Conclusions:** Our study emphasizes the importance of psychological survey of patients with CHD. On a preliminary basis, we found that CHD increases the likelihood to have a psychiatric disorder, suggesting also that a severe form of disease and many physical limitations can enhance this proneness and worse emotional adjustment such as withdrawal and isolation.

## P-83

### Medical attendance of adult patients with congenital heart disease.

Bauer U., Niggemeyer E. (1), Vigil M. (1), Paulick M. (1), Lux A. (2)  
 (1) Competence Network for Congenital Heart Defects, Berlin, Germany; (2) Otto-von-Guericke-Universität Magdeburg, Institut für Biometrie u. Medizinische Informatik, Magdeburg, Germany

**Background:** Although the number of adults with congenital heart defects is rising steadily, there is a lack of data concerning the medical attendance provided for this patient group.

**Hypothesis:** It cannot be assumed that adult patients with congenital heart disease are followed up by adult cardiologists exclusively.

**Methods:** Descriptive cross-sectional study including 1396 adult patients aged 18 or older from the German National Registry for congenital heart defects.

**Results:** The majority of the surveyed patients is followed up by cardiologists or paediatric cardiologists. Only 12.4% are not followed up continuously by physicians of relevant subspecialties. Of the patients with severe or moderate congenital heart defects, 66.5% are still followed up in the paediatric cardiological setting, with the degree of severity turning out to be a highly significant factor. Furthermore, these patients tend have been under the same physician since the time of first diagnosis. In contrast, those patients with mild congenital heart defects are rather followed up by adult cardiologists or non-cardiologic physicians (33.5%). In this context, a significant connection with the patients' age was also detected. However, this correlates with the fact that the older patients tend to be those with less severe congenital heart defects. Neither sex nor educational status nor stratum could be shown to be significant influencing factors on the type of medical attendance.

**Conclusion:** Patients with congenital heart disease tend to remain in paediatric cardiology care when entering adulthood. Generally, transition to adult cardiology does not take place. Further studies are needed to find out the reasons for this.

## P-84

### Testing a new disease specific quality of life measure for children and adolescents with heart disease in the United Kingdom (UK)

Wray J. (1,2), Franklin R. (2), Brown K. (1), Casale L. (2), Guy S. (1), Blyth J. (3), Cassedy A. (4), Marino B. (4)  
 Great Ormond Street Hospital for Children NHS Trust, London, UK (1); Royal Brompton and Harefield NHS Foundation Trust, London, UK (2); Birmingham Children's Hospital NHS Trust, Birmingham, UK (3); Cincinnati Children's Hospital Medical Center, Cincinnati, US (4)

**Objectives:** With the continued successful treatment of children/adolescents with congenital (CHD) or acquired heart disease, quality of life (QoL) outcomes assessment is needed, rather than simple mortality. The Pediatric Cardiac Quality of Life Inventory (PCQLI) is a new self administered disease specific QoL measure, comprising both patient and parent proxy reporting and recently shown to be reliable and valid in the United States. The purpose of this study was to assess reliability and validity of PCQLI scores in the UK.

**Methods:** Following translation of the PCQLI into "British English", 3 UK tertiary paediatric cardiac centres recruited patients aged 8 to 18 years with heart disease. Patient/parent pairs completed the PCQLI, a generic QoL measure (PedsQL4.0) and non QoL instruments [Self Perception Profile for Children/Adolescents (SPPC/A); Achenbach Youth Self Report Form/Child Behavior Checklist]. Test/retest reliability was performed by administering the PCQLI at two time points and assessed by correlation coefficients. Validity was assessed by correlation of PCQLI scores between patients and parents and with severity of CHD, medical care utilisation, and PedsQL, SPPC/A and Achenbach scores.

**Results:** This preliminary study was completed by 125 child/parent pairs and 125 adolescent/parent pairs. 54% of patient respondents were male and 84% of parent respondents were female. Diagnostic categories were biventricular repair (40%), Fontan palliation (20%), unrepaired CHD (20%), heart transplantation (2.8%) and acquired heart disease (17.2%). Correlations evaluating test/retest reliability ranged from 0.64 to 0.90. Correlations for patient and parent scores were moderate for both child and adolescent groups (0.49 to 0.72). Lower PCQLI scores were associated with Fontan palliation ( $p < .001$ ) and increasing number of hospitalisations ( $p < .005$ ) and cardiac catheterisations ( $p < .001$ ). Correlations for the PCQLI and generic QoL measure were moderately high (0.66 to 0.77). Lower PCQLI total scores were associated with lower global self worth on the SPPC/A ( $p < .001$ ) and lower total competency scores on the Achenbach measures ( $p < .001$ ).

**Conclusions:** Preliminary analysis of PCQLI data collected in the UK suggests it is a reliable and valid disease specific QoL measure for children/adolescents with heart disease. This novel tool allows for outcomes assessment in the paediatric cardiac population, with further prospective testing in progress.

#### P-85

##### **Aortic valve-sparing operations for ascending aortic aneurysmal disease: promising midterm results in young patients with congenital cardiovascular pathology.**

Francois K., Bove T., Van Belleghem Y., De Backer J., De Wolf D., Suys B., Van Nooten G.

Cardiac Centre, University Hospital Gent, Belgium

**Objectives:** Since the introduction of valve-sparing operations for ascending aortic aneurysms in the 1990s, these techniques have become more widespread and the applications have been expanded. We reviewed our 10-year experience with aortic valve-sparing operations in a variety of congenital cardiovascular pathology.

**Methods:** Between 2000 and 2009, 30 patients with ascending aortic aneurysmal dilatation and underlying congenital pathology (21 Marfan, 2 Loeys-Dietz, 5 bicuspid aortic valves, 2 other) underwent an aortic valve-sparing operation. Mean age was  $36 \pm 13.6$  years (range 15.4–58.2 years), mean sinus of Valsalva diameter was  $54 \pm 7$  mm (range 43–81 mm), mean aortic valve incompetence was  $1.4 \pm 1.1$  (range 0–4). Two patients presented with acute aortic dissection, 2 were reoperations after previous corrective cardiac surgery during childhood. A David reimplantation procedure was performed in the majority of patients (27/30; 90%) to reduce the dilated aortic annulus and stabilize the valve repair.

**Results:** No early or late deaths occurred. Postoperative morbidity was limited (2 revisions for bleeding, 1 pericardial drainage, 1 pacemaker insertion). Aortic incompetence at discharge was grade 0 to 1 in 27 patients (90%). Follow-up was complete for a mean of  $4.4 \pm 2.9$  years. One Marfan patient with a bicuspid valve required insertion of a mechanical aortic valve prosthesis 20 months after David repair for increasing aortic incompetence. One patient underwent mitral valve repair 5 years after the David procedure. There were no thromboembolic events nor reoperations on the residual aorta. Aortic regurgitation at latest follow-up was none in 7, a trace in 5, grade 1 in 13 and grade 2 in 4 patients.

**Conclusions:** Aortic valve reimplantation for aortic root aneurysmal dilatation with or without aortic valve incompetence in young patients with tissue disorders and other congenital cardiovascular pathology proves to offer a stable repair and a good quality of life. The midterm results are promising and permit extending the indications in a young patient cohort, avoiding the need for life-long anticoagulation therapy.

#### P-86

##### **Prevalence of reflex, vaso-vagal syncope in young adults long term follow-up correction of d-transposition of great arteries by Senning atrial switch.**

Pietrucha A. Z(1), Pietrucha B.J. (2), Czubek U. (1), Mroczek-Czernecka D. (1), Węgrzynowska M. (1), Piwowarska W. (1), Nessler J. (1)

(1) Coronary Disease Department, Medical School of Jagiellonian University John Paul II Hospital, Cracow, Poland; (2) Children Cardiology Department, Children University Hospital, Medical School of Jagiellonian University, Cracow, Poland.

**The aim of study:** evaluation of prevalence of reflex reaction on orthostatic stress in patients with benign forms of sinus node dysfunction in near-asymptomatic young adults after reparation of d-transposition of great arteries d-TGA by Senning atrial switch (d-TGA-S).

**Study population:** we observed 16 pts (13 men) aged 18–21 yrs with d-TGA-S, with electrocardiographic signs of sinus node dysfunction (SND) and/or mild bradyarrhythmia-related symptoms.

**Methods:** All pts underwent head-up tilt test (HUTT) according to Westminster protocol. Transoesophageal atrial stimulation (RAS) for evaluation of total (SNRT) and corrected (CNRT) sinus node recovery times before and after pharmacological blockade (PHB) with iv propranolol and atropine administration was also made in all pts. SNRT > 1500 ms and CNRT > 525 ms were assumed as abnormal.

**Results:** HUTT was positive in 4 pts (25%): one cardiodepressive with max RR pause 6,7s and three mixed with hypotonia and significant bradycardia as a response to the orthostatic test. The following 7 pts had negative results of HUTT, whereas in 5 pts results of the test was doubtful, mild bradycardia and/or hypotonia occurred, but without complete loss of consciousness (only prodroms).

Mean rest HR value was 62,8 bpm, IHR 77,8 bpm. Before PHB mean values of SNRT was 1569,3 ms and CNRT- 698,2 ms. After PHB SNRT shortened to 1108,2 ms and CNRT to 362,8. Prolongation of SNRT < 1000 ms and/or CNRT < 850 ms) with normalization after PHB (functional SND) was observed in 8 pts (53,3%). Rest of patients presented normal SNRT/CNRT values.

Positive or doubtful result of HUTT significantly corresponds to symptoms previously described as bradycardia related. It may suggest that bradyarrhythmias, specially symptomatic, may be due reflex usually leading to vaso-vagal syncope, than primary sinus node dysfunction.

**Conclusions:**

1. Reflex vaso-vagal syncope relatively frequently occurs in patients after physiological correction of d-TDGA by atrial switch.
2. Electrophysiological parameters of sinus usually be in the normal range in spite of electrocardiographic signs of sinus node dysfunction in young adults with d-transposition of great arteries corrected by Senning atrial switch.
3. Observed in this group of patients bradyarrhythmias, specially symptomatic, may be due reflex usually leading to vaso-vagal syncope, than primary sinus node dysfunction.

#### P-87

##### **Early changes in Right Ventricular Function and their Clinical Consequences in Childhood Dilated Cardiomyopathy**

Grosse-Wörtmann L., Roche S.L., Yoo S.-J., Seed M., Kantor P.  
The Hospital for Sick Children, Toronto, Canada

**Introduction:** There is increasing evidence that the right ventricle (RV) plays a vital role in the clinical status in patients with dilated cardiomyopathy (DC) and their survival. We aimed to 1.) determine whether abnormalities in RV function are also seen by magnetic resonance imaging (MRI) in children with this disease and 2.) investigate whether any alterations in RV size and function are clinically important.

**Methods:** We measured biventricular size and function in eleven patients with dilated cardiomyopathy as well as in twelve normal paediatric controls. Late gadolinium enhancement images and phase contrast flow velocity mapping across both atrioventricular valves were obtained. Blood for N-terminal pro-brain natriuretic peptide (NT-pro-BNP) was collected at the time of MRI, and the results from the most recent echocardiogram and exercise test were reviewed.

**Results:** Patients with DC had significantly faster heart rates (85 versus 65 bpm) to maintain a normal cardiac output. They had lower LV ejection fractions (EF) (42 versus 61%), RV EF (44 versus 54%) and lower LV and RV stroke volumes (35.5 versus 49.5 ml/m<sup>2</sup> and 40.9 versus 56.4 ml/m<sup>2</sup> respectively). Relative to normal controls, RV EF was more severely reduced than LV EF (30.3 versus 18.7%). Lower RV EF and higher RV volumes were associated with NT-pro-BNP ( $r = -0.67$ ,  $p = 0.03$ , and  $r = 0.65$ ,  $p = 0.04$ , respectively), RV EF correlated with the anaerobic threshold ( $r = 0.67$ ,  $p = 0.049$ ). In contrast, neither LV EF nor LV end-diastolic volume correlated with NT-pro-BNP or exercise tolerance. Patients had lower mitral and tricuspid valve inflow e/a wave velocity ratios than controls (2.02 versus 2.80 and 1.25 versus 2.58, respectively). E/a wave ratios by MRI were not different from those by echocardiography. Tricuspid valve annulus velocity by tissue doppler echocardiography, correlated with RV EF by MRI ( $r = 0.60$ ,  $p = 0.05$ ). LV diastolic dysfunction, as shown by a lower MV e/a ratio correlated with lower LV EF ( $r = 0.90$ ,  $p = 0.0062$ ), both by MRI.

**Conclusions:** Right ventricular systolic function is decreased in the early stages of DC and may, in fact, be more severely affected than the LV. Right ventricular size and EF may be important indicators of subclinical heart failure and should be monitored by MRI.

#### P-88

##### Utility of Whole-Blood Aggregation for Evaluation of Anti-Platelet Therapy in Chronic Stage of Kawasaki Disease

Suzuki C., Yahata T., Hamaoka A., Fujii M., Ozawa S., Hamaoka K.

Department of Pediatric Cardiology and Nephrology, Graduate School of Medical Science, Kyoto Prefectural University of Medicine, Kyoto, Japan

**Introduction:** We think that part of patients in the chronic stage of Kawasaki disease (KD) may exhibit aspirin resistance; therefore, it is necessary to evaluate the efficacy of anti-platelet agents by monitoring systems. Platelet function is conventionally determined by measuring platelet-rich plasma (PRP) aggregation. However, the conditions for the preparation of PRP have not yet been standardized, and it requires skill to prepare PRP during clinical practice. Additionally, only platelet aggregation can be measured with the PRP methods; however other blood cells and prostaglandins are also involved in thrombus formation. Whole-blood analysis was recently established. Therefore, we compared whole-blood aggregation with PRP aggregations.

**Methods:** The subjects were 24 chronic KD patients. 13 patients underwent anti-platelet therapy mainly with aspirin for coronary artery lesions (CALs), and the remaining did not undergo the

medication because they did not exhibit the CALs. Whole-blood aggregation was analyzed using collagen as the stimulus and was compared with the PRP aggregation measured with an optical aggregometer. Whole-blood aggregation was evaluated on the basis of the platelet-aggregation threshold index (PATI), which was defined as the putative agonist-concentration giving half-maximal-aggregation. PRP aggregation was categorized into 5 classes; -2, -1, 0, +1, and +2.

**Results:** The whole-blood PATI was significantly decreased in subjects who did not undergo therapy when compared with that of all the subjects in the anti-platelet therapy, except for one non-compliance patient (no therapy,  $0.35 \pm 0.20$ ; anti-platelet therapy,  $0.83 \pm 0.28$ ). PRP aggregation in no therapy group was of class  $\pm 0$  to +2, and that in anti-platelet therapy group was of class -2 to  $\pm 0$ . In the non-compliance patient, whole-blood and PRP aggregations were similar to those of the other patients in the no-therapy group. (PATI, 0.19; class, +1). In addition, we found a negative correlation between PATI and the PRP aggregation class.

**Conclusions:** This study revealed that whole-blood aggregation contributed to the efficacy of aspirin in patients in the chronic stage of KD. Furthermore, the whole-blood analysis is easier because preparation of PRP is not required; therefore, it is highly useful for monitoring the effectiveness of anti-platelet drugs during clinical practice.

#### P-89

##### Complete anatomical and physiological assessment of partial anomalous pulmonary venous connection with MRI

Grosse-Wörtmann L., Sellar N., Grant B., Yoo S  
The Hospital for Sick Children, Toronto, Canada

**Introduction:** Diagnosis of partial anomalous pulmonary venous connection (PAPVC) is usually made by echocardiography. However, complete anatomical delineation can be challenging. Magnetic resonance imaging (MRI) can provide a comprehensive assessment of patients with PAPVC. Controversy regarding the patient selection for surgical intervention exists. In particular, the effect of the location of the anomalous connection on the amount of blood flow via that vein is unclear.



**Methods:** We reviewed the MRI studies performed on patients with PAPVC in our institution over the last 5 years. Patients with Scimitar syndrome were excluded. Patients were assessed with cine imaging, contrast-enhanced angiography, and phase-contrast

velocity mapping. The number of anomalous veins was recorded, and the proportion of anomalously draining lung was estimated by relating the number of abnormally draining pulmonary veins to the total number of ipsilateral veins. The magnitude of the systemic-to-pulmonary shunt (Qp:Qs), and ventricular volumes were measured. *Results:* Nineteen patients (13 male, mean age  $11.1 \pm 5.8$  years) with an MRI diagnosis of PAPVC. In four patients, PAPVC had not been detected by echocardiography. In a further three cases, an echocardiographic suspicion was confirmed. Twelve patients had right-sided PAPVC of the upper  $\pm$  middle pulmonary vein (Fig. 1), six had PAPVC of the left upper pulmonary vein, and one patient had non-Scimitar right-sided unilateral total anomalous pulmonary venous drainage. In 8 patients, there was an associated sinus venosus atrial septal defect (SVASD). Mean Qp:Qs was  $1.75 \pm 0.4$ , and increased to  $2.5 \pm 0.6$  in the presence of a SVASD. Qp:Qs correlated with right ventricular end diastolic volume index ( $\rho = 0.54$ ;  $p = 0.02$ ) and the proportion of anomalous to normally draining veins ( $\rho = 0.77$ ;  $p = 0.01$ ). We found no increased differential pulmonary arterial blood flow to lungs with PAPVC as compared to normal controls.

*Conclusions:* For patients with PAPVC, MRI provides a complete anatomical and functional assessment, which may provide important additional information in surgical decision making.

### P-90

#### Quantification of left to right shunt through patent ductus arteriosus by color Doppler—Clinical and experimental studies.

Harling S. (1), Jansson T. (2), Pesonen E. (3)  
Department of Paediatrics, Halmstad Hospital, Halmstad, Sweden (1); Department of Electrical Measurements, Lund University, Lund, Sweden (2); Department of Paediatrics, Section of Paediatric Cardiology, Lund University Hospital, Lund, Sweden (3)

*Background:* The increased number of surviving extremely low birth weight (ELBW) infant has led to a higher number of babies requiring medical or surgical intervention for patent ductus arteriosus. There is no consensus as to optimal management strategy in this high-risk and vulnerable population.

*Methods:* We have developed a noninvasive method to quantify the ductal shunt by color Doppler echocardiography which could serve as a guideline in the treatment decision. An image is obtained over the main pulmonary artery longitudinal cross section (PALS). A presentative frame image during maximal color intensity in diastole is selected and analysis in a custom-designed program written in MAT LAB® (The Math Works, Natick, MA). The program separates the pixels in PALS into three matrices, each containing one of the three base colors established from the color bar in the image. Measurements are derived from the color information we have collected from both an animal study and from children admitted to our unit for device closure of PDA. We looked at the numbers of pixels that indicated flow towards or away from the transducer, or turbulence, each color added together and divided with the area in PALS, expressed as total percentage of color pixels.

*Results:* We found in our experimental study in lambs (sensitivity of 80% and specificity of 100%  $n = 24$ ) as well as in our clinical study, made retrospectively (sensitivity of 92% and specificity of 71%  $n = 20$ ) a significant correlation between percentage of green pixels in PALS and the ratio of pulmonary to systemic flow. When the Qp/Qs was  $>1.5:1$  the percentages green pixels in PALS, representing ductal flow, was  $>50\%$ .

*Conclusions:* The target group for our developed method is extremely low birth weight infants. A noninvasive method to

evaluate the size of ductal shunt would be of great value for detection of infants that need treatment. This could reduce the morbidity and complications of treatments. Interventional closure of ductus arteriosus could be limited to infants and children with shunt sizes  $>1, 5:1$ .

### P-91

#### Tissue Doppler Imaging study in long-term follow-up of patients who underwent surgical mitral valvuloplasty in pediatric age

Milanesi O. (1), Varotto E. (1), Biffanti R. (1), Maschietto N. (1), Padalino M. (2), Vida V. (2), Stellin G. (2)  
Pediatric Cardiac Unit (1) and Pediatric Cardiac Surgery Unit (2), Department of Paediatrics, University of Padova. Italy

*Objectives:* Surgical valvuloplasty is the preferred treatment for congenital mitral anomalies in paediatric age. This study evaluates the functional result of such surgical procedure using 2D-Doppler and Tissue-Doppler Echocardiography (TDE).

*Methods:* Thirty patients affected by MV stenosis (MS, 12) or insufficiency (MI, 18) who had surgical valvuloplasty at a mean age of 4,9 years (range 0,2–18,7) underwent echocardiographic evaluation at a mean follow-up of 10,4 years (range 1,9–20,2). Patients with A-V canal defects, hypoplastic left heart syndrome, anomalies of the A-V, or V-A junction, were excluded. We recorded the following parameters, assessed before surgery and at the follow-up. For prevalent MI: LV diastolic diameter, FS, Pisa index, systolic RV pressure, medical therapy, heart failure. For prevalent MS: LA dilatation, FS, transmitral inflow peak velocity, RV pressure, medical therapy, heart failure. We produced a severity score system (SS, higher number = more severe condition) to make clinical and hemodynamic data comparable. TDE evaluation, performed only at the follow-up, considered the following parameters: peak E1, peak A1, E1/A1, E/E1, peak S'1, peak S1, ICA1. TDE data were compared to those obtained from a group of 22 healthy subjects, sex-, age- and body surface area-related to our patients.

Global population's TDE data trend compared to healthy group values

	<25th percentile		25th–75th percentile		>75th percentile	
	n	%	n	%	n	%
LV diastolic function						
E1	24	80.0	5	16.7	1	3.3
A1	16	53.3	8	26.7	6	20.0
E/E1	18	60.0	5	16.7	7	23.3
LV systolic function						
S'1	15	50.0	8	26.7	7	23.3
S1	27	90.0	2	6.7	1	3.3
ICA1	15	50.0	13	43.3	2	6.7
RV diastolic function						
E1	23	88.5	3	11.5	0	0.0
A1	20	76.9	6	23.1	0	0.0
RV systolic function						
S'1	20	76.9	5	19.2	1	3.8
S1	25	96.1	1	3.8	0	0.0
ICA1	22	84.6	4	15.4	0	0.0

LV: left ventricle.

RV: right ventricle

**Results:** SS decreased after surgery in 23 patients (83%), was stable in 3 (10%) and increased in 2 (6, 7%). SS was not related to the length of follow-up. Thirteen patient with MI (72%) lowered their SS, 3 (16,7%) remained stable, and 2 (11%) worsened. All the MS patients had higher SS before surgery than those with MI. They all improved and 4 became asymptomatic. TDE study showed a reduction of systolic and diastolic function of both ventricles (table). We observed a direct correlation between LV TDE abnormalities, but not RV, and SS. LV dysfunction, detected by TDE, was more frequent in MS group, and at mid-term follow-up.

**Conclusion:** Functional result of mitral valvuloplasty in paediatric patients at a long follow-up is satisfying. However TDE shows persistent systolic and diastolic impairment of the LV function, more evident in shorter follow-up patients. We could infer that the ventricular mass requires longer time to adapt to the new loading condition.

#### P-92

##### **Changes of natriuretic peptide type B levels after exercise in children with congenital heart defects compared with healthy children**

Holmgren D. (1), Westerlind A. (2), Lundberg P.A. (3), Wähländer H. (4)

The Departments of Paediatrics, Skövde Hospital (1); Cardiothoracic Surgery (2); Clinical Chemistry (3); Sahlgrenska University Hospital and The Queen Silvia Children's Hospital (4); Göteborg, Sweden

The natriuretic peptide type B (BNP) is synthesized mainly in the ventricles of the heart and released into the circulation in response to increased stretch of myocytes. In the circulation, BNP act as reliever of volume and pressure overload by inducing diuresis, natriuresis and vasodilation. The activation of BNP may precede clinical symptoms caused by the overload. One may speculate that additional strain produced by exercise will unmask a possible pre-clinical overload of the heart.

**Aim:** To compare changes of BNP levels after exercise in children with congenital heart defects (CHD) with right or left ventricular overload with healthy children.

**Methods:** Plasma (P)-BNP was analysed before, immediately after and 2 hours after exercise (bicycle ergometer) in 20 patients, aged median 13.1 years, (range 7.7–18.9) with CHD of whom 13 had left ventricular (LV) overload (aortic stenosis/regurgitation) and 7 right ventricular (RV) overload (conduit stenosis/regurgitation). Nine healthy children, 5 girls and 4 boys, with a median age of 13.8 years, (12.7–14.1) served as controls. (Reference values for P-BNP was, 0–18.4 ng/L).

**Results:** In children with CHD, P-BNP increased from median 31.9 ng/L (5.3–304.6) to 44.7 (8.6–523.5) immediately after exercise and decreased to 24.2 (3.7–280.1) 2 hours after exercise ( $p < 0.0001$  and  $p < 0.0001$ , respectively). The same pattern was observed in the controls, 14.3 ng/L (2.6–38.7) at baseline, 26.5 (5.8–66.7) immediately after and 9.1 (2.6–57.6), 2 hours after exercise ( $p = 0.004$  and  $p > 0.99$ , respectively). The changes in P-BNP immediately after exercise correlated with maximal workload in children with CHD ( $p = 0.01$ ). The changes in P-BNP after exercise were the same in the LV and RV groups, respectively. The pressure gradient were the same in the LV and RV groups, 42 mmHg (13–96) and 67 (13–80) respectively ( $p = 0.80$ ). Maximal workload reached approximately the same level in the RV, LV and control groups, 2.6 W/Kg (2.0–3.0) 3.1 (2.2–4.1) and 3.0 (2.3–3.8), respectively ( $P = 0.09$ ).

**Conclusion:** P-BNP increases immediately after exercise and returns to baseline 2 hours after exercise in both healthy children and those with CHD. The magnitude of changes (e.g. increase) of BNP after exercise was the same in those with and without cardiac overload.

#### P-93

##### **Assessment of ventricular function and dyssynchrony before and after stage II operation for hypoplastic left heart syndrome using 2D speckle tracking**

Petko C., Hoffmann U., Möller P., Kramer H.-H., Uebing A.

Klinik für Kinderkardiologie, Universitätsklinikum Schleswig-Holstein, Kiel, Germany

**Background:** 2D speckle tracking is a new echocardiographic technique that is both geometry and angle independent and yields data on global and regional myocardial function. Using this technique, we investigated differences in global and regional systemic right ventricular (RV) function as well as intraventricular dyssynchrony in hypoplastic left heart syndrome (HLHS) patients before and after stage II operation.

**Methods:** We enrolled 31 patients with HLHS who underwent stage II operation (28 Hemi Fontan, 3 bidirectional Glenn) at our institution between 9/2007 and 9/2009. From apical 4-chamber views of echocardiograms taken before (day of admission) and after (day of discharge) surgery, we assessed changes in global strain (S) and strain rate (SR) as well as regional peak systolic longitudinal S, SR and velocity (V) in 6 RV segments (basal septal, mid septal, apical septal, apical lateral, mid lateral and basal lateral) using 2D speckle tracking. Additionally, we investigated changes in the time intervals from the beginning of electrical activation to each peak event (S, SR and V) and assessed intraventricular dyssynchrony by calculating the 3 standard deviations of the 6 segmental times to each peak event before and after surgery.

**Results:** Global S ( $-16.7 \pm 5.0$  vs.  $-15.6 \pm 5.5\%$ ) and global SR ( $-1.2 \pm 0.3$  vs.  $-1.2 \pm 0.3$  s<sup>-1</sup>) were not different before and after operation. Peak systolic longitudinal velocities decreased in mid lateral ( $p = 0.01$ ) and basal lateral ( $p = 0.001$ ) segments while peak longitudinal systolic strain was lower in the basal lateral segment ( $p = 0.005$ ) after surgery. Intraventricular dyssynchrony was prevalent at both times but did not change after surgery. The time to peak systolic velocity was prolonged after surgery in the basal lateral segment ( $p = 0.04$ ) while time to peak strain rate was prolonged in the mid lateral segment ( $p = 0.03$ ).

**Conclusions:** After stage II operation myocardial velocities and strain were decreased in the RV free wall of HLHS patients. The same region showed prolongation of the times from electrical activation to peak V and to peak SR. Longitudinal follow-up is necessary to assess whether this impairment of regional myocardial function is an early postoperative phenomenon or permanent.

#### P-94

##### **Increased left atrial performance in asymptomatic patients and ventricular analysis of diastolic function in children after ToF-Repair**

Koenigstein K., Hurst T., Hosse M., Hauser M., Abdul-Khaliq, H.

Clinic for Paediatric Cardiology, Saarland University Hospital, Homburg/Saar, Germany

**Aim:** The aim of our prospective study was to evaluate left atrial and left ventricular diastolic function as well as the interaction between both in asymptomatic children and adolescent after ToF-Repair.

**Methods:** 25 children with TOF-repair and age-matched 30 children and young adults of normal health were studied using non-invasive conventional echo and 2D-speckle-Tracking parameters. Atrial function was assessed by registration of inflow-patterns in right pulmonary veins during ventricular systole (S), diastole (D) and atrial contraction (A). Global atrial performance was evaluated by measurement of LAEF, regional systolic and diastolic myocardial performance in left atrium and

ventricle (Strain S and G, Strain Rate S, E and A) were analyzed using 2D speckle tracking (Vivid VII, EchoPac GE). Furthermore we collected data about the haemodynamic situation of the left heart during ventricular systole (volumetric LVEF, Tei-Index, E/E') and diastole (LAEF, ventricular enddiastolic volume, atrial volume). To verify atrial timing and possible electromechanical atrial delay we assessed several time intervals in the atrium (PA-time, QD-time, QA-time, S-time, D-time, SDA-time). Statistical analysis included Mann-Whitney-U-Test and two-tailed Spearman-Correlation.

**Results:** Systolic inflow of the pulmonary veins did not change in ToF-patients compared to the controls. Diastolic LA-inflow patterns (D, A) were significantly higher ( $p < 0,001$ ), ventricular EF appeared to be slightly lower ( $p = 0,09$ ) and Tei-Index significantly increased ( $p = 0.006$ ). E/E', which is closely related to LVEDP, was significantly higher in ToF-patients ( $p < 0,001$ ) and correlated with the diastolic pulmonary venous parameters ( $p < 0,04$ ). The myocardial parameters such as Strain and Strain Rate did not show significant differences between the ToF-patients and controls. The time intervals to the active atrial contraction parameters such as time to peak of Strain Rate A, time to peak atrial wave in pulmonary veins were shortened ( $0,007 < p < 0,075$ ,  $p < 0,049$ ). VTI of the pulmonary venous A wave was significantly increased indicating higher LA contractile performance in patients in contrast to controls.

**Conclusions:** In asymptomatic young patients after ToF-Repair increased atrial performance indicates adaptive compensatory mechanisms to overcome altered systolic and diastolic ventricular function. Extensive assessment of left atrial parameters should be considered in terms of an entire evaluation of left heart performance in patients after ToF-repair.

#### P-95

##### **Early detection of anthracycline cardiotoxicity in children with cancer using plasma levels of NT proBNP and pulsed-tissue Doppler echocardiography.**

Atalay S., Uçar T., Tutar E., Uysal Z., Yavuz G.  
Ankara University Faculty of Medicine, Ankara, Turkey

**Objectives:** To evaluate left and right ventricular myocardial performance after anthracycline treatment using pulsed-tissue Doppler imaging (TDI) and its relation to NT proBNP levels in children with cancer.

**Methods:** We enrolled 60 children with cancer with normal left ventricular systolic and diastolic functions by conventional echocardiography and 30 healthy control subjects ( $11.9 \pm 3.1$  years). Thirty patients (Group 1,  $9.0 \pm 3.4$  years) were eligible for the analysis of subacute and 30 (Group 2,  $14 \pm 3.0$  years) of late cardiotoxicity. The mean follow-up duration from end of anthracycline treatment were  $4.2 \pm 2.5$  months in group 1 and  $69.8 \pm 39.7$  months in group 2. The cumulative dose of anthracyclines were  $319.2 \pm 196$  mg/m<sup>2</sup> in group 1 and  $275.6 \pm 149.6$  mg/m<sup>2</sup> in group 2. Myocardial performance indexes (MPI) of mitral lateral annulus (MLA), interventricular septum (IVS) and tricuspid lateral annulus (TLA) were calculated with TDI. Plasma NT proBNP levels were measured in all patients and control group. MPIs, other echocardiographic parameters, and plasma NT proBNP levels of patients were compared with control group.

**Results:** MPI of MLA, IVS and TLA of patients in group 1 were significantly higher than control group ( $p < 0.01$  and  $p < 0.001$ , and  $p < 0.001$ , respectively). NT proBNP levels of patients in group 1 were significantly higher than control group ( $p < 0.05$ ). The cumulative dose of anthracyclines of patients in group 1 were significantly correlated with MPI of MLA, IVS and TLA.

MPIs of patients in group 2 were significantly higher than control group ( $p < 0.001$  and  $p < 0.001$ , and  $p < 0.001$ , respectively). The cumulative dose of anthracyclines of patients in group 2 were significantly correlated with MPIs. The follow-up duration from end of anthracycline treatment in group 2 were significantly correlated with MPIs. NT proBNP levels of patients in group 2 were not different from control group.

**Conclusion:** Our study confirms that MPI obtained by TDI seems to be an early sensitive parameter of cardiac dysfunction in both subacute and late anthracycline cardiotoxicity. NT proBNP levels seems to be an early sensitive marker of cardiac dysfunction only subacute cardiotoxicity.

#### P-96

##### **The use of advanced imaging modalities in the management of patients with congenital heart disease – a European multicenter survey**

Valsangiaco Buechel E.R. (1), Hoesli D. (1), Mertens L. (2), Helbing W.A. (3), Simpson J.M. (4), Sieverding L. (5), Milanesi O. (6)  
(1) University Children's Hospital Zurich, Switzerland; (2) Hospital for Sick Children, Toronto Canada; (3) Sophia Children's Hospital, Rotterdam The Netherlands; (4) Evelina Children's Hospital, London UK; (5) Universitätsklinikum, Tübingen Germany; (6) Cardiac Unit, University of Padua, Italy

**Background:** During the last decade different imaging techniques have been introduced for diagnosis and management of congenital heart disease (CHD). We wanted to assess how the different imaging modalities are currently used in the evaluation of the most common forms of CHD.

**Methods:** Multicentric, web-based survey addressed to 153 tertiary paediatric cardiology centres in 30 European countries. In the questionnaire the most common CHD were classified in groups with similar pre- and postoperative conditions. For each group the centres were asked which imaging modalities were used beyond standard transthoracic echocardiography. These included cardiac catheterisation (cath), all advanced echocardiographic (echo) techniques and cross-sectional imaging, such as cardiovascular magnetic resonance (CMR) and computer tomography (CT).

**Results:** 67 centres from 20 different countries participated in the survey. For all different groups of CHD the most frequently used additional imaging technique was cath in  $57 \pm 24\%$  of the cases, followed by CMR in  $38 \pm 14\%$  and transoesophageal echocardiography (TEE) in  $20 \pm 7\%$ . Cath was most frequently used (% always/sometimes) for evaluation of univentricular hearts before cavopulmonary connection (84/14) or the Fontan procedure (84/16), for assessment of pulmonary hypertension (87/13) and of pulmonary atresia with multiple aortopulmonary collaterals (89/11). CMR was mainly utilized for evaluation of aortic arch anomalies and vascular rings (59/36), after repair of tetralogy of Fallot in presence of pulmonary regurgitation (80/18) or RVOT obstruction (50/40) and after Fontan operation in case of failing haemodynamics (37/58). The most common indications for performing TEE were anomalies of the aortic valve (34/52) and LVOT obstruction (27/65), failing Fontan (29/58), shunt defects (24/65) and patients after the Senning operation (17/75).

**Conclusions:** In Europe, cardiac catheterisation is still the most commonly used additional imaging technique, however mainly for specific indications, such as univentricular CHD. Besides cath, CMR is the most frequently utilized emerging modality, mainly for evaluation of the intrathoracic arteries and of selected haemodynamic parameters. TEE is the most commonly used additional echocardiographic technique.



## P-97

### Added value of Real-time Three-dimensional Trans-thoracic Echocardiography in the Assessment of Congenitally Malformed Aortic valve

Al-Barakati M, Anwar A, MNosir Y, F Galal AN, Chamsi-Pasha H  
Cardiology dep. King Fahad Armed Forces Hospital, Jeddah, Saudi Arabia

**Objectives:** To study the feasibility and additional value of real-time three-dimensional transthoracic echocardiography (RT3D-TTE) for evaluation of patients with malformed aortic valve (AV) compared to conventional two-dimensional TTE (2D-TTE).

**Methods:** Twenty five patients (mean age  $18 \pm 9.5$  years, 70% male) were evaluated for malformed AV by both 2D-TTE and RT3D-TTE. Visualization of aortic cusps (number, direction and commissures) was evaluated by a 3-point score (1: non-visualized, 2: inadequate, 3: adequate). Measurements obtained by both 2D-TTE and RT3D-TTE included AV area by planimetry and diameters of aortic annulus, left ventricular outflow tract (LVOT), and aorta at multiple levels (root, ascending, arch and descending).

**Results:** The mean and median visualization score obtained by RT3D-TTE were higher than by 2D-TTE (Table). Malformed AV was bicuspid in 19 patients, quadricuspid in 2 patients, tricuspid in 3 patients and unicuspid in 1 patient. Associated lesions were ventricular septal defects in 3 patients, coarctation in 2 patients and supra-ventricular stenosis in 1 patient and subvalvular stenosis in 1 patient. RT3D-TTE and 2D-TTE measurements of aortic annulus, LVOT and aorta at multiple levels were well correlated ( $r = 0.85$ ;  $p < 0.001$ ) but the RT3D-TTE measurements of aortic annulus and LVOT diameters were significantly larger than that obtained by 2D-TTE ( $2.05 \pm 0.7$  cm and  $2.5 \pm 0.86$  vs.  $1.94 \pm 0.67$  and  $1.98 \pm 0.74$ ;  $p < 0.01$ ). Measurement of AV area was obtained in 76% and 44% of patients by RT3D-TTE and 2D-TTE respectively.

**Conclusion:** RT3D-TTE is a feasible technique that allows comprehensive quantitative and qualitative assessment of malformed AV.

Score	Number of cusps		Cusp direction	
	RT3D-TTE	2D-TTE	RT3D-TTE	2D-TTE
Adequate (3)	22 (88%)	16 (64%)	22 (88%)	16 (64%)
Inadequate (2)	2 (8%)	4 (16%)	2 (8%)	4 (16%)
Not visualized (1)	1 (4%)	5 (20%)	1 (4%)	5 (20%)
Median score	2.84	2.44	2.84	2.44
Mean score	$2.7 \pm 0.2$	$2.4 \pm 0.6$	$2.7 \pm 0.2$	$2.4 \pm 0.6$
	Commissures		AV area	
	RT3D-TTE	2D-TTE	RT3D-TTE	2D-TTE
Adequate (3)	21 (84%)	15 (60%)	19 (76%)	11 (44%)
Inadequate (2)	3 (12%)	4 (16%)	3 (12%)	6 (24%)
Not visualized (1)	1 (4%)	6 (24%)	3 (12%)	8 (32%)
Median score	2.80	2.36	2.64	2.12
Mean score	$2.6 \pm 0.5$	$2.1 \pm 0.6$	$2.3 \pm 0.4$	$1.8 \pm 0.7$

## P-98

### Integrated assessment of diastolic and systolic ventricular function using diagnostic MRI-catheterization: Validation in pigs and application in a clinical pilot study

Kuehne T. (1), Schmitt B. (1), Steendijk P. (2), Lunze K. (1), Ovroutski S. (1), Falkenberg J. (1), Rahmanzadeh P. (1), Maarouf N. (1), Ewert P. (1), Berger F. (1)  
German Heart Institute Berlin, Berlin, Germany (1); Leids Universitair Medisch Centrum, Leiden, The Netherlands (2)

**Objectives:** To develop and validate a method for the integrated analysis of systolic and diastolic ventricular function.

**Background:** An integrated approach to assess ventricular pump function, myocontractility (ESPVR) and diastolic compliance (EDPVR) is of high clinical value. MRI is well established for measuring global pump function and catheterization combined MRI was previously demonstrated to accurately measure ESPVR but not, yet, the EDPVR.

**Methods:** In 8 pigs the MRI technique was compared to conductance catheter methods (gold standard) for measuring the EDPVR in the left and right ventricle. Measurements were performed at rest and during dobutamine. For MRI, the ESPVR was estimated with a single-beat approach by synchronizing invasive ventricular pressures with cine-MRI derived ventricular volumes. The EDPVR was determined during preload reduction from additional volume data that was obtained from real-time velocity encoded MRI pulmonary/aortic blood flow measurements. Preload reduction was achieved by transient balloon occlusion of the inferior vena cava. The stiffness coefficient  $\beta$  was calculated by an exponential fit from the EDPVR. After validation in the animal experiments, the EDPVR was assessed in a pilot study of 3 patients with single ventricle using identical MRI and conductance catheter techniques.

**Results:** Bland-Altman tests showed good agreement between conductance catheter and MRI derived EDPVR. In both ventricles of the pigs, dobutamine enhanced myocontractility ( $p < 0.01$ ), increased stroke volume ( $p < 0.01$ ) and improved diastolic function. The latter was evidenced by shorter early relaxation ( $p < 0.05$ ), a downward shift of the EDPVR and a decreased stiffness coefficient  $\beta$  ( $p < 0.05$ ). In contrast, in the patients early relaxation was inconspicuous but the EDPVR shifted left-upwards and the stiffness constant remained unchanged. The observed changes in diastolic function were not significantly different when measured with conductance catheter and MRI.

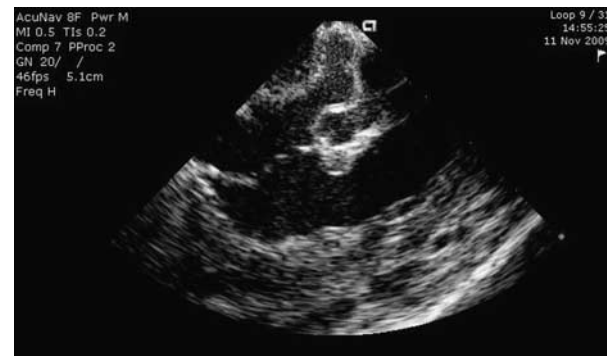
**Conclusions:** This novel MRI method provides differential information about diastolic function in conjunction with parameters of systolic contractility and global pump function.

## P-99

### Use of Monoplane Intracardiac Echocardiographic Transducer as a Transesophageal Probe in Low - Weight Infants Undergoing Complex Congenital Heart Surgery: Feasibility and Effectiveness.

Arcidiacono C., Micheletti A., Piazza L.P., Negura D.G., Butera G., Chessa M., Frigiola A., Carminati M.  
Department of Pediatric Cardiology and Cardiac Surgery. Policlinico San Donato IRCCS, Milan, Italy.

**Introduction:** Transesophageal echocardiographic (TEE) imaging provides valuable information on anatomy and function during congenital heart surgery. However, conventional multiplane transducers are usually unsuitable for small infants weighing less than 4 kg, or with abnormal oropharyngeal anatomy.



**Methods:** We intraoperatively studied 9 infants (4 males, 5 females, weight range 2.6–6.5 kg) with the commercially available 8F monoplane intracardiac ultrasound – tipped catheter (AcuNav, Acuson/Siemens) used as a TEE probe. 1 patient had ventricular septal defect (VSD), 1 had Tetralogy of Fallot (ToF), 2 had transposition of the great arteries (TGA), 1 had atrioventricular septal defect (AVSD), 2 had hypoplastic left heart syndrome (HLHS), 1 had anomalous origin of the left coronary artery from the pulmonary trunk (ALCAPA) and 1 had double outlet right ventricle (DORV) with VSD and coarctation. The probe was placed in the esophagus via the mouth or the nostrils.

**Results:** Despite the limitation of monoplane imaging, satisfactory views were obtained in all cases. Preoperative evaluation showed important anatomic details, such as anatomy of the pulmonary infundibulum and valve in the case of ToF or partial anomalous pulmonary venous return in 1 case of HLHS. Post-operative imaging allowed assessment of ventricular and valvar function in all cases and detection of a small residual VSD in one case. It showed good result of the Damus-Kaye-Stansel anastomosis in both cases of HLHS, successful re-implantation of the left coronary artery with good Color Doppler flow in the case of ALCAPA and good result of coarctation repair in the case of DORV. The probe steering system was generally unnecessary, and used occasionally only to improve contact with the esophageal wall. Transgastric views were usually very difficult to obtain. No complications related to handling or overheating of the probe were observed.

**Conclusions:** TEE imaging with the intracardiac probe is a feasible and safe technique that enables reliable intraoperative imaging in small infants undergoing cardiac surgery, who are unsuitable for conventional TEE imaging.

#### P-100

##### **Comparison of Radiation Dose in Paediatric Diagnostic Cardiac Catheter and Cardiovascular CT Angiography**

*Narayanan A., Tawil M.*

*Alder Hey Children's Hospital NHS Trust Liverpool United Kingdom.*

**Background:** Cardiac catheterisation and cardiovascular CT angiography provide substantial diagnostic information on congenital heart disease. However, these techniques are not without risk as they require use of ionising radiation. Based on Ionising Radiation (Medical Exposure) Regulation IR(ME)R, employers are required to optimise their procedures to ensure that doses are kept to the minimum. IR (ME) R requires employers to set Diagnostic reference levels (DRL) for radiological procedures.

**Objective:** To compare the effective doses of radiation in diagnostic cardiac catheter and cardiac CT angiography in children.

**Design:** Retrospective observational cohort study.

**Methods:** Data collected on 100 patients – 50 each from CT angiogram and diagnostic cardiac catheter. Following details were collected: Age at time of examination, body weight, CT dose index (CTDI) and Dose Length Product (DLP) from CT, and Dose Area Product (DAP) from Catheter Lab. Patients categorised into 5 different age groups. The mean CTDI, DLP, and DAP for each age group were measured. The data were submitted to the Integrated Radiological Services (IRS) for calculation of the effective dose. Cardiac CT scans were performed using retrospective ECG gated 16 multi-slice Philips machine. Biplane Philips projector was used in catheter Lab. The DLP of Cardiac CT and the DAP of cardiac catheters were converted into effective dose in mSv using conversion factor based on Schmidt et al study<sup>1</sup>.

**Results:** Patients categorised into the following age groups: <1 year, 1–5 years, 6–10 years, 11–15 years, and >15 years. Number of cardiac CT examinations and cardiac catheter for each age group: 25, 11, 2, 3, 3 and 11, 19, 7, 7, 6 respectively. Mean effective dose in mSv for each age group: CT (3.36, 3.17, 3.32, 3.57, 3.75), cardiac catheter (18.08, 18.19, 19.12, 9.68, 10.87).

**Conclusion:** There is no published data comparing Cardiac catheter and CT angiography in children. Our data revealed that CT provides significantly lower effective doses compared to the diagnostic cardiac catheter.

Large variation in radiation dose for cardiac catheter was noted.

#### P-101

##### **Detection of coronary complications after coronary reimplantation surgery: first experience with multislice computer tomography in children.**

*Marini D. (1), Defilippi C. (2), Di Rosa G. (2), Abbruzzese P. (1), Agnoletti G. (1)*

*(1) Paediatric Cardiology, Ospedale Infantile Regina Margherita, Torino; (2) Paediatric Radiology, Ospedale Infantile Regina Margherita, Torino*

**Introduction:** The main cause of long-term morbidity and mortality after successful coronary reimplantation of coronary arteries are complications at the ostial segments and/or proximal segments. The purpose of this study was to investigate the clinical usefulness of multislice computed tomographic angiography in detecting ostial and proximal coronary lesions in children having undergone coronary reimplantation surgery for transposition of the great arteries (TGA) and anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA).

**Methods:** Twenty-four children (aged 6,4 ± 2,2 years) having had arterial switch operation for TGA (N = 17) and direct reimplantation of the left coronary artery for ALCAPA (N = 7) underwent systematic selective conventional and multislice computed tomographic angiography at a mean follow-up of 6,2 ± 2,3 years from surgery. The ability of multislice computed tomography in detecting stenosis and other modifications of the coronary arteries' course was analyzed by 2 independent investigators.

**Results:** Multislice computed tomography, as compared with selective conventional coronary angiography, permitted the assessment of ostial and proximal coronary segments in every patient (N = 41 coronary arteries). It correctly identified 3 (12%) patients with significant coronary lesions (N = 1 proximal severe stenosis of the left coronary artery with post stenotic aneurysm, N = 1 sub occlusion and N = 1 occlusion of the right coronary artery) whom had been identified by means of conventional angiography.

**Conclusions:** These results indicate that multislice computed tomographic angiography is fully accurate in detecting ostial coronary artery stenoses, proximal coronary artery stenoses, or both type of lesions in paediatric patients having undergone surgical reimplantation of the coronary arteries. Our results suggest that multislice computed tomography could be used as a screening technique for detecting coronary complications in the follow-up of these patients before having recourse to conventional angiography.

#### P-102

##### **Stiffness and Wall Motion Velocities of Abdominal Aorta Measured by Tissue Doppler Imaging in Obese Children**

*Gurses D. (1), Ozyurek A.R. (2), Ulger Z. (2), Coker M. (2), Darcan S. (2), Levent E. (2)*

(1) Pamukkale University Faculty of Medicine, Department of Pediatric Cardiology, Denizli, Turkey; (2) Ege University Faculty of Medicine, Department of Pediatrics, izmir, Turkey.

Obesity in childhood has been associated with the development of early cardiovascular abnormalities. Evaluation of the aortic stiffness may be an indicator of early vascular changes. Tissue Doppler imaging (TDI) offers a new technique for assessing aortic wall expansion and contraction velocities and may provide a noninvasive approach to aortic wall mechanics. The aim of this study was to evaluate the stiffness and wall motion velocities of the abdominal aorta in obese children. The study was performed in 85 obese children and 45 healthy children as a control group. The mean ages were  $12.4 \pm 1.9$  and,  $12.3 \pm 2.1$  years in the study and control group respectively ( $p > 0.05$ ). All children were noninvasively evaluated with M-mode, 2-D, and Doppler echocardiography. Annulus of aorta and abdominal aorta diameters were measured. Abdominal aorta diameters were recorded at maximum systolic expansion and at minimum diastolic pressure. Aortic strain (S), pressure strain elastic module (Ep), pressure strain normalized by diastolic pressure (Ep\*), aortic stiffness  $\beta$  index ( $\beta$ ) and, aortic dispensability (DIS) were calculated using the measured data. Using TDI, maximum wall expansion velocity during systole (peak S) and maximum wall contraction velocity during diastole (peak D) were measured.

Aortic mechanical parameters were found significantly different in obese children as compared to controls: S ( $0.302 \pm 0.12/0.393 \pm 0.13$ ,  $p < 0.001$ ) and, DIS ( $1.4 \pm 0.8/2.2 \pm 0.8$ ,  $p < 0.001$ ) parameter was found significantly lower and, Ep ( $236 \pm 143/171 \pm 117$ ,  $p < 0.001$ ), Ep\* ( $2.36 \pm 1.4/1.7 \pm 1.1$ ,  $p < 0.05$ ) and  $\beta$  ( $1.9 \pm 0.9/1.2 \pm 0.4$ ,  $p < 0.001$ ) were found significantly higher than the control group. Significantly correlation was found aortic mechanical parameters and body mass index (BMI) ( $p$  values  $< 0.01$ , and  $r$  values  $-0.357$ ,  $-0.386$ ,  $0.410$ ,  $0.297$ ,  $0.316$  for the S, DIS, Ep, Ep\* and  $\beta$  respectively. The mean values peak S and D were found significantly higher in the obese group (peak S:  $5.30 \pm 0.103/4.89 \pm 0.9$ ,  $p < 0.05$ ; peak D:  $3.26 \pm 0.1/2.93 \pm 0.1$ ,  $p < 0.05$ ). Peak S and peak D values were significantly correlate with BMI ( $p < 0.05$ ,  $r = 0.240$ , and  $0.512$  respectively). The mean abdominal aortic flow was higher in the obese children than the controls ( $1.23 \pm 0.05/0.94 \pm 0.4$  L/m<sup>2</sup>,  $p < 0.05$ ).

Our data demonstrated that aortic stiffness was increased in obese children. Further pathophysiologic studies in larger population size are needed to confirm these findings in obese children.

#### P-103

##### Altered Biophysical Properties of the Aorta Following Kawasaki Disease

Dahdah N. (1), Fournier A. (1), Houde C. (2), Sandor G. (3)  
Centre hospitalier Universitaire Sainte-Justine, Montreal, Canada (1);  
Centre Hospitalier Universitaire de Quebec, Quebec, Canada (2);  
British Columbia Children Hospital, Vancouver, Canada (3).

**Background:** With the exception of the aneurismal coronary artery lesions, the long term sequelae of Kawasaki disease (KD) remain uncertain. Since KD is a generalized vasculitis, aneurisms may occur including the femoral, iliac, renal, axillary and brachial arteries. Attempts to detect endothelial dysfunction have been controversial lately and the hypothesis suggesting that KD is a precursor of atherosclerosis has not been proven yet. There are reports indicating an increase of the intima media thickness in the carotid arteries but a global assessment of the systemic arterial vasculature has not been fully studied yet. Aim: to study

the aortic biophysical properties (ABP) in cases with a history of KD.

**Methods:** specific echocardiographic and Doppler measurements were obtained from KD patients and healthy subjects. The ABP parameters were calculated by extrapolating from equations used in invasive techniques of ABP assessment.

**Results:** There were 70 KD and 165 controls aged  $8.18 \pm 5.42$  vs  $7.96 \pm 5.08$  yrs ( $p = 0.76$ ) with comparable weight, height and body surface area. The interval between the acute onset of KD and the current evaluation was  $4.7 \pm 4.8$  yrs (median 3.4 yrs). Among the KD patients only 7/70 (10%) had significant coronary artery sequelae. There were no statistical differences between cases and controls with respect to systolic or diastolic blood pressures ( $105.1 \pm 13.9$  vs  $104.9 \pm 12.8$  mmHg and  $60.0 \pm 9.1$  vs  $61.4 \pm 8.1$  mmHg;  $p = 0.92$  and  $0.23$ , respectively). The ABP demonstrated significant increase in KD vs controls for the following parameters: the Pulse Pressure  $45.0 \pm 11.5$  vs  $25.0 \pm 17.4$  mmHg ( $p < 0.001$ ), the Elastic Pressure Strain Modulus  $390.0 \pm 610.3$  vs  $153.8 \pm 116.6$  mmHg ( $p < 0.001$ ), the Beta Stiffness Index  $2.48 \pm 0.53$  vs  $2.11 \pm 0.37$  ( $p < 0.001$ ), and the Input Impedance  $244.4 \pm 101.5$  vs  $47.6 \pm 138.3$  dyne.s.cm<sup>-5</sup> ( $p < 0.001$ ), implying increased aortic stiffness, strain and impedance.

**Conclusion:** Our data strongly suggest ABP alteration in a large cohort of KD patients despite the large proportion of those without major coronary artery complications. These results suggest that the impact of the systemic vasculitis upon onset of the disease persists many years after contracting KD. The implication of these findings in early and mid adulthood are yet to be elucidated.

#### P-104

##### Surgical correction of congenital heart disease has no influence on left ventricular dyssynchrony

Klitsie L, Hazekamp M.G., Gesink-Van der Veer B.J., Blom N.A., Ten Harkel A.D.J.

Leiden University Medical Center, Leiden, The Netherlands

**Introduction:** In adults who previously had surgery for a congenital heart defect a considerable number of patients will develop heart failure. The amount of dyssynchrony has been considered one of the most important risk factors. It is, however, unknown if dyssynchrony will be present during childhood, shortly after correction of congenital heart defects. We hypothesize that patients who underwent surgical correction of congenital heart defects develop dyssynchrony shortly after operation, and that the amount of dyssynchrony is related to a decrease in strain values.

**Methods:** We studied 73 children aged 0–17 years, who underwent biventricular correction of their congenital heart defect. All patients were in sinus rhythm at the time of the study. Speckle Tracking Strain Analysis was used to measure strain and a dyssynchrony index was defined as the standard deviation of time to peak values for 6 myocardial segments. Longitudinal, radial, and circumferential strain and dyssynchrony were analyzed. Echocardiographic measurements were done before operation, to establish the baseline velocities, a day after operation and 10 days afterwards.

**Results:** Longitudinal strain significantly ( $P < 0.01$ ) decreases from  $19.5 \pm 4.2\%$  at baseline to  $14.9 \pm 3.6\%$  one day after surgery. After 10 days the strain was  $16.2 \pm 3.3\%$  which was unchanged as compared to immediately after surgery (NS). Radial and circumferential strain also significantly decrease one day after surgery compared to baseline (from  $37.1 \pm 15.0\%$  to  $26.4 \pm 12.5\%$  and from  $20.3 \pm 5.4\%$  to  $16.3 \pm 5.3\%$ , respectively).

However, these values returned to preoperative values 10 days after surgery. Neither the longitudinal, nor the radial or circumferential dyssynchrony indexes did significantly change immediately after surgery or after the 10-days follow-up period (NS).

**Conclusions:** This study shows for the first time that correction of congenital heart defects has no influence on left ventricular dyssynchrony. There was, however, a decrease in overall strain that partly returned to preoperative values after 10 days. If dyssynchrony will develop during longer-term follow-up remains to be established.

#### P-105

##### **Myocardial velocities and 2-d strain in 26 healthy children and adolescents before and immediately after a triathlon**

Schön P (1), Petzuch K (1), Kühn A (1), Elmenhorst J (1), Röhlig Ch (1), Schönfelder M (2), Oberhoffer R (2), Vögt M (1).

Department of Paediatric Cardiology and Congenital Heart disease, German Heart Centre Munich, Technische Universität München, Germany (1); Lehrstuhl für Sport und Gesundheitsförderung, Technische Universität München, Germany (2).

**Introduction:** Chronic endurance stress has been shown to have effects on systolic and diastolic myocardial function in adults. Very little is known on the normal "physiological" reaction of a growing heart in childhood under such conditions.

**Aim of the study:** We therefore decided to investigate 19 boys and 7 girls (8 to 16 years old) who participated in a childhood triathlon competition in Munich in 2009.

**Methods:** 2–4 weeks before the competition all participants underwent a first examination under rest with ECG and echocardiography. The heart was examined with 2-d echo, M-mode, PW-Doppler, colour Doppler, Tissue Doppler and 2d-Strain. All studies were stored digitally. Within 15 minutes after the competition all participants underwent the same echo-protocol. On off line analysis we measured left and right ventricular dimensions on 2-d and M-mode, all parameters of global systolic and diastolic function on conventional Doppler, myocardial velocities in systole and diastole as well as global and regional right and left ventricular strain and stroke volumes. To find out the effect of endurance stress on the growing heart we compared the two studies under rest and after the competition.

**Results:** Comparing haemodynamic parameters of the two conditions we found a significant increase in heart rate, shortening fraction and ejection fraction of the left ventricle under stress. Global systolic strain of the left ventricle increased significantly ( $p = 0.01$ ) as well as global systolic strain of the right ventricle ( $p = 0.002$ ). Also diastolic parameters like peak E-wave of LV decreased significantly ( $p = 0.003$ ).

**Conclusions:** Endurance training seems to have a considerable effect on the ventricular systolic and diastolic function in children. If these findings can be verified in a bigger normal population we decided to look at patients with congenital heart formations and find out differences in myocardial performance under stress.

#### P-106

##### **Two Dimensional and Color Doppler Derived Systolic Strain in Children: a Comparison Study using 2 Different Ultrasound Systems and 3 Analysis Software Packages.**

Koopman L.P., Slorach C., Manlhiot C., McCrindle B.W., Friedberg M.K., Jaeggi E.T., Mertens L.

Labatt Family Heart Center, Hospital for Sick Children and University of Toronto, Toronto, Canada

**Background:** Different ultrasound systems (US) and analysis software packages are used to measure 2-dimensional (2D) and

color tissue Doppler (CTD) derived myocardial deformation. However, data on whether these US are comparable in terms of deformation results are limited, especially in children.

**Methods:** During the same session children underwent an echo examination by the same operator using a Vivid 7 (General Electric-GE) and IE 33 (Philips) scanner to acquire B-mode and CTD images. Mean frame rate ( $\pm$  SD) was  $69 \pm 17$  fps for EP,  $72 \pm 16$  for QL and  $191 \pm 28$  for SP. Off-line analysis was performed using GE Echopac (EP) and Philips QLAB (QL) for 2D strain ( $\epsilon$ ) and Leuven/Speqle (SP) for CTD  $\epsilon$ . Left ventricular (LV) radial and circumferential systolic  $\epsilon$  was measured at papillary muscle level in the short axis (6 segments for 2D  $\epsilon$ , posterior wall for radial CTD  $\epsilon$ ). Longitudinal systolic  $\epsilon$  was measured in 6 segments from the apical 4 chamber view. Global  $\epsilon$  values were calculated from the average of the segmental values. Inter- and intra observer variability was examined in 15 participants. Agreement between the US was assessed by Bland-Altman analysis.

	Mean EP $\pm$ SD	Mean QL $\pm$ SD	Mean SP $\pm$ SD
Radial $\epsilon$ posterior wall (%)	55 $\pm$ 21	28 $\pm$ 13*	60 $\pm$ 15
Global radial $\epsilon$ (%)	47 $\pm$ 15	23 $\pm$ 6†	×
Global circumferential $\epsilon$ (%)	-18 $\pm$ 3	-21 $\pm$ 3†	×
Global longitudinal $\epsilon$ (%)	-19 $\pm$ 3	-20 $\pm$ 3	-19 $\pm$ 3
	Mean % $\Delta$ EP-QL (LA)	Mean % $\Delta$ EP-SP (LA)	Mean % $\Delta$ QL-SP (LA)
Radial $\epsilon$ posterior wall (%)	66 (-23 to 154)	-14 (-101 to 72)	-76 (-167 to 15)
Global radial $\epsilon$ (%)	63 (-5 to 130)	×	×
Global circumferential $\epsilon$ (%)	-13 (-47 to 20)	×	×
Global longitudinal $\epsilon$ (%)	-4 (-30 to 121)	1 (-33 to 35)	2 (-29 to 34)

LA = limits of agreement ( $\pm$ 2SD). \* $p < 0.001$  for QL versus EP&SP; † $p < 0.001$  for QL versus EP

**Results:** 34 children (12 controls and 22 with heart disease; mean age  $11 \pm 3$  yrs) were studied. Shortening fraction was  $36 \pm 6\%$ . Results are shown in the table. Radial  $\epsilon$  measured by QL was lower compared to EP and SP and variability between the 3 methods within participants was high. Circumferential  $\epsilon$  measured by EP and QL showed reasonable agreement, as did longitudinal  $\epsilon$  measured by the 3 methods. Intra/inter coefficient of variation was: radial  $\epsilon$  posterior wall: 11/24% (EP), 39/56% (QL), 12/12% (SP); global radial  $\epsilon$ : 10/17% (EP), 18/18% (QL); circumferential  $\epsilon$ : 10/10% (EP), 6/10% (QL); longitudinal  $\epsilon$ : 6/7% (EP), 5/5% (QL), 12/12% (SP).

**Conclusions:** Agreement between US for circumferential and longitudinal strain is reasonable, as is their reproducibility. 2-D radial  $\epsilon$  results are unacceptably different between different US and have moderate to poor reproducibility. These results are important when interpreting deformation imaging and have significant implications for serial evaluation of myocardial function in individual patients.

#### P-107

##### **Echocardiographic evaluation of right ventricle function after percutaneous balloon valvuloplasty in infants with pulmonary valve stenosis.**

Khanenova V.A., Kurkevych A.K., Maksymenko A.V., Boyko O.P., Stogova O.V., Rudenko N.M., Yemets I.M.

Ukrainian Children's Cardiac Center, Kyiv, Ukraine

**Objectives:** Percutaneous balloon pulmonary valvuloplasty (BPV) is the treatment of choice in children with moderate to severe pulmonary stenosis (PS). The aim of this study was to assess the RV function in patients with valvular PS before and after BPV by using new quantitative echocardiographic parameters.

**Methods:** We studied 65 infants with PS (the group 1) and 59 healthy infants (group 2). We used new complex echocardiographic method, which consisted of new parameters (measurements of LV, RV, LA chambers, thickness of walls of LV and

pressure in RV, PA) and special formulas based on this parameters for calculation of RV end-diastolic volume (RVEDV) and ejection fraction (RVEF) as well as Tissue Doppler velocities and TAPSE measurement. RV systolic and diastolic function was assessed by Tissue Doppler according to the following parameters: peak systolic velocity (Sm), peak early diastolic velocity (Em), peak late diastolic velocity (Am) and the Em/Am ratio. All infants with PS underwent complex echocardiographic study before BPV (subgroup 1a), early after BPV (mean 7 days, subgroup 1b) and at the mid-term follow-up (mean 6–12 months, subgroup 1c).

**Results:** All parameters of RV systolic and diastolic function (RVEF, Sm, Em/ Am ratio, TAPSE) were significantly decreased in subgroup 1a compared to the same parameters in the group 2 ( $p < 0.001$ ). In addition, the pressure gradient across pulmonary valve was inversely correlated with RVEF ( $r = -0.784$ ) and Em/ Am ratio ( $r = -0.638$ ). Early after BPV (subgroup 1b) RVEF and Em/ Am ratio decreased even further, while peak Sm and TAPSE slightly increased. At mid-term follow-up evaluation (subgroup 1c) all these parameters significantly increased ( $p < 0.001$ ).

**Conclusions:** After temporary decrease in RV diastolic function early after BPV for PS in infants, we observed improving of systolic and diastolic function in 6–12 month follow-up. Our complex echocardiographic quantitative method could be used for non-invasive RV function evaluation before and after in pediatric surgery.

## P-108

### Two Dimensional and Color Doppler Derived Radial Strain Rate in Children: a Comparison using 2 Ultrasound Systems and 3 Analysis Software Packages.

Koopman L.P., Slorach C., Manlhiot C., McCrindle B.W., Jaeggi E.T., Friedberg M.K., Mertens L.

Labatt Family Heart Center, Hospital for Sick Children and University of Toronto, Toronto, Canada

**Background:** Various ultrasound (US) and software systems are available to measure 2-dimensional (2D) and color tissue Doppler (CTD) derived myocardial strain rate (SR). However, data on the comparability of these methods are limited, especially in children.

**Methods:** Thirty-four children ( $11 \pm 3$  years) without ( $N = 12$ ) and with ( $N = 22$ ) heart disease were scanned to acquire B-mode and CTD images by Vivid 7 (GE) and iE33 (Philips) US for off-line analysis. EchoPAC (EP) and QLAB (QL) were used to obtain 2D SR. Leuven/Spqle software (SP), using GE images, was used to obtain CTD SR. Mean frame rates were  $69 \pm 17$  for EP,  $72 \pm 16$  for QL and  $191 \pm 28$  for SP. Radial systolic SR (S-SR) and diastolic SR (D-SR) were measured at papillary muscle level in the short axis (6 segments for 2D, posterior wall for CTD). Global SR was calculated as averaged segmental values. Inter- and intra observer variability was evaluated in 15 pts and reported as the coefficient of variation (COV). Agreement between systems was assessed by Bland-Altman analysis.

	Mean EP $\pm$ SD	Mean QL $\pm$ SD	Mean SP $\pm$ SD
Radial S-SR posterior wall (1/s)	$1.8 \pm 0.4$ †	$1.5 \pm 0.7$	$3.5 \pm 1.4$ †
Radial D-SR posterior wall	$-2.3 \pm 0.8$ #	$-4.7 \pm 2.3$	$-5.5 \pm 2.4$ *
Global S-SR	$1.7 \pm 0.3$	$1.5 \pm 0.7$	×
Global D-SR	$-2.0 \pm 0.7$ #	$-3.0 \pm 0.8$	×
	Mean% $\Delta$ EP-QL (LA)	Mean% $\Delta$ EP-SP (LA)	Mean% $\Delta$ QL-SP (LA)
Radial S-SR posterior wall (1/s)	27 (-70 to 125)	-54 (-105 to -4)	-77 (-167 to 13)
Radial D-SR posterior wall	-60 (-159 to 39)	-78 (-161 to 4)	-13 (-150 to 123)
Global S-SR	$14.4$ (-45 to 74)	×	×
Global D-SR	$-43$ (-98 to 12)	×	×

LA = limits of agreement ( $\pm 2SD$ ), † $p < 0.001$  SP versus EP&QL, \* $p < 0.001$  SP versus EP, ‡ $p = 0.01$  EP versus QL, #  $p < 0.001$  EP versus QL.

**Results:** Table 1 summarizes the main results. Intra/inter observer COV was: Radial S-SR posterior wall: 16/24% (EP), 34/39% (QL) and 20/20% (SP); Radial D-SR posterior wall: 32/33% (EP), 30/57% (QL) and 26/23% (SP); global S-SR: 13/16% (EP) and 34/38% (QL); global D-SR: 20/26% (EP) and 21/30% (QL).

**Conclusions:** Reproducibility of radial SR values was moderate to poor for EP and SP and poor for QL. Systolic and diastolic SR values by 2D techniques were significantly lower compared to CTD and lower compared to values from previously published clinical and experimental studies. Limits of agreement between the different techniques were unacceptably wide. Current 2D speckle tracking techniques use low frame rates and may be inadequate to accurately and reliably measure fast deformation events like SR in children.

## P-109

### Tissue Doppler Velocity Imaging in Children: a Comparison Between two Ultrasound Systems

Koopman L.P., Slorach C., Manlhiot C., McCrindle B.W., Friedberg M.K., Mertens L., Jaeggi E.T.

Labatt Family Heart Center, Hospital for Sick Children and University of Toronto, Toronto, Canada

**Background:** Measurement of ventricular long-axis myocardial velocities by pulse-wave (PTVI) or color (CTVI) Doppler velocity imaging is widely used to quantify systolic and diastolic function. However, there are limited data as to whether TVI-derived measurements from different ultrasound systems (US) are comparable. We prospectively compared TVI-results from 2 commonly used US and examined inter/intra observer reliability.

**Methods:** Forty-nine children ( $11.9 \pm 4$  years; 12 without and 37 with cardiac disease) with normal systolic function and heart rates were scanned with Vivid 7 (GE) and Philips IE33 (PH) US during the same exam by a single operator. PTVI and CTVI images were acquired from an apical 4-chamber view and stored for off-line analysis using Echopac (GE) and QLAB (PH) using default settings. CTVI frame rates were  $171 \pm 36/s$  for GE versus  $227 \pm 50/s$  for PH ( $p < 0.001$ ). Diastolic (E', A') and systolic (S') velocities were obtained from the annulus of right (TV) and left (MV) atrio-ventricular valve and ventricular septum (IVS) and results from the 2 US compared by paired t-test. Intra- and inter-observer variability was calculated in 15 participants using the coefficient of variation (COV).

Table: comparison of PTVI and CTVI velocities using GE and PH ultrasound systems

	PTVI-GE	PTVI-PH	Mean $\Delta$ PTVI	CTVI-GE	CTVI-PH	Mean $\Delta$ CTVI
MV-E'	$16.9 \pm 3.3$	$17.3 \pm 3.5$	$-0.3 \pm 2.9$	$12.0 \pm 2.9$	$8.9 \pm 1.7$	$3.4 \pm 2.8$ *
MV-A'	$6.1 \pm 1.7$	$5.5 \pm 1.4$	$0.7 \pm 1.2$ *	$3.1 \pm 1.4$	$2.5 \pm 0.6$	$0.7 \pm 1.3$ *
MV-S'	$9.1 \pm 2.2$	$9.5 \pm 2.1$	$-0.4 \pm 1.5$	$6.2 \pm 2.0$	$4.4 \pm 1.1$	$1.9 \pm 1.4$ *
IVS-E'	$12.9 \pm 2.3$	$12.3 \pm 2.4$	$0.7 \pm 1.7$ *	$9.6 \pm 1.9$	$8.7 \pm 2.0$	$1.0 \pm 1.6$ *
IVS-A'	$6.0 \pm 1.6$	$5.3 \pm 1.2$	$0.7 \pm 0.8$ *	$3.3 \pm 1.2$	$3.1 \pm 0.9$	$0.3 \pm 1.1$
IVS-S'	$7.5 \pm 1.2$	$7.6 \pm 1.3$	$-0.1 \pm 1.0$	$5.2 \pm 1.0$	$4.1 \pm 1.0$	$1.2 \pm 0.8$ *
TV-E'	$13.1 \pm 3.3$	$13.4 \pm 3.7$	$-0.2 \pm 2.7$	$9.4 \pm 2.7$	$7.4 \pm 1.8$	$2.4 \pm 2.1$ *
TV-A'	$8.0 \pm 2.7$	$7.4 \pm 2.5$	$0.7 \pm 1.7$ *	$5.1 \pm 2.3$	$3.4 \pm 1.1$	$1.8 \pm 1.5$ *
TV-S'	$10.9 \pm 3.0$	$11.5 \pm 2.9$	$-0.7 \pm 1.3$ *	$8.1 \pm 2.7$	$6.2 \pm 2.0$	$2.2 \pm 1.1$ *

Velocities in Cm/s ( $\pm$ SD), \* $p \leq 0.01$  for difference between GE and PH

**Results:** The intra- and inter-observer COV for PTVI-derived measurements ranged from 4–10% for GE and 1–14% for PH. COV for CTVI- derived measurements ranged from 4–19% for GE and 6–25% for PH. The table summarizes the main findings.

**Conclusions:** There is a good agreement between GE and PH-derived PTVI velocities with low mean differences and no systematic bias. In contrast, GE-derived CTVI velocities are systematically higher when compared to PH, questioning the interchangeability of CTVI findings between these systems.

**P-110**

**Evaluation with Echo-Dipyridamole or supine bicycle exercise stress echocardiography to authorize physical activity without restriction in patients with a history of Kawasaki disease**

De Lucia V. (1), Mazzoni A.M. (2), Terrazzi M. (2), Lunardini A. (1), Recla S. (1), Spadoni I. (1), Giusti S. (1)  
 Pediatric Cardiology and GUCH Unit (1); Department of Cardiology (2); "G. Monasterio" Tuscan Foundation, "G. Pasquinucci" Heart Hospital, Massa, Italy

**Introduction:** Kawasaki Disease (KD) is a systemic self-limiting vasculitis of childhood with an endothelial damage of small-medium-sized arteries and coronary arteries. Despite recommendations on KD of the American Heart Association (AHA), there is a restriction for physical activity and very often the physical suitability for athletic sports is not granted.

**Methods:** 30 consecutive patients (pts) (16 males) with previous KD and a mean age of 12 (3,3–26) years underwent stress echocardiography with Dipyridamole (DSE), or exercise with supine bicycle (ESE) if Dypiridamole was contraindicated. DSE protocol was: Dypiridamole 0.56 mg/kg over 4 minutes followed by a second dose of 0,28 mg/Kg over 2 minute, Aminophyllin (1 mg/kg) at the end of the test. Personal history was reviewed. All pts had also physical examination, basal EKG and echographic measurement of the diameter of the coronary arteries.

**Results:** mean age of pts at the time of the acute phase of KD was 3 yrs (range 2 months –7 yrs). The coronary involvement had been documented by echocardiography. The AHA risk level was I in 12 pts, II in 12 pts, III in 5 pts and IV in 1pt. At the time of the study all pts were asymptomatic. Stress-echocardiography was performed in 29 pts (DSE: 23 pts, ESE: 6 asthmatic pts) 4,7 years (range 1,2–23,4) after the acute phase of the disease. One pt with hystory of asthma could not perform ESE because he was too short for the bicycle. All stress-echocardiographic studies were judged of good quality. In 1 pt the test was interrupted because of a mild reaction to dipyridamole (tachycardia and vomit). All diagnostic stress tests were negative for inducible ischemia.

**Conclusions:** in our experience, DSE and ESE are feasible and well tolerated methods for the evaluation of inducible ischemia in pts with KD. In absence of inducible ischemia, after the acute phase of the disease pts in risk level I-III can safely perform physical activity without restriction, according to the AHA 2004 guidelines.

**P-111**

**Left and right ventricular deformation and mechanical dyssynchrony in young patients after repair of Tetralogy of Fallot**

Friedberg M.F., Fernandes F.P., Roche S.L., Fackoury C., Kantor P.F.  
 Hospital of Sick Children, Toronto, Canada

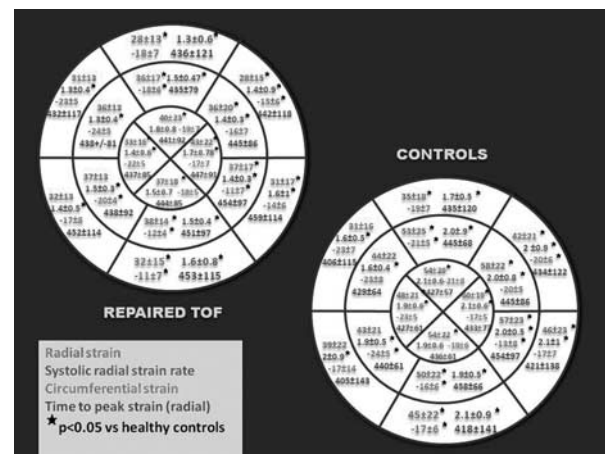
**Background:** Right (RV) and left (LV) ventricular dysfunction can develop late after Tetralogy of Fallot (TOF) repair and are important clinical problems. Mechanical dyssynchrony (MD) and impaired myocardial deformation may be important contributory factors. The aim of this study was to investigate LV and RV myocardial deformation and MD in young patients with repaired TOF using 2-dimensional (2D) speckle tracking.

**Methods:** Healthy controls and repaired TOF pts with normal LV EF (>55%) and qualitatively good RV function prospectively underwent 2-D echo. Apical 4-chamber and short axis views at basal, mid and apical levels were acquired at frame rates of 60–90 fps (Vivid 7, GE). Strain (S) and strain rate were measured by 2D speckle tracking (Echopac, GE). Longitudinal S was

measured at the basal and mid third of the lateral RV and LV walls. Radial and circumferential S and radial S rate were measured at 16 basal, mid and apical LV segments. LV and RV longitudinal MD (absolute difference in time to peak longitudinal S between basal septum and basal LV and RV lateral walls respectively), LV radial MD (septal-posterior wall delay and 6 segment SD of time to peak radial strain at mid-ventricular level) were calculated and corrected for heart rate (delay/ R-R interval X1000). Non-paired t-tests were used to compare results between groups.

**Results:** Forty one repaired TOF pts (median age 12; range 5–16 yrs) and 60 healthy controls (median age 13; range 2–16 yrs) were studied. Results are shown in the table. TOF pts had reduced RV longitudinal S and increased RV intraventricular MD. LV radial S and S rate were decreased compared to controls. LV longitudinal deformation and MD were similar between the groups.

**Conclusions:** RV MD in association with decreased RV and LV deformation are present in young pts after TOF repair. Serial evaluation of MD and myocardial performance as possible contributors to later development of overt ventricular dysfunction and as potential therapeutic targets warrants further prospective study in this population.



	TOF	Controls	P value
<b>Myocardial Deformation</b>			
<b>LV Radial strain</b>			
base*	30 ± 14	39 ± 20	<0.01
mid*	36 ± 16	50 ± 22	<0.01
apex†	38 ± 19	54 ± 20	<0.01
<b>LV Radial strain rate</b>			
base*	1.5 ± 0.74	1.9 ± 0.82	<0.01
mid*	1.4 ± 0.39	1.9 ± 0.61	<0.01
apex†	1.6 ± 0.77	2.0 ± 0.65	0.01
<b>LV Circumferential strain</b>			
base*	-16 ± 6	-18 ± 7	0.1
mid*	-16 ± 5	-19 ± 6	<0.01
apex†	-19 ± 6	20 ± 6	0.4
<b>Longitudinal strain</b>			
RV (base+mid)	-23 ± 5	-27 ± 9	0.03
LV (base+mid)	-16 ± 5	-17 ± 4	0.4
<b>Mechanical Dyssynchrony</b>			
Longitudinal RV-IVS	139 ± 163	76 ± 99	0.04
Longitudinal LV-IVS	62 ± 56	45 ± 51	0.1
Radial LV Septal-posterior wall delay	42 ± 42	36 ± 32	0.4
SD of time to peak radial strain (6 segments)	24 ± 19	24 ± 19	0.5
SD of time to peak circumferential strain (6 segments)	52 ± 40	51 ± 28	0.8

\*Values represent the average of 6 myocardial segments.

†Values represent the average of 4 myocardial segments

## P-112

**Usefulness of cardiovascular magnetic resonance in children with suspected myocarditis**

Werner B. (1), Piotrowska Kownacka D. (2), Kownacki L. (2),  
Wójcicka Urbanska B. (1), Rowinski O. (2)

Department of Pediatric Cardiology and General Pediatrics, Medical University of Warsaw, Warsaw, Poland (1); II Department of Clinical Radiology, Medical University of Warsaw, Warsaw, Poland (2)

**Objective:** The aim of the study was to assess the usefulness of cardiovascular magnetic resonance (CMR) in the diagnosis of myocarditis in children.

**Methods:** 11 boys aged 13–17 years were admitted to cardiology department due to sharp, stabbing chest pain. 7 of them had a history of intensive physical training and respiratory infections. In all of them laboratory data, ECG, Echo-2D and CMR were performed. CMR protocol included short axis cine, T2 weighted “black blood” imaging with fat saturation (myocardial oedema imaging) and delayed enhancement imaging after Gd-DTPA infusion (0.1 mmol/kg b.w.).

**Results:** In 9 patients myocardial enzymes including troponin-I levels were elevated. In all patients ST segment elevations (myocardial infarction-like patterns) in ECG were observed. Echocardiography revealed impaired regional left ventricular function in 4 and pericardial effusion in 3. Based on CMR findings acute myocarditis and/or acute pericarditis were confirmed in 9/11 and 2/11 patients respectively. Left ventricular EF was mildly decreased in 5/11 children. Myocardial oedema and delayed gadolinium enhancement regions were present in subendocardial layers in 9/11 patients, mainly in lateral segments. Whole heart, navigator-gated 3D MR coronary angiography (post contrast) performed in 5/11 patients revealed normal proximal segments of main coronary arteries. Additionally whole heart 3D datasets gave a similar information as delayed enhancement images regarding myocardial injury in patients with acute myocarditis.

**Conclusions:** Cardiovascular magnetic resonance is a useful, non-invasive and well tolerated method in pediatric patients with suspected myocarditis. Our initial findings suggest that post-contrast navigator gated 3D whole heart angiography could be used as “one stop shop” alternative in pediatric population with suspected myocarditis.

## P-113

**Evaluation of Left Ventricular Force Frequency Response Using Systolic Tissue Velocities and Radial Strain in a Novel Rabbit Model of Heart Rate Dependent Contractile Remodeling**

Slorach C., Mertens L. Hui W. Jiang J. Gross G. Friedberg M.  
Hospital for Sick Children, Toronto, Canada

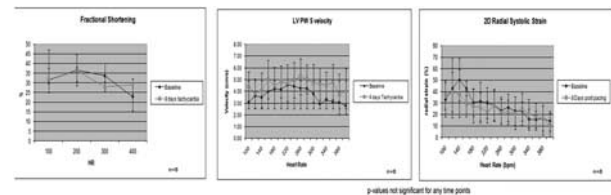
**Background:** Assessment of myocardial contractility in small animal models has traditionally necessitated invasive measurements, limiting serial acquisition of data. The use of newer echo methods to study myocardial performance in small animal experimental models has important potential implications for study of heart disease in children and fetuses, but there is little experience in this area. We report initial experience studying effects of chronic tachycardia pacing on myocardial performance in a novel rabbit model using 2-D speckle tracking strain and tissue Doppler (TDI) velocities to characterize the myocardial force-frequency response (FFR).

**Methods:** After induction of complete heart block (CHB) by A-V node ablation in a rabbit model, tachycardia was instituted for 8

days by continuous right ventricular pacing at 280 bpm. FFR was evaluated (by pacing from 100–400 bpm at 20 bpm increments) at baseline (day 0 after ablation, before tachycardia pacing) and after 8 days of tachycardia pacing. At each HR increment, M-mode, 2D and color TDI parasternal short axis views were acquired. Shortening fraction (100 beat increments), posterior wall TDI systolic velocity and radial strain by 2-D speckle-tracking (20 beat increments) were measured off-line (Echopac, GE). These were compared at baseline and after tachycardia pacing, at each FFR HR, using the paired student's t-test.

**Results:** Of 11 rabbits studied, ablation was successful in 8. Short-axis image acquisition, TDI and deformation analysis was feasible in all animals. Frame rates were 200–300 fps for color TDI and 80–120 for 2-D imaging. FFR by M-mode, tissue velocities and radial strain are shown in the figure. Standard deviations for M-mode, TDI velocities and strain were relatively high. Overall, there were no significant differences in myocardial velocities or deformation FFR after tachycardia pacing.

**Conclusions:** These pilot data demonstrate high feasibility of deformation and TDI imaging for serial study of myocardial performance at high heart rates in small animals, although measurement variability is a concern. While further validation of deformation imaging for assessment of myocardial contractility in small animals is required; newer echo-based applications have important translational potential for fetal and pediatric cardiac research.



## P-114

**Intraventricular Timing of Strain and Strain Rate in healthy children**

Laser K.T. (1), Hauffe P. (1), Beck C. (1), Haas N. (1),  
Kececioglu D. (1), Körperich H. (2)

Center of Congenital Heart Disease (1), Institute for Radiology, Nuclear Medicine and Molecular Imaging (2), Heart and Diabetes Center NRW, Bad Oeynhausen

**Background:** Timing events can be measured by speckle tracking techniques with good reproducibility. Knowledge of normal LV-intraventricular delay (ID) in children is the basis for understanding abnormalities.

**Methods:** We prospectively enrolled 174 healthy children/adolescents (85 males, 0–20.3y., median = 9y.) performing 2DE with a Vivid 7 machine including 4-chamber and short axis loops. Longitudinal, circumferential and radial event timing of Strain (S) and Strain Rate (Sr) were measured using Echopac Software (2D-Strain). Intraventricular delay was assessed using all opposite segments of each view. Timing was normalized to a heart rate of 60 bpm. Children were divided into 3 subgroups (I:50–89, II:90–129, III:130–180 bpm) to test the influence of heart rate, statistical analysis was done using the Anova test.

**Results:** Shortest ID was detected in group III (radial S  $10,3 \pm 12,4$ , Sr  $14,3 \pm 16,3$ , circumferential S  $24,3 \pm 19,6$ , Sr  $16,5 \pm 15,7$ , longitudinal S  $15,6 \pm 7,9$ , Sr  $23,7 \pm 16,3$  ms). Statistical differences ( $p < 0,05$ ) between group III and both other groups could be detected in the short axis views for all combinations of segments using radial S and Sr. Circumferential Sr and S only showed corresponding changes for the lateral-septal

and antero-septal-posterior delay, longitudinal S and Sr for the mid-septal-midlateral delay.

**Conclusions:** ID in children is heart rate dependent with inversely proportional relation. Timing of radial S and Sr detect most differences regardless of segment combination used. Longitudinal S and Sr should preferably be measured in mid-ventricular segments. These results may be important for the diagnosis of dyssynchrony in children.

#### P-115

##### Assessment of global and regional volumes by RT3DE and CMR

Laser K.T. (1), Barth P. (2), Beck C. (1), Haas N.A. (1), Kececioglu D (1), Burchert W. (2), Körperich H. (2)

Center of Congenital Heart Disease (1), Institute for Radiology, Nuclear Medicine and Molecular Imaging (2), Heart and Diabetes Center NRW, Bad Oeynhausen

**Background:** Left ventricular function as well as dyssynchrony can be described by quantification of entire ventricular volumes as well as subvolumes and their timing within one cardiac cycle. We investigated a new model based tool for CMR-volumetry in comparison to RT3DE.

**Methods:** We prospectively enrolled 24 healthy children/adolescents (12 males, 5,3–18,8y., median = 11,8y.) performing RT3DE and CMR with a Vivid 7 machine (GE) and a 1.5 Tesla Scanner (Philips) respectively. Volumetric measurements were quantified using the LV-Analysis tool (Tomtec) for Echo and CMR. The RT3DE quantification process had been harmonized to our standard CMR volumetry software as published before. Global and regional volumetric parameters (17 segment model) were calculated twice by RT3DE using different datasets. Comparison to CMR was carried out using the Bland Altman method.

**Results:** TOMTEC-RT3DE calculated larger EDV ( $-9,8 \pm 15,1\%$   $r = 0,94$ ) and smaller ESV ( $9,6 \pm 25,3\%$   $r = 0,80$ ) leading to higher SV ( $-22,2 \pm 17,3\%$   $r = 0,93$ ) than measured by TOMTEC-CMR. Interstudy-reproducibility of RT3DE was excellent (EDV  $3,8 \pm 8,5\%$   $r = 0,96$ , ESV  $0,0 \pm 15,2\%$   $r = 0,93$ , SV  $5,8 \pm 16,1\%$ ,  $r = 0,87$ ). Differences in timing of the 17 segmental subvolumes ranged from  $-1,4 \pm 7\%$  to  $1,3 \pm 8,9\%$  by RT3DE and  $-1,8 \pm 8,3\%$  to  $13,8 \pm 9,4\%$  compared to CMR. Calculation of the maximal difference within the subvolumes between systole and diastole ranged from  $0,17 \pm 7,6\%$  to  $8,4 \pm 20,2\%$  by RT3DE and from  $-2,3 \pm 32,4\%$  to  $25,4 \pm 19,9\%$  compared to CMR.

**Conclusions:** Reproducibility of global RT3DE data was excellent, the correlation with the CMR tool was especially less for SV because of underestimation of EDV and overestimation of ESV by CMR. Regional timing and quantitative changes in subvolumes could reproductively be measured by RT3DE. Comparison to CMR revealed acceptable values. Higher standard deviations increase the dyssynchrony indices in this setting.

#### P-116

##### Preserved aortic elasticity in patients with Marfan syndrome during five years of follow-up

Baumgartner D., Baumgartner C. (2), Karall T (1), Schweigmann U (1), Stein J.I. (1)

Clinical Division of Pediatrics III, Innsbruck Medical University, Innsbruck, Austria (1); Department of Biomedical Sciences and Engineering, UMIT, Hall in Tirol, Austria (2)

Aortic wall dysfunction caused by loss of elasticity is the primary cause of premature death in patients with Marfan syndrome. We

sought to determine aortic elastic parameters noninvasively during a follow up of 5,4 years in 3 different age groups. 46 Patients with Marfan syndrome according to the Ghent criteria aged  $16,9 \pm 11,8$  years were investigated prior to medical and surgical therapy (1) and  $5,4 \pm 2,9$  years later (2) under medication with a betablocker  $\pm$  losartan. 17% ( $n = 8$ ) had prophylactic aortic root replacement surgery. Diameter measurements were obtained in the aortic root, the ascending and abdominal aorta from M-Mode echocardiographic images by a special autocontour finding software. After simultaneous blood pressure measurements aortic elastic parameters were calculated.

Examination	age 0–10	age 10–20	age 20–44
	years group A (n = 17)	years group B (n = 16)	years group C (n = 13)
	1 2	1 2	1 2
Aortic root diameter (mm)	25,6 $\pm$ 4,3 29,4 $\pm$ 6,1*	36,7 $\pm$ 6,2 37,8 $\pm$ 5,1	44,6 $\pm$ 14,1 37,0 $\pm$ 5,3
Ascending aortic diameter (mm)	18,1 $\pm$ 3,0 21,6 $\pm$ 3,7*	26,1 $\pm$ 5,1 28,3 $\pm$ 3,6	32,9 $\pm$ 6,5 30,1 $\pm$ 3,0
Ascending aortic distensibility ( $\text{kPa}^{-1} \cdot 10^{-3}$ )	43 $\pm$ 19,8	28,4 $\pm$ 16,1	23,9 $\pm$ 17,2
Abdominal aortic diameter (mm)	44,2 $\pm$ 20,8 9,6 $\pm$ 2,3 12,2 $\pm$ 3,1*	29,4 $\pm$ 15,8 13,5 $\pm$ 1,8 16,8 $\pm$ 2,1	22,3 $\pm$ 13,0 16,8 $\pm$ 5,1 19,0 $\pm$ 3,3*
Abdominal aortic distensibility ( $\text{kPa}^{-1} \cdot 10^{-3}$ )	76,0 $\pm$ 33,2	46,7 $\pm$ 15,8	30,3 $\pm$ 17,5
Aortic root diameter/BSA	70,1 $\pm$ 22,7 30,1 $\pm$ 8,9 22,8 $\pm$ 5,7*	42,1 $\pm$ 21,3 21,9 $\pm$ 3,8 19,7 $\pm$ 2,8*	30,4 $\pm$ 13,8 22,9 $\pm$ 6,3 19,1 $\pm$ 2,9
Asc. aortic diameter/BSA	21 $\pm$ 4,8 16,8 $\pm$ 3,9*	15,6 $\pm$ 2,6 14,8 $\pm$ 2,3	17,1 $\pm$ 3,4 15,5 $\pm$ 1,5

\* $p < 0,05$ ; BSA body surface area

Ascending and descending aortic distensibility remained unchanged during follow-up in all age groups. Young patients showed higher values than older ones. Lowest distensibility values ( $< 20 \text{ kPa}^{-1} 10^{-3}$ ) were observed in patients before operation. In unoperated patients ( $n = 38$ ), especially in age group A, absolute aortic diameters increased, while all diameters normalized for BSA decreased. In older patients (group C) absolute and normalized aortic diameters did not change significantly. No acute vascular complications occurred during follow-up.

Loss of aortic elasticity can be accurately quantified from echocardiographic images in patients with Marfan syndrome. During 5 years of medical therapy aortic wall function was preserved in all age groups, young patients showed the greatest benefit. Low distensibility ( $< 20 \text{ kPa}^{-1} \cdot 10^{-3}$ ) can be recommended as an additional indicator for optimized timing of aortic root replacement surgery.

#### P-117

##### Value of exercise systolic blood pressure and systolic pressure gradient in assesment of children after repair of coarctation of the aorta

Djukic M., Vukomanovic G., Ilisic T., Stefanovic I., Dabetic M., Ilic S., Jovanovic I.

University Childrens Hospital, Belgrade, Serbia

**Objective:** To assess the diagnostic value of exercise systolic blood pressure (SBP) and maximal systolic pressure gradient across the descending aorta (SPG) in children after repair of the coarctation of the aorta (CoAo).



**Method:** Study group included 20 healthy children (Group I) and 20 patients after repair of CoAo, without significant residual deformation of aortic arch and/or isthmus (according to 2D echo and/or MRI). Those patients were subdivided on the basis of SPG at rest in Group II (<25 mmHg) and Group III (PG ≥ 25 mmHg). Mean age in the coarctation group was 14.3 ± 4 yrs, with aged and sex matched controls. The subjects underwent bicycle exercise test using the modified McMaster protocol. SBP was measured on the right arm both at rest and at the maximal exercise. SPG across the descending aorta was measured by CW Doppler during exercise.

**Results:** The most important data are presented in the Table:

	I group (Healthy children) n = 20	II group (PG < 25 mmHg) n = 11	III group (PG ≥ 25 mmHg) n = 9
SBP at rest (mmHg)	123 ± 13	127 ± 11	136 ± 19
Max exercise SBP (mmHg)	163 ± 17	174 ± 33	188 ± 30
SPG at rest (mmHg)	8 ± 3	18 ± 5	32 ± 3
Max exercise SPG (mmHg)	20 ± 5	46 ± 17	75 ± 11
	P value Gr.I vs. II	P value Gr.II vs. III	P value Gr.I vs. III
SBP at rest (mmHg)	0,097	0,087	0,016*
Max exercise SBP (mmHg)	0,205	0,165	0,028*
SPG at rest (mmHg)	9,6E-05**	2,0 <sup>E</sup> -06**	9,1E-11**
Max exercise SPG (mmHg)	0,0004**	0,0001**	1,46E-10**

\*P value < 0,05, \*\* P value < 0,01

**Conclusion:** This study suggests that exercise systolic blood pressure is not very sensitive variable either in their difference from controls or in their relation to increased repair site gradient. Systolic pressure gradient across the descending aorta, however is much more informative and sensitive parameter, particularly in a group of patients with increased repair site gradient. Exercise testing may have a role in assessing patients after correction of CoAo, particularly for the follow up and risk stratification of these patients, the early diagnosis of hypertension and “vascular dysfunction”.

#### P-118

##### Right ventricular function assessed by Tissue Doppler imaging before and after transcatheter closure of atrial septal defect.

Laskari C.V., Kiaffas M.G., Tsutsinos A.I., Rammos S.  
Onassis Cardiac Surgery Center, Athens, Greece.

Patients with an atrial septal defect (ASD) have long-standing right ventricular dilation and dysfunction. We used Tissue Doppler Imaging (TDI) to assess right ventricular systolic and diastolic function in patients with ASD before and after closure with the Amplatzer septal occluder.

**Methods:** 95 pts with ASD secundum, mean age 23 yrs (5–65), 32 male and 63 female underwent transcatheter closure of their ASD with an Amplatzer septal occluder. ASD size was 8–35 mm by echocardiography and mean Amplatzer size was 24 mm (8–40). After conventional echocardiographic assessment, pulsed TDI was obtained from the apical 4-chamber view at the basal RV free-wall-tricuspid annular junction. The following measurements were made: peak early systolic myocardial tissue velocity (e), peak late systolic myocardial tissue velocity (a), e/a ratio, peak systolic myocardial tissue velocity (s). Tricuspid annular motion was measured by M-Mode echocardiography in the 4-chamber view.

Results	Pre-Amplatzer	Post-Amplatzer	p
E (cm/sec)	19 ± 2	16 ± 2	<0.01
A (cm/sec)	15 ± 4	11 ± 4	0.05
E/A	1.38 ± .2	1.45 ± 2	NS
S (cm/sec)	18 ± 2	16 ± 2	<0.01
TV M-Mode (mm)	33.7 ± 6	25.2 ± 3	<0.02

**Conclusion:** There is dramatic improvement in both systolic and diastolic right ventricular function, measured by TDI indices the next day after transcatheter ASD closure, even in pts with long-standing RV dysfunction. Very high myocardial tissue velocities tend to normalize soon after the device placement. This is probably due to the sudden relief from the long-standing volume overload of the right ventricle secondary to the abrupt removal of the left to right shunt.

#### P-119

##### Echocardiographic morphological parameters for detection of the coarctation aorta in neonates

Ilisić T., Ilić S., Parezanović V., Đukić M., Vukomanović G., Stefanović I., Raus M., Vulićević I., Jovanović I.  
University Children's Hospital, Belgrade, Serbia.

**Objectives:** Neonatal coarctation of the aorta (CoA) diagnosis is still presenting a challenge in everyday practice. The aim of this study is to identify easy obtainable 2D echocardiographic parameters for detection of the CoA in neonatal period.

**Methods:** Echocardiographic evaluation was performed in 30 newborns with CoA and 20 healthy neonates (control group). Among 30 newborns in CoA group, 15 patients had isolated CoA (group I), and 15 newborns had ventricular septal defect – VSD (group II). Measurements of the aortic arch were obtained by two-dimensional echocardiography at the end of systole. Proximal transverse arch (PTL) diameter was measured at the origin of the left carotid artery (LCA) and distal transverse arch (DTL) at the origin of the left subclavian artery (LSC). Distance between LCA and LSC was measured between origins of the LCA and LSC. Index I was calculated as a ratio of PTL and distance between origins LCA-LSC, and Index II was calculated as a ratio of DTL and distance between LCA-LSC origins.

**Results:** The main results are presented in the following Table:

	Group I (CoA) n = 15	Group II (CoA+VSD) n = 15	Control group n = 20
PTL (mm)	4.59 ± 1.10	4.61 ± 1.60	6.52 ± 0.84
DTL (mm)	3.83 ± 1.01	3.48 ± 1.27	5.73 ± 0.73
LCA-LSC (mm)	6.47 ± 0.92	5.97 ± 1.66	4.14 ± 0.48
Index I	0,54 ± 0,10	0,47 ± 0,05	1,39 ± 0,24
Index II	0,66 ± 0,13	0,63 ± 0,11	1,60 ± 0,30
	P value group I vs. control	P value group II vs. control	P value group I vs. II
PTL (mm)	p < 0,0001	p < 0,0001	0.9563
DTL (mm)	p < 0,0001	p < 0,0001	0.3520
LCA-LSC (mm)	p < 0,0001	p < 0,0001	0,252
Index I	p < 0,0001	p < 0,0001	p > 0.05
Index II	p < 0,0001	p < 0,0001	p > 0.05

A cut-off point at 0.39, for Index I, showed a sensitivity of 92% and specificity of 99% for detection CoA, and for Index II a cut-off point at 0.44 showed a sensitivity of 96% and specificity of 99% for the diagnosis of neonatal CoA.

**Conclusions:** By using these echo indexes, 2D echocardiographic aortic arch measurement becomes a simple, reliable noninvasive method for the evaluation of aortic coarctation in neonatal period. These measurements have a high sensitivity and specificity for diagnosis of the CoA in population of newborns, and may lead to earlier diagnosis and subsequent surgical correction.

#### P-120

##### **Strain Rate Imaging for the Evaluation of Left Ventricular Function after Patent Ductus Arteriosus Closure**

Ghonimy R.\*, Adel M.\*, Hegab M.\*\*\*, Khairee E.\*\*

\* Cardiology and \*\* Cardiothoracic Surgery department at Tanta University Hospital, and \*\*\* Mahala Cardiac Center. EGYPT.

**Background and objectives:** Ductal closure may affect systolic and diastolic left ventricular (LV) function. We planned this study to evaluate the effect of surgical ligation of patent ductus arteriosus (PDA) on LV systolic and diastolic function, correlating traditional methods to Doppler tissue imaging (DTI) based mitral annulus velocities and its new application strain rate imaging.

**Methods:** The study population consisted of 20 children with PDA who had full Doppler, two-dimensional-echocardiography studies with measurement of mitral inflow velocities in early and late diastole (E and A wave), peak systolic mitral annular velocity (Sm) early and late diastolic mitral annular velocity (Em and Am) by DTI, Tei index, strain rate imaging and standard chamber dimensions. All children were studied before and with in one month after surgical closure of PDA that done by direct surgical ligation.

**Results:** Surgical closure of PDA produces significant decrease in E wave, Em and strain rate E (SR E), while there was no significant changes in A wave, Am or strain rate A (SR A). Ejection fraction (EF) and Sm decreased significantly while peak systolic strain rate (PSSR) and Tei index increased significantly. There was a good correlation between Sm and EF and fraction shortening (FS), and PSSR showed good correlation with Tei index and EF. There is a significant positive correlations between SR E, E and Em, and between SR A, A and Am.

**Conclusions:** Changes in LV volume and function caused by PDA are reversible after surgical closure. Strain rate imaging can assess both LV systolic and diastolic function after PDA closure.

**Key words:** patent ductus arteriosus, left ventricular function, strain rate imaging.

#### P-121

##### **Use of the new Amplatzer duct occluder II in comparison with Amplatzer duct occluder – single center experience**

Venczelova Z., Masura J., Tittel P.

National Institute of Heart Diseases – Children's Cardiac Center, Bratislava, Slovakia

**Objectives:** Determine the safety and efficacy, advantages and disadvantages in using Amplatzer duct occluder (ADO) or Amplatzer duct occluder II (ADO II).

**Methods:** All patients with a device duct closure between September 2005 and September 2009 were included. We analyzed the catheterization data and echocardiographies in 41 patients with ADO closure and 42 patients with ADO II closure.

**Results:** Between September 2005 and September 2009, 41 arterial ducts were closed with ADO (group I) and between April 2008 and September 2009, 42 ducts were closed with ADO II (group II). In group I, the mean age was 3 years 4 months, the mean weight was 14,97 kg. The mean minimal ductal diameter

was 2,4 mm. The closure with ADO was successfully performed in all patients. Immediate complete closure was obtained in 26 of 41 patients (63,41%), closure at 24 hours was complete in 39 of 41 patients (95,12%). At 1 month, the complete closure was in all examined patients (100%). 1 patient was recatheterised for device protrusion into the aortic isthmus 26 months post closure with an invasive gradient of 14 mmHg.

In group II, the mean age was 4 years and 7 months, the mean weight was 19,79 kg. The mean minimal ductal diameter was 1,78 mm. The closure with ADO II was successfully performed in all but one patient, in this patient, the occluder protruded significantly into the aortic isthmus and was replaced by ADO. Immediate complete closure was in 36 of 42 patients (85,71%), closure at 24 hours was complete in 41 of 42 patients (97,62%). At 1 month, there was 1 small residual shunt, which was completely closed at 6 month follow – up. There are no significant aortic protrusions and left pulmonary artery stenosis during follow – up period.

**Conclusion:** The duct closure with the ADO II is a safe and effective method. The advantages of using this device are smaller sheath sizes and softer shape of the device, however the use of this device is discutable in small patients with small aortic isthmus and short duct.

#### P-122

##### **Clinical, echocardiographic and histopathologic findings in nine patients with surgically explanted ASD/PFO devices: Do we know enough about the healing process in humans?**

Vögt M. (1), Kühn A. (1), Hörer J. (2), Schreiber Ch. (2), Schneider H. (3), Foth R. (3), Hess J. (1), Sigler M. (3)

(1) Department of Pediatric Cardiology and Congenital Heart Disease, German Heart Center Munich, Technical University, Germany; (2)

Department of Cardiovascular Surgery, German Heart Center Munich, Technical University, Germany; (3) Department of Pediatric Cardiology and Pediatric Intensive Care Medicine, Georg-August University,

Goettingen, Germany

**Background:** Atrial septal defects (ASD) and persistent foramen ovale (PFO) are managed in increasing numbers by catheter interventions as an attractive alternative to surgery. Early complications have been described in clinical series whereas late complications are rare. No series are reported with clinical, echocardiographic and histological data.

**Methods and results:** We collected clinical, echocardiographic, and histological data of nine patients with surgically explanted devices. Occlusion devices were explanted after a mean interval of  $3.4 \pm 2.4$  y (range 0.9–8.3). Indications were recurrent thromboembolic events in five, residual shunt/dislocation in three, and growing mass on echocardiography despite oral anticoagulation in one patient. Two patients suffered potentially life threatening events due to coronary embolism. One of them had to be resuscitated due to ventricular fibrillation. Histologically, residues of superficial thrombus formation could be demonstrated in two of the devices. In another patient, hyperplastic tissue formation was related to a local inflammatory process but not to a thrombus as suspected by echocardiography.

**Conclusion:** Late complications after device implantation may occur up to eight years after device implantation and may be potentially life threatening. Echocardiographic controls should be prolonged beyond the first year after implantation and every explanted device should be histologically worked up in an experienced center. Up to now, the mechanisms of late thrombogenesis are not fully understood.

## P-123

**The Italian Society of Pediatric Cardiology Registry on transcatheter Melody pulmonary valve implantation**

Butera G. (1), Milanese O. (2), Piazza L. (1), Donti A. (3), Spadoni I. (4), Giusti S (4), Formigari R. (3), Ricci M. (1), Pongiglione G. (5), Papa M. (6), Pangrazi A. (7), Carminati M. (1)  
 Policlinico San Donato IRCCS, Università Padova, Policlinico San Donato IRCCS (1); Università Padova (2); Università Bologna (3); Ospedale Pasquinucci Massa (4); Ospedale Bambino Gesù Roma (5); Ospedale San Raffaele (6); Ospedali Riuniti Marche (7), Italy

**Aims:** To analyse indications, results, follow-up data of patients treated in Italy by using the Melody Medtronic valve.

**Methods:** Observational, multi-centric survey by using a web-based database.

**Results:** Forty-six patients were included in the registry (58% female, median age 25 years (range 11–63 years); median weight 60 kg (range 20–140 kg)). Sixty-eight percent were in NYHA class I-II. Patients had a history of a median 3 previous surgeries (range 1–5).

Original diagnosis was tronco-conal disease in 58% (tetralogy of Fallot in 18%, truncus arteriosus in 10%, pulmonary atresia with VSD in 30%), Ross surgery in 18%, TGA+VSD in 10%, others in 18%. Right ventricular outflow tract was reconstructed with homograft in 48%, an Hancock conduit in 30% and other type in 22%. Indication to valve implantation was pure stenosis in 30%, pure regurgitation in 21%, association of stenosis and regurgitation in 49%. In all except two, a femoral approach was used. Pre-stenting was performed in 80% of cases. Ensemble delivery system was used. The system was post-dilated in 60% of the procedures. Median procedure time was 170 minutes (range 85–360). No significant regurgitation was recorded after procedure while the trans-pulmonary gradient reduced significantly. Early complications occurred in 6 subjects (13%): one retro-peritoneal bleeding which required blood transfusion, one femoral bleeding requiring blood transfusion, two surgeries of the femoral artery, and minor complications in two subjects. No early death occurred. Hospital stay was a median of 5 days (range 3–30 days). Mortality: at a median follow-up of 6 months (range 0–36 months) two patients died due to underlying disease. Complications at follow-up were reported in 5 patients: one subject needed EEC due to atrial fibrillation, and 4 experienced fractures of the stent. In two patients these were minor fractures while in the other two subjects a second Melody valve was required. Pre-stenting was a major protective factor for the occurrence of stent fracture.

**Conclusion:** Early results of the SICP registry shows that the procedure is safe and successful. A major concern is related to the occurrence of stent fracture. Longer follow-up and larger series are needed.

## P-124

**Aortic coarctation associated with wall aneurysm: The role of covered stent**

Butera G., Heles M., Shridar A., Piazza L., Negura D., Chessa M., Micheletti A., Arcidiacono C., Abella R., Giamberti A., Pomè G., Bussadori C., Frigiola A., Carminati M.  
 Policlinico San Donato IRCCS -Italy

**Background:** Aortic wall aneurysm associated with aortic coarctation carries a significant risk of rupture and lethal outcome. Surgery is associated with significant in-hospital mortality and morbidity (paraplegia, injury to the central nervous system, bleeding). Trans-catheter dilation of aortic coarctation carries the risk of aneurysm or rupture.

**Objectives:** To evaluate safety and efficacy of the covered stent in treatment of native coarctation and re-coarctation of the aorta associated with aneurysm.

**Methods:** Between January 2005 and December 2009, 160 patients with aortic coarctation were treated in our unit by using percutaneous stent implantation. Twelve subjects had aortic coarctation associated with aneurysm. Patients' distribution was as follows: native coarctations (n=3), previous coarctation surgery (n=3), previous bare stent implantation (n=3), previous balloon dilatation (n=3).

All subjects had systemic arterial hypertension. Procedures were performed under general anaesthesia and oro-tracheal intubation. Expanded-PTFE covered Cheatham-Platinum stents were used. During follow-up all subjects underwent physical examination and echocardiography at 1,6,12 months and yearly thereafter. A EKG effort test was performed at 6 and 12 months and yearly thereafter. A 64-slices spiral CT scan was performed at 6 months after procedure.

**Results:** Thirteen covered stents were placed in 12 patients (median age 15 years (range 9–42 years)). Successful device deployment was achieved in all patients. In all subjects gradient reduced significantly from baseline median value of 48 mmHg (range 35–68 mmHg) to a median post-procedural gradient of 5 mmHg (range 0–10 mmHg) (p<0.01). Aortic diameter increased from a median value of 3 mm (range 2–6 mm) to a median of 12 mm (10–14 mm) (p<0.01).

No early complications occurred. During a median follow-up of 12 months (range 1–48 months) no problems occurred. Due to somatic growth, three subjects underwent uneventful re-dilatation of the covered stent 24 months after the first procedure. Four out of 12 subjects were still under anti-hypertensive drugs at latest follow-up.

**Conclusions:** These results show that percutaneous implantation of covered stents is safe and effective in the treatment of aortic coarctation associated with wall aneurysm.

## P-125

**Italian Patent Foramen Ovale Survey (I.P.O.S.): early results**

Butera G. (1), Sangiorgi G. (2), Lucarelli K. (3), Ussia G. (4), Aprile A. (2), Spadoni I. (5), Onorato E. (6), Caputi L. (7), Benassi A. (8), Chessa M. (1), Ramondo A. (9), Anzola G. (6), Zanchetta M. (10), Bortone A. (11), Santoro G. (12), Vignati G. (13), Rigatelli G. (14), Presbitero P. (15), Pierli C. (16), Varotto L. (17), Papa M. (18), A.Colombo (18) Carminati M. (1)  
 Policlinico San Donato IRCCS (1); Policlinico di Modena (2); Miulli, Acquaviva delle Fonti (3); Università di Catania (4); Massa (5); Brescia (6); Istituto Neurologico Besta (7); Hesperia Hospital Modena (8); Università di Padova (9); Ospedale di Cittadella (10); Università di Bari (11); Firenze (12); Ospedale Niguarda (13); Ospedale Rovigo (14); Humanitas Rozzano (15); Università di Siena (16); Ospedale Vicenza (17); Ospedale San Raffaele (18) – Italy

**Background and aims:** PFO percutaneous closure is gaining wide acceptance. Aims of the study were: (a) to analyse clinical practice regarding PFO closure in Italy; (b) to study indications, devices used, results of percutaneous PFO closure; (c) to evaluate follow-up of large series of patients treated by percutaneous closure.

**Methods and patients:** IPOS is a prospective, observational, multi-centric survey that uses a web-based database. The survey lasted 12 months. Follow-up evaluations will be recorded yearly up to 3 years after procedure.

Between November 2007 and October 2008, 50 centres accepted to participate. One thousand and ninety patients were

included in the registry (58% females; median age 45 years (range 5–75 years). An aneurysm of the interatrial septum was associated in 41% of patients.

The large part of subjects were treated due to a previous history of TIA/Stroke (85% of pts). Fifteen percent of subjects had an associated migraine with aura. Procedures were monitored by using trans-esophageal echocardiography and fluoroscopy in 70% of subjects while 30% were monitored by using intracardiac echocardiography. Procedures were performed under general anesthesia in 54% and under local anesthesia/conscious sedation in 46%. Devices used for PFO closure were PFO/Cribriform Amplatzer devices in 68%, Cardiastar devices in 8%, Starflex/Biostar in 7%, Premere in 11%, Helex in 2%, other in 4%.

Early complications occurred in 27 subjects (2.5%): 7 experienced transient atrial fibrillation, 1 had pericardial effusion, 2 needed vascular surgery due to the occurrence of femoral artero-venous fistula, 1 developed blood effusion due to oro-tracheal intubation, in 2 subjects a device was removed due to device malposition, 1 patient needed surgery due to device malposition, other minor complications in 13 subjects.

**Conclusions:** Early results of the IPOS study shows that percutaneous PFO closure is a safe procedure. Follow-ups are under way.

#### P-126

##### **Five-year Follow-up After Balloon Pulmonary And Aortic Valvuloplasty In Benghazi Cardiac Center**

*Feituri.R (1),(2),(3), Rahuma.S (1),(2),(3), Gobhaa.N (1),(2),(3), Kasmi.A (1); Benghazi Cardiac Center, Benghazi,Libya (1), El Fateh Children Hospital, Benghazi, Libya (2); Garyounis University, Benghazi,Libya (3)*

**Objective:** To assess results 5 years after balloon pulmonary and aortic valvuloplasty in Benghazi Cardiac Center.

**Background:** Since the technique of balloon pulmonary and aortic valvuloplasty was first reported in 1982, it has become the treatment of choice for pulmonary and aortic valve stenosis.

**Methods:** Between January 2004 and February 2009 we performed percutaneous pulmonary valvuloplasty with a balloon catheter in 39 patients 8 months to 37 years of age 20 males and 19 females, and aortic valvuloplasty in 6 patients 4 months to 13 years of age 3 males and 3 females with using of pacing in right ventricle. follow up studies were performed 0.3 to 5.2 years after the procedure for pulmonary stenosis and 0.1 to 4.1 years after the procedure for aortic stenosis by echocardiography in all patients and by cardiac catheterization and angio for one patient with restenosis of aortic valve.

**Results:** short term result after balloon pulmonary valvuloplasty the right ventricular systolic pressure decreased from  $100 \pm 60$  mmHg before valvuloplasty to  $60 \pm 39$  mmHg after balloon dilatation. After aortic valvuloplasty the pressure gradient across aortic valve decreased from  $95 \pm 45$  before valvuloplasty to  $50 \pm 20$  after valvuloplasty. Follow up: the pressure across the pulmonary valve and the aortic valve significantly decreased in all patients with pulmonary and aortic stenosis except one of 39 patient with pulmonary stenosis post operative Tetralogy of fallout with right pulmonary branch stenosis who needs to be operated, and one of 6 patients with aortic stenosis with thick bicuspid aortic valve develop restenosis after 4.2 years and developed syncopal attack we performed 2nd aortic valvuloplasty the pressure gradient across the aortic valve decreased from 50 mmHg to 35 mmHg after balloon dilatation. Mild incompetence of pulmonary valve was noted in 9 of 39 patients, and mild incompetence of aortic valve in 2 of 6 patients after aortic

valvuloplasty, but it had disappeared at follow up in almost all of them. **Conclusions :** patients with congenital pulmonary and aortic stenosis can be treated with percutaneous balloon valvuloplasty with excellent short and long term results.

#### P-127

##### **A comparative study on validation of a novel device for percutaneous closure of atrial septal defects: Figulla ASD Occluder versus Amplatzer Septal Occluder**

*Pac FA., Polat T.B., Oflaz M.B., Topaloglu S., Balli S., Cetin I.I., Kibar E., Ece I.*

*Clinic of Pediatric Cardiology, Turkiye Yuksek Ihtisas Education and Research Hospital, Ankara, Turkey*

**Objectives:** Occlutech Figulla ASD Occluder (FSO) was recently introduced as an alternative to Amplatzer Septal Occluder (ASO) for treatment of small to very large atrial septal defects. Therefore, a comparison of their use in the closure of ASDs is needed.

**Methods:** Between December 2005 and December 2009, 95 patients diagnosed as secundum atrial septal defects underwent transcatheter closure. The FSO device was used in 53 patients, and the ASO was used in 42.

**Results:** There were no significant differences among the groups regarding age, weight, sex ratio, mean pulmonary arterial pressure or Qp/Qs ratio. Stretch size of the defect, device size and device left disc size, procedure and fluoroscopy time were also similar between the groups. However, the difference between device waist size and stretched diameter of the defect was significantly higher, and device delivery sheath was significantly larger in FSO group and device left disc size was significantly lower in the FSO group. In all subjects the residual shunt was small to trivial during follow-up and the reduction in prevalence of residual shunt with time was similar in both groups ( $P = 0.68$ ). We found no differences in complication rate between the 2 devices, however device embolization to the pulmonary bifurcation in one patient was recorded as major complication in FSO device group.

**Conclusions:** Both devices are clinically safe and effective in ASD closure. FSO device has similar outcomes when compared to ASO device. However, difficulties in selecting the correct device size in larger defects and larger venous sheath requirement are needed to be evaluated in further studies.

#### P-128

##### **Atrial Septal Defect Occlusion by the Transcatheter Patch: Can Patch Release Time Influence the Results?**

*Sideris E., Zanjani K., Zeinalloo A., Kalachanis M., Coulson J., Sideris B., Pursanov M., Kumbasar D.*

*Athenian Institute of Pediatric Cardiology, Athens, Greece*

The transcatheter patch (TP) is a wireless bio-absorbable device originally released in 48 hours (48 HR) with good results despite the inconvenient release; accelerated patch release was later performed using surgical adhesives (SA) with 45 min–3 hours release time. Recently, an immediate release patch model (IRP) was introduced. Purpose of this study was to compare the results of the ASD occlusion in the three TP release groups.

**Methods:** The Transcatheter patch is made by polyurethane foam and is balloon supported until it is attached by fibrin (48HR group) or by polyethylene glycole (SA group); withdrawal of the supportive balloon catheter is necessary for TP release. In the IRP group the balloon/patch was immediately released after occlusion of the ASD; the catheter shaft was withdrawn and the

device was immobilized in the groin by a bio-absorbable thread. The results of successfully implanted patients from the international registry were reviewed. They included 74 patients in the 48HR group, 9 patients in the SA group and 12 patients in the IRP group.

**Results:** The mean defect size was 25 mm in the 48HR group, 18 in the SA group and 23 in the IRP group. The mean age was 30 years, 33 years and 17 years respectively. The full occlusion rate on discharge was 66% in the 48HR group, 78% in the SA group and 100% in the IRP group. The effective occlusion (full occlusion+trivial shunt) was 88%, 100% and 100% respectively. The effective occlusion improved further on latest follow-up to 96% (48HR), 100% (SA) and 100% (IRP). No serious complications were seen in any of the groups, although minor problems including temporary attachment problems, balloon deflation and echocardiographic strands were noticed in the 48HR group and unreliable patch attachment in large ASDs in the SA group. No minor complications have been seen in the IRP group so far.

**Conclusions:** The IRP is not only conveniently placed but it appears to have better occlusion rates and no minor complications in comparison to the other release methods. Careful recording of the results in the international registry is necessary for future assessment.

#### P-129

##### **Immediate and long-term results of balloon aortic valvuloplasty of congenital valvular aortic stenosis in 98 patients**

Gamillscheg A. (1), Mileder L.P. (1), Knez I. (2), Koestenberger M. (1), Heinzl B. (1), Fandl A. (1), Finding K. (1), Fritsch P. (1), Nagel B. (1)

Department of Pediatrics, Division of Pediatric Cardiology (1), Department of Surgery, Division of Cardio-Thoracic Surgery (2), Medical University Graz, Austria

**Introduction:** Long-term follow-up data of patients with congenital valvular aortic stenosis after balloon valvuloplasty are limited.

**Methods:** Retrospective follow-up study of all 98 patients at the median age of 52.4 months (0–31.8 years, 24 patients <1 month) in whom BAV was attempted at our institution between 3/1986 and 4/2009. Immediate success was defined as a residual systolic peak-to-peak gradient <50 mmHg or a gradient reduction of  $\geq 50\%$  after BAV.

**Results:** In two cases BAV had to be abandoned because of technical problems. Two neonates with severely compromised left ventricular function died during the procedure. In the remaining 94 patients the peak-to-peak systolic pressure gradient decreased from a mean of  $57.2 \pm 25.6$  mmHg (25–140) to  $19.2 \pm 13.5$  mmHg (0–60) corresponding to a mean gradient reduction of  $65.5 \pm 22.7\%$  after BAV ( $p < 0.001$ ). Only 4/94 patients (4.2%) had a residual pressure gradient  $\geq 50$  mmHg. After BAV aortic regurgitation (AR) increased in 27 (28.7%) patients (19  $\times$  grade 1, 7  $\times$  grade 2, 1  $\times$  grade 3). In 88/94 patients (93.6%) follow-up was possible with a mean of  $91.2 \pm 67.4$  (0.07–250.8) months. The mean systolic Doppler gradient at latest follow-up or before re-intervention was  $46.4 \pm 24.1$  (15–120) mmHg, with 32 patients (36.4%) having gradients  $\geq 50$  mmHg. AR grades 0, 1, 2, 3 and 4 were detected in 19 (21.5%), 33 (37.5%), 21 (23.9%), 14 (15.9%) and 1 (1.1%) patients, respectively. After the median interval of 55.1 (0.1–168.6) months 35/88 patients (39.8%) required re-BAV (n = 10) or surgery (n = 25) including valvulotomy (n = 2), Ross procedure (n = 11), valve replacement (n = 10) and Norwood

procedure (n = 2) mainly due to re-stenosis (n = 11) or AR (n = 10). Twelve of these 35 patients (34.3%) had undergone BAV as neonates. During follow-up 7/88 (7.9%) patients (two patients with neonatal BAV) died at a median interval of 47.2 (0.7–213.5) months after BAV (2 of non-cardiac causes).

**Conclusions:** BAV is an effective and safe first line procedure of congenital valvular aortic stenosis. However, in the long-term follow-up the incidence of re-interventions in particular in neonatal BAV has to be noted.

#### P-130

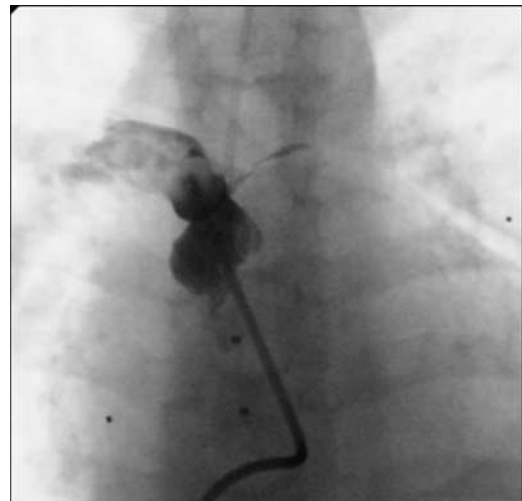
##### **Reduction of right ventricular afterload improves right ventricular function**

Kaestner M., Schneider K., Toussaint-Götz N., Wiebe W., Zartner P., Schneider M.B.E.

German Paediatric Heart Centre, St. Augustin, Germany

**Objectives:** To evaluate the effect of different strategies for the treatment of peripheral pulmonary artery stenoses (pPS) on the right ventricular function.

**Methods:** The study was designed as prospective multi centre clinical treatment trial as part of the competence network for congenital heart disease. Patients were followed with a standardized protocol for echocardiography, electrocardiography (ECG), Holter-ECG, exercise test, magnetic resonance imaging and a questionnaire for quality of life, at time of inclusion (visit 1) and one year later (visit 3). Visit 2 was planned with echocardiography and ECG three month after treatment. Different treatment strategies as balloon angioplasty (BAP) or stent implantation and surgical approaches were allowed.



**Results:** 53 patients, 26 female and 27 male, aged 6 to 38 years from five paediatric heart centres were analysed. Most patients (n = 32) had malformations from the spectrum of tetralogy of Fallot, 8 had common arterial trunk, six patients had transposition of the great arteries and the remaining patients had miscellaneous disorders with pPS after surgical procedures. At visit 1 all patients had pPS in 87 locations. Treatment was BAP in 55 locations (63.2%), implantation of stents in 17 locations (19.5%) and surgery in 11 locations (12.6%) respectively. In 4 locations no treatment was necessary. At visit 3 in 15 patients 25 sites of pPS were treated with BAP in 16 locations and stent implantation in 9 respectively. Haemodynamic measurements before and after treatment were available in the interventional group only. These showed significant reduction of the systolic pressure ratio from the right and left ventricle before and after intervention with  $p = 0.004$  at visit 1 and  $p = 0.003$  at visit

3 respectively. The diameter of the treated vessels increased significantly ( $p < 0.001$ ). In all patients the stress tolerance test showed significant increase of the oxygen pulse, measured at visit 1 and visit 3 ( $p = 0.06$ ), which can be used as an indirect measure of stroke volume.

**Conclusions:** Catheter interventions and surgery are effective methods for reduction of right ventricular afterload. Reduction of right ventricular afterload improves the stroke volume. Therefore stress tolerance testing with measurement of oxygen pulse is an easy method for follow up.

#### P-131

##### Percutaneous aortic valvuloplasty in patients older than 1 month with congenital aortic valve stenosis: a good alternative to surgery.

Corzani A., Donti A., Formigari R., Bronzetti G., Balducci A., Fabi M., Testa G., Bonvicini M., Picchio F.M.  
Institute of Pediatric Cardiology, S.Orsola-Malpighi Polyclinic Hospital, Bologna, Italy

**Objectives:** Percutaneous aortic valvuloplasty is an effective means of treatment for congenital aortic valve stenosis. The aim of this study was to evaluate the immediate results of valvuloplasty, to analyze the medium to long term outcome and to assess its efficacy in preventing or postponing a new percutaneous valvuloplasty or aortic valve surgery.

**Methods:** We retrospectively analyzed the reports of 37 patients > 1 month (mean age of 6,3 years) who underwent aortic valvuloplasty for severe aortic stenosis. Associated congenital cardiac defects were present in 16% of the patients. The average time of follow up was  $5,07 \pm 4,23$  years. Particular attention was focused on occurrence and progression of aortic regurgitation.

**Results:** Hemodynamic gradient after aortic valvuloplasty decreased from 58,5 to 22,5 mmHg, with an average decrease of 61,5%. On echography, the maximum gradient decreased from 93,0 to 40,5 mmHg, with an average decrease of 56,5%; mean gradient decreased from 52,0 to 20,5 mmHg with an average decrease of 60,6%. 1 patient died because of sepsis 11 days after the procedure. At last follow up the average maximum and mean gradient on echo were 50,0 and 27,0 mmHg. 2 patients (5,4%) showed severe aortic regurgitation immediately after valvuloplasty and 4 (17,4%) at last follow up. A reintervention was needed in 8 patients (21,6%): a second valvuloplasty in 3 and aortic surgery in 5 patients for severe aortic regurgitation.

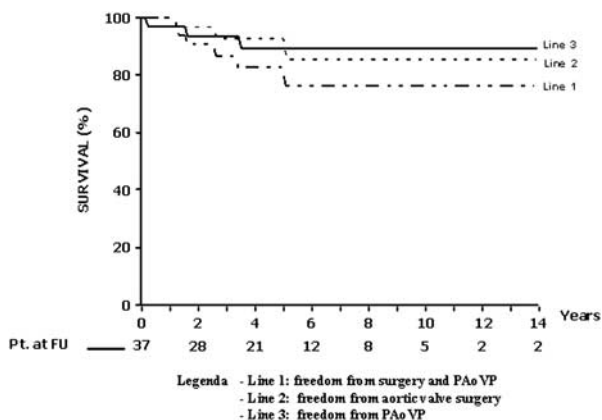


Fig. 1 – Freedom from different type of intervention on aortic valve during follow up after valvuloplasty

Survival after 14 years was 97,2%; freedom from aortic valve surgery was 85,5%, from a second valvuloplasty 89,5% and from any type of procedure 76,1%.

**Conclusions:** Percutaneous aortic valvuloplasty is a safe and effective treatment for congenital aortic stenosis in patients older than 1 month. Aortic regurgitation is the main concern in the follow up. Nonetheless, 14 years after valvuloplasty, over 75% of patients are free from any type of aortic valve reintervention.

#### P-132

##### Stent Expansion of Stretch Gore-Tex® Grafts in Children with Congenital Heart Lesions

Gewillig M., Brown S.C., Boshoff D.E., Heying R., Gorenflo M., Rega F., Eyskens B., Meyns B.  
University Hospitals Leuven, Belgium

**Objective:** To evaluate the efficacy and safety of expanding previously placed vascular shunt grafts beyond nominal value by means of stents.

**Methods:** Bench testing confirmed the expandability of 3.5 and 4.0 mm vascular Gore-Tex® stretch grafts. We attempted to stent-expand beyond nominal value 11 systemic to pulmonary artery shunts with diminished flow in 10 patients. Stents were selected to be at least 1 mm larger than the nominal diameter of the implanted graft.

**Results:** On the bench, the graft could be expanded up to 4.5 and 5.8 mm respectively. Fourteen stents were implanted in 11 stretch grafts a median of 18.9 (range: 0.9–62.1) months after shunt surgery. All shunts could be dilated beyond nominal value. There was a median increase in diameter of 3.5 mm (3.0–6.0) to 4.9 mm (3.5–7.7) from nominal to final stented diameter of the shunts with a median gain of 136% ( $p < 0.001$ ; 95% CI 0.9 to 1.5). Saturations improved from a median of 73% (62–82) to 87% (79–89) [ $p = 0.003$ ; 95% CI 6.5 to 20.1]. No complications were experienced during the procedures.

**Conclusion:** In our limited experience, stretch Gore-Tex® vascular grafts can be safely expanded beyond nominal diameters using high pressure vascular stents. This leads to improvement in saturation and pulmonary blood flow. It allows the clinician to tailor pulmonary flow in relation to pulmonary artery size and somatic growth, ensuring best possible timing for the next surgical procedure.

#### P-133

##### Bailout stenting for critical coarctation in premature/critical/complex/early re-coarcted neonates

Gewillig M., Boshoff D.E., Heying R., Eyskens B., Rega F., Meyns B., Gorenflo M.  
University Hospitals Leuven, Belgium

**Aims:** Surgical repair of critical coarctation can be problematic in premature, critical, complex or early postoperative neonates. We aimed to review our experience with stent implantation to defer urgent surgery to an elective time (early or late).

**Methods:** 15 neonates with severe aortic coarctation: 5 premature-hypotrophic (1400–2000 g), 6 critical & complex cardiac malformation, 4 early (1 day [0–2 days]; median [range]) after surgical coarctectomy or complex arch reconstruction. Bare coronary stents (diameter 4.0 [3.5–5.0] mm; length 10 [8–16] mm) were used. Stents were removed surgically depending on clinical needs.

**Results:** Adequate aortic flow was obtained in 15 patients. The femoral artery was preserved in 13/15 patients. 2 deaths occurred before stent removal and were non-procedure related. In patients with simple stented coarctation the stent was removed after

2.8 (0.2–5.0) months. In complex cardiac malformation, stents were finally removed 3.0 (0.2–78) months after implantation. Surgical technique: simple coarctectomy end-to-end in 8, extensive arch patch reconstruction in 4. 1 patient is awaiting stent removal. The final maximum systolic velocity (cw-Doppler) across the aortic arch was 1.7 [1.2–2.5] m/sec.

**Conclusion:** In premature/critical/complex neonates with severe coarctation, bailout stenting followed by early or late surgical coarctectomy appears a promising concept with a very competitive final result.

#### P-134

##### Stent Implantation in Newborns and Infants:

###### A Current Reality

*Ferreira Martins J.D., Costa G., Gouveia S., Parames F., Freitas I., Rebelo M., Trigo C., Pinto F.*

*Hospital de Santa Marta, Lisbon, Portugal*

**Introduction:** Since introduction of Rashkind septostomy that percutaneous intervention assumed a central role in the management of congenital heart disease in newborns and infants. In these, stent implantation is limited by iatrogenic “stenosis” that its maximum diameter imposes, the need of vascular access/line of large dimension and technical difficulties related with the size of patients and hardware availability. However, progress of technology and knowledge has extended the offer of these procedures to a selected sub-group of newborns and infants.

**Objectives:** To report the experience of our center in stent implantation in newborns and infants with congenital heart disease.

**Methods:** Retrospective evaluation of clinical processes of newborns and infants that have been submitted to stent implantation, with special focus in catheterism data.

**Results:** In the past 10 years, 12 stents have been implanted in 9 patients (4 male), with a median age of 23 [10–142] days. Diagnoses were hypoplastic left heart syndrome (4), cyanotic congenital heart disease palliated with systemic pulmonary anastomosis (3), pulmonary atresia with intact ventricular septum (1) and repaired truncus arteriosus. The vascular access was percutaneous femoral vein (2), or artery (5) or surgical access to pulmonary trunk (5). Stent implantation performed in all cases as palliative procedure was in the ductus arteriosus (6), in systemic pulmonary shunt (4) or pulmonary artery branches (1). Stents had a median diameter of 6.2 [4–8] mm. All procedures were under general anaesthesia, and were immediately successful. The complications were early embolization with surgical removal (1), early hypofunctioning (2), transient arrhythmias (3) and need of elective transfusion (3). With a median follow-up of 23.1 [0.6–143] months, eight patients are alive and without significant residual lesions, and have been submitted to ulterior surgery that included stent removal or ligation. There was no significant stent stenosis.

**Conclusions:** Stent implantation is a useful, effective and safe palliative option in selected groups of newborns and infants. Technology evolution, careful selection of patients, anticipated planning and multidisciplinary approach allowed to assure the success of interventions performed in our center.

#### P-135

##### Stenting of native and re-coarctation with a new chromium-cobalt stent (Andra Stent®) in children and young adults

*Beck C., Laser T.K., Kececioğlu D., Haas N.A.*

*Clinic for Congenital Heart Defects, Heart and Diabetes Center NRW, Bad Oeynhausen, Germany*

**Introduction:** Stenting of native and re-coarctation after surgical repair in children and young adults is a well established procedure with excellent outcome. Theoretical stents used for this procedure should be suitable to be delivered in small children and re-expandable to adult size. We present our experience in stenting these lesions with a new laser cut chromium-cobalt stent with a semi-open cell design (Andra Stent®, Andramed, Reutlingen, Germany).

**Methods:** Between October 2008 and November 2009 we implanted 14 Andra Stents® in 14 patients. Patient age ranged from 4 to 17 years (mean 10.6 years) and weight from 17 kg to 95 kg (mean 40.9 kg). The stent was delivered through a 9 Fr to 14 Fr sheath (mean 10 Fr) using a high pressure balloon. In 11 of 14 patients the stent was re-dilated with a larger balloon with a final balloon diameter from 12 mm to 25 mm (mean 17.3 mm).

**Results:** The mean gradient could be reduced from 47 mmHg to 5 mmHg (range 0 to 15 mmHg) after implantation. The stent shorting ranged from 1.5% to 15.4% (mean 8.8%). No complications did occur during the procedures.

**Conclusion:** In our limited experience the Andra Stent® produced an excellent clinical result with the added benefits of the chromium-cobalt technology for excellent visibility on MRI follow up. The semi-open cell design of the Andra stent® seems to be beneficial when placing the stent in an angulated part of the aorta.

#### P-136

##### Monitoring unfractionated heparin during cardiac catheterization in children – results from the HEARTCAT study

*Kitzmüller E. (1), Hanslik A. (1), Karapetian H. (1), Salzer-Muhar U. (1), Michel-Behnke I. (1), Male C. (1)*

*Division of Pediatric Cardiology, Department of Pediatrics, Medical University of Vienna, Austria (1)*

**Introduction:** Monitoring unfractionated heparin (UFH) in children is problematic because of variable dose-response.

**Objectives:** To assess (i) the relationship between UFH dose and UFH plasma levels as tested by various assays and (ii) the correlation between these assays.

**Methods:** Randomized controlled trial comparing two UFH protocols (high dose: 100 u/kg body weight bolus, followed by continuous infusion of 20 u/kg/hour; low dose: 50 u/kg bolus) for prevention of thrombosis during elective cardiac catheterization. Children with congenital heart disease at a tertiary care pediatric cardiology centre. Clinical outcome were vascular complications associated with cardiac catheterization assessed by vascular ultrasound. Blood samples were taken before and 30, 60 and 90 minutes after UFH administration. Assays: anti-Xa (aXa), activated partial thromboplastin time (APTT), activated clotting time (ACT), thrombin generation (TG).

**Results:** Interim laboratory analysis of 65 patients. Thirty-two patients were in high dose group, 33 patients in low UFH dose. All four assays showed significant discrimination between UFH dose groups which was best for aXa, followed by ACT, TG, and APTT. Correlation of aXa versus ACT and TG was fair but aXa versus APTT was poor. Low dose heparinization reached therapeutic levels, whereas high dose heparinization led to supratherapeutic levels of anticoagulation.

**Conclusion:** In children during elective cardiac catheterization, vascular complications were less frequent than anticipated. High and moderately high doses of UFH were well discriminated using aXa, APTT, ACT and TG. However, correlation between the results of these assays moderate to poor.

## P-137

**Outcome of device closure of ASD in intermediate and long term**

Sadiq M. (1), Kazmi T. (1), Rehman A.U. (1), Latif E. (1), Hyder N. (1), Qureshi S.A. (2)

The Children's Hospital and Punjab Institute of Cardiology, Lahore, Pakistan (1); Evelina Children's hospital, Guy's NHS Trust, London UK (2)

**Background:** Device closure of Secundum ASD is an accepted mode of treatment in selected patients. There are however, concerns over the long term outcome especially complications like erosions and AR.

**Aim:** To assess the intermediate and long term outcome of device closure of ASD with special reference to complications.

**Methods:** Between October 1999 and March 2009, 204 patients underwent transcatheter closure of ASD with the Amplatzer septal occluder. Ages were 1.5–55 (median 8) years and weight 10–97 (median 18) kg. Four patients had multiple defects. Patients above 10 years of age underwent TOE before the procedure. Follow up including ECG and echocardiogram was performed at 1, 3, 6, and 12 months and thereafter every year. Holter recording was done in selected patients.

**Results:** Device closure of ASD was done successfully in 200/204 patients (98%). In 2 patients, devices could not be implanted despite multiple attempts. The device embolized immediately after implantation in 2 patients and a larger device was implanted in the same setting while in another 2 device embolized in the next 24 hours and both were taken for surgical removal and closure of ASD. The procedure time was 20–167 (median 46) minutes and fluoroscopy time was 2.5–48 (median 10) minutes. The immediate procedure related major complications were device embolization (4 pts), pericardial effusion (1 pt) and 2:1 heart block (1 pt). At a mean follow up of 4.9 (90 days to 9.6, median 5.3) years complete closure was documented in all except 2 patients with small residual shunt. There were no late embolizations. Two patients have developed mild AR (1%) and there were no erosions or thromboembolizations. The child with 2:1 heart block has not needed a pacemaker. Atrial fibrillation occurred in 5 adult patients (2.5% of all). Four responded to antiarrhythmic treatment which could be stopped, while one needed cardioversion and is on treatment.

**Conclusions:** Device closure of Secundum ASD using Amplatzer septal occluder is safe and effective in intermediate and long term follow up. The risk of development of AR is 1%. Atrial fibrillation occurs in adults (2.5%) but majority respond to medical treatment.

## P-138

**Our Experience of ADO2 Device Occlusion of PDA**

Narayanan A., Mehta C. (1), Ladusans E., Gladman G., Misra S., Peart I.

Alder Hey and Royal Manchester (1) Children's Hospital NHS Trust Liverpool United Kingdom

**Background:** The Amplatzer Duct Occluder II (ADOII) (AGA Corporation, MN) is a new self-expanding nitinol device for occlusion of a patent ductus arteriosus (PDA). This device is designed for ease of placement and higher rates of occlusion for all sizes and types of PDAs. Clinical results using the ADOII demonstrated low rates of embolisation and extremely high occlusion rates with a single device and procedure.

**Objective:** To evaluate our experience of using the new ADO II device for PDA closure and document the immediate/early closure rate, complications and device behavior during implantation.

**Method:** Retrospective cohort study from July 2008 to December 2009. Review and evaluation of data of all patients on whom we attempted closure of PDA using the new device.

**Results:** Total 34

Demographics	Median	Range	Ductal Morphology <sup>1</sup>
Age	17.5 months	6 months–10.8 yrs	A (47%)
Weight	10.2 Kg	5.2–39 kg	B (23%)
Narrowest ductal diameter	2.5 mm	1.5 mm–3.6 mm	C (17%)
Length of duct	6 mm	4 mm to 15 mm	D (03%)
Diameter of aortic ampulla	7 mm	4 mm–13 mm	E (10%)
Procedure time	49 min	36–104 mins	
Fluoroscopy time	7.3 min	4.7–19.2 mins	

Devices used were 14–4/4, 9–3/4, 7–5/4, I – 6/4 and 2 4/6, 1 8/4. All the devices implanted through venous side. Follow-up echocardiography was done on D1 and 4–6 weeks post implantation. All except two implantations were technically successful with immediate complete angiographic closure in 24, trivial contrast flow in 8 patients and mild flow in one patient. On 32 patients, echocardiography the next day and on follow up scan at 6 weeks showed no residual shunts. One patient showed moderate flow with device embolisation the next day. In one patient, the device did not articulate well and hence was not deployed. There were no procedure-related complications.

**Conclusion:** ADO2 is a safe and effective device for PDA occlusion and has a high early closure rate with fewer complications.

Ref: 1. Krichenko A, Benson LN, Burrows P, et al.

## P-139

**Balloon angioplasty of branch pulmonary arteries following arterial switch operation for transposition of great arteries: Is it effective?**

Shastri A, Naqvi N, Maiya S, Magee AG  
Royal Brompton Hospital, London, UK.

**Objectives:** To investigate the incidence of significant branch pulmonary artery (PA) stenosis after arterial switch operation (ASO) and assess the results of balloon angioplasty and stenting of branch PAs.

**Methods:** Single institution retrospective study over 11 years (1998–present). Arterial switch procedure was performed in 120 patients. Branch PA stenosis was detected by echocardiography. Success was identified by: 1) Immediate response to balloon dilatation assessed via analysis of angiograms in a standardised method by two experienced paediatric cardiologists: the criterion of successful dilatation was a  $\geq 50\%$  increase in diameter. 2) Clinical progress and echocardiography findings.

**Results:** At follow up 28 children had echocardiographic evidence of branch PA stenosis which was mild in 11. Seventeen had significant branch PA stenosis (Doppler gradient  $> 45$  mm Hg), evident within 2 months of ASO in 82% of cases. All 17 underwent catheterisation with an intention to treat. Two had surgery (1 supravalvar pulmonary stenosis, 1 supravalvar aortic stenosis). Fifteen underwent a total of 20 balloon angioplasties. The median age at dilatation was 12 months (range 2–75 months). Six had isolated Left PA (LPA) stenosis, 1 had isolated Right PA (RPA) stenosis; 8 had both. Balloon angioplasty alone was successful in 47% (7/15). Comparing to pre-dilatation diameter, RPA size increased by a median of 36% (range 15–92%) and LPA by median of 27% (0–208%). Stenting was required in 27% due to significant re-stenosis. (4/15, all LPA). There were no catheter related complications. Four patients required surgical revision.



**Conclusions:** Success of balloon angioplasty for significant PA stenosis after ASO is modest but should be first line treatment given the potential benefits and low complication rate. Importantly, surgery can be avoided in 74% of patients.

#### P-140

##### **Electrocardiographic and Rhythm Changes Following Percutaneous Atrial Septal Defect Device Closure**

Donald J. Hagler, Jonathan N. Johnson, Michelle L. Marquardt, Michael J. Ackerman, Samuel J. Asirvatham, Guy S. Reeder, Allison K. Cabalka, Frank Cetta.  
Mayo Clinic Rochester, MN USA

**Introduction:** Percutaneous device closure of atrial septal defect (ASD) has become the standard of care. Electrocardiogram (ECG) anomalies associated with device placement have been reported. However, a large cohort comparison of pre- and post-procedure ECGs in these patients has not been reported.

**Methods:** We reviewed records for all patients who underwent percutaneous device closure of an ASD at our institution from 1999–2008. Pre-procedure ECG and Holter studies were compared to those obtained on the day after device placement and >2 months after placement. The PR, QRS, and QT intervals were measured as well as P wave duration, height, and PR segment (end of P wave to start of QRS). All measured time intervals were indexed to heart rate.

**Results:** A total of 341 patients underwent ASD device closure. Both pre- and post-procedural ECGs were available in 223 patients (142 females, mean age  $43 \pm 21$  years, range 1–83 years). Median device size was 20 mm (range 8–38 mm). There were no differences between pre- and post-procedure measurements of QRS interval, PR interval, P-wave duration, or QTc. The PR interval index was longer in post-procedure ECG (average  $2.59 \pm 0.8$ ) compared to pre-procedure ECG ( $2.42 \pm 0.77$ ,  $p < 0.04$ ). The P-wave duration index also was longer in post-procedure ECG ( $1.54 \pm 0.47$ ) compared to pre-procedure ( $1.41 \pm 0.40$ ). Only 9 patients (4%) developed arrhythmias within 4 months of device placement, of whom 8 had atrial fibrillation/flutter. One patient with repaired double-outlet right ventricle (DORV) developed 2:1 heart block 2 months post-device closure. The device size and number of devices placed in each patient were not statistically associated with risk of post-procedure arrhythmias.

**Conclusion:** We report a low (4%) risk of post-procedure arrhythmias after device closure of ASDs. Clinically significant heart block occurred in only 1 patient that had DORV. There appears to be relative lengthening of the PR interval at higher heart rates in some patients of no recognized clinical significance. This finding may require further elucidation.

#### P-141

##### **The Role of Real-Time Three-Dimensional Transoesophageal Echocardiography in Guiding Device Closure of Atrial Septal Defect in Children**

Chen F.L. (1, 2), Kuo S.M. (3), Liao C.P. (1), Weng C.Y. (1)  
(1) Division of Pediatric Cardiology, Department of Pediatrics, Chung Shan Medical University Hospital, Taichung, Taiwan; (2) Institute of Medicine, Chung Shan Medical University, Taichung, Taiwan; (3) Division of Cardiovascular Surgery, Chung Shan Medical University Hospital, Taichung, Taiwan

**Background and Objective:** Percutaneous closure of secundum ASD with Amplatzer septal occluder (ASO) is becoming a standard alternative to surgery. Accurate evaluations of the dynamic

geometry, location and spatial relationship of ASD are essential for successful ASD occlusion. By the advances in computer and transducer technologies, a novel matrix array three-dimensional (3D) TEE probe (X7-2t, Philips Medical Systems), has permitted real-time 3D image acquisition and display of the exact imaging of cardiac structures. Our aim was to evaluate the utility and feasibility of real-time 3-dimensional transesophageal echocardiography (RT3D-TEE) for guiding transcatheter closure of ASD with Amplatzer septal occluder (ASO) in children.

**Methods:** 22 patients were prospectively enrolled for ASD device closure under general anesthesia with fluoroscopy and RT3D-TEE (transducer X7-2t, Royal Philips Electronics, Andover, MA). A total of 30 patients under 2D-TEE guidance were used for comparison. The assessment of ASD structure and size before and during balloon stretching, their relationship to surrounding structures, and utility for guiding deployment of ASO were compared.

**Results:** All 22 interventions under RT3D-TEE guidance were successfully completed without complications or dislodgement of ASO. RT3D-TEE is more easy to 1) delineate ASD geometry and size for selection of appropriate ASO device size for ASD closure, 2) assist in spatial orientation and manipulation of the exchange guide wire and advancing catheter to pass through ASD, 3) without fluoroscopy, completely guide effective device localization, and 4) to confirm effective closure of ASD. Fluoroscopy time was significantly shortened from  $15.3 \pm 2.9$  minutes to  $1.2 \pm 0.4$  minutes. The maximal diameter of ASD under balloon stretching ( $21.5 \pm 7.3$  mm) correlated well with the size of ASO device ( $21.9 \pm 7.5$  mm) and better than ASD size before balloon stretching ( $19.0 \pm 5.6$ ) ( $r = 0.999$  vs  $r = 0.873$ , respectively).

**Conclusions:** RT3D-TEE can be feasible, useful to guide ASD closure by selecting appropriate size of ASO, especially for larger size of ASD. After a learning curve, transcatheter ASD closure only under 3D-TEE guidance may be performed at bedside as the procedure of Rashkind septostomy without the need of fluoroscopy.

#### P-142

##### **Five years follow-up of stenting of aortic coarctation**

Brzezińska-Rajszyś G., Zubrzycka M. (1), Rewers B. (1), Książek J. (1), Jagiełłowicz D. (2), Kościeszka A. (3), Kany A. (4)  
The Children's Memorial Health Institute, Warsaw, Poland  
Heart Catheterization Laboratory (1), Cardiology Department (2), Computed Tomography (3), Cardiosurgery Department (4)

**Background:** Stent implantation for treatment of coarctation of the aorta has been widely used for the last years.

**Aims:** The aim of the study was to analyse the single centre results of stent implantation in patients with native and postoperative aortic coarctation performed in single.

**Material/Methods:** Fifty-five patients with native (41pts) and postoperative (14pts) coarctation were treated with stent implantation. Fifty-six stents were implanted for primary treatment (7 Palmaz stents, 35 Cheatham-Platinum, 14 covered Cheatham-Platinum). Covered stents were primarily implanted in 3 patients after balloon angioplasty due to small aneurysms, in 2 due to coexistence of PDA, in 7 due to severe coarctation (one with Down and one with Turner syndrome).

**Results:** The mean patient age was  $13.4 \pm 3.08$  yrs (range 5–20, median 14 yrs). The diameter of coarctation increased from a mean  $5.2 \pm 2$  mm (range 2–11, median 5 mm) to  $16.2 \pm 2.4$  mm (range 12–22 mm, median 16 mm) ( $p < 0.001$ ). The systolic

gradient across the coarctation decreased from a mean 32.44 +/- 12.02 mmHg (range 10–84, med.30 mmHg) before to a mean of 2.78 +/- 4.94 mmHg (range 0–18, med.0) after the procedure ( $p < 0.000001$ ). CT angiography, performed in 54pts (98%) in the first year after procedure, confirmed good anatomical result of stent implantation. During mean follow-up period 4.95 +/- 2.59 yrs (range 0.6–11, med.5) 28 patients (51%) did not need antihypertensive treatment and all the other patients have better control of arterial hypertension on lower doses of medications. Arterial systolic pressure of mean 124 +/- 13 mmHg (range 85–146, med.125), diastolic pressure of mean 66.4 +/- 9.7 mmHg (range 40–80, med.66) were recorded in whole group. Stent redilation was performed successfully in 5 pts during follow up period due to neointimal hyperplasia or patients somatic growth. Eight Covered Cheatham-Platinum stents were additionally implanted in 5 patients due to stents fracture and in-stent stenosis, in two patients due to small aneurysm detected 4 yrs and 6 yrs after stent implantation, in one patient due to aortic arch hypoplasia.

**Conclusions:** Stent implantation in native and postoperative coarctation of the aorta has good immediate and mid-term results.

Arterial hypertension therapy can be easily modified after stent implantation.

During follow-up period, stents redilation and additional covered stent implantations may occasionally be needed.

#### P-143

##### **Transcatheter recanalization of occluded modified Blalock-Taussig shunts**

Rewers B. (1), Brzezińska-Rajszyś G. (1), Zubrzycka M. (1), Książek J. (1), Kąsy A. (2), Birbach M. (2), Zuk M. (3)  
The Children's Memorial Health Institute, Warsaw, Poland  
Heart Catheterization Laboratory (1), Cardiosurgery Department (2), Cardiology Department (3)

**Background:** Modified Blalock-Taussig shunt occlusion results in a sudden reduction of pulmonary vascular perfusion, causing dramatic deterioration with decrease of oxygen saturation and metabolic acidosis in patients. The results of local recombinant tissue plasminogen activator (rt-PA) infusion, balloon angioplasty and additional stent implantation of obstructed modified Blalock-Taussig shunts were studied to assess their role.

**Material and methods:** From April 2003 to December 2009, various emergency treatments for occlusion or critical stenosis of modified Blalock-Taussig shunts were performed in 11 pts (7 male and 4 female), age 7.53 +/- 7.62 mths (range 0.5–22 months), weight 6.25 +/- 5.35 kg (range 3.3–9.5 kg). The shunt diameter was 3.5 mm in 10 pts and 4 mm in 1 pt. The mean time from surgical shunt placement to intervention was 4.8 +/- 5.2 mths (range 13 days–13 mths). Occlusion of the shunts was diagnosed on the basis of acute decreases in systemic arterial saturation (from 80–95% mean 85.5% to 30–75% mean 51%) and was confirmed in all pts by Doppler echocardiography. The emergency transcatheter recanalisation procedures were performed under general anesthesia with mechanical ventilation via femoral artery approach. Two to three boluses of rt-PA (dose 0.3–0.5 mg/kg, intervals 10 min) were given into the proximal end of the shunt in 9/11pts. Balloon angioplasty with Tyshak balloons (NuMed, Canada) of diameter of 4 mm was performed in 10/11 children (in addition to rt-PA infusion in 8 pts) and stent implantation in 1/11 pt. After the procedure all pts received heparin infusion (100–150 mg/kg/24h) for 2–3 days or low molecular weight heparin (Clexane)(1 mg/kg/24 h) for 3–5 days, followed by oral aspirin (2–3 mg/kg/day) until further surgery was needed electively.

**Result:** All the shunts were successfully recanalized. Mean arterial saturation increased from 51 to 83% Mean arterial saturation increased from 51% (range 30–75%) to 83% (range 70–95%) This corresponded to angiographic improvement in the shunt diameter. One child had stent implantation 7 mths after balloon angioplasty due to shunt restenosis.

**Conclusion:** Our experience demonstrates that the acute occlusion of modified Blalock-Taussig shunts can be successfully recanalized with transcatheter methods.

#### P-144

##### **Aorto-pulmonary collaterals causing complicated postoperative course after cardiac surgery – results of percutaneous treatment.**

Zubrzycka M. (1), Brzezińska-Rajszyś G. (1), Rewers B. (1), Książek J. (1), Kąsy A. (2), Birbach M. (2), Ziolkowska L. (3)  
The Children's Memorial Health Institute, Warsaw, Poland.  
Heart Catheterization Laboratory (1); Cardiosurgery Department (2); Cardiology Department (3)

**Introduction:** The aim of the study was to assess the role of percutaneous embolization of aorto-pulmonary collaterals in patients with complicated postoperative course after cardiac surgery.

**Material and methods:** Over a period of 13 years (1996–2009), in 30 patients with a complicated postoperative course after cardiac surgery aorto-pulmonary collaterals were found on aortography. Seventeen pts were after arterial switch for TGA, 9 pts after ToF correction, 1 pt after DORV+IAA repair, 1 pt after TAC+IAA, 1 pt after TAPVC to the portal vein repair, 1 after reoperation for VSD recanalization after DORV repair. Mean age of pts was 111.4 ± 141.7 days (range 12–500 days), mean body weight was 4.8 ± 2.4 kg (range 2–10 kg). All pts required mechanical ventilation for mean 18.2 ± 12.9 days (range 1–47 days) after operation, and received inotropic support and diuretics for heart failure. Good result of correction was confirmed by echo and heart catheterization. One to five aorto-pulmonary collaterals were found on aortography, diameters of collaterals were 1–2 mm (mean 1.5 mm). Collaterals were closed from the femoral artery approach (28 pts) and carotid artery cut-down (2 pts) using Cook microcoils in most cases. Microcatheter was inserted through the guiding catheter positioned in the collateral and coils were delivered. Gianturco coils were used for closure of 9 collaterals and PDA coils for 5 collaterals.

**Results:** Collaterals were closed successfully in all pts using 1–8 coils. Clinical improvement was observed in all pts and mechanical ventilation could be stopped 1–9 days (mean 2.7 ± 2.2 days) after the procedure. One microcoil embolized to the femoral artery and was successfully snared and retrieved from the artery. No other complications occurred.

**Conclusions:** 1. Aorto-pulmonary collaterals may result in prolonged mechanical ventilation and heart failure after cardiac surgery. 2. Percutaneous embolization is safe and effective method of treatment in these cases.

#### P-145

##### **Occlusion of Fontan fenestrations using Amplatzer Duct Occluder II**

Briales C., Zunzunegui J.L., Ballesteros F., Alvarez T., Maroto E., Centeno M., Medrano C.  
Gregorio Marañón Hospital. Madrid, Spain.

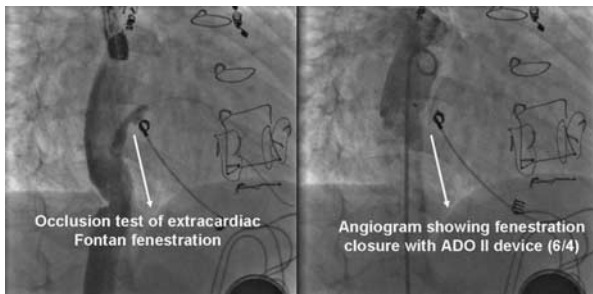
**Objectives:** Different devices have been used to close a patent fenestration after a modified Fontan operation. The aim of this

study was to evaluate the efficacy and safety of the Amplatzer Duct Occluder II for occlusion of Fontan fenestrations.

**Methods:** A retrospective review was used to study 12 patients, median age 8.16 years (range, 5 to 19 years old) who underwent Amplatzer Duct Occluder II device transcatheter closure of Fontan fenestration in our institution between May 2008 and June 2009. Each patient had complete right and left heart catheterization and fenestration test occlusion to assess haemodynamic suitability for closure. Patients were considered for the ADO II closure if the Fontan pressure was less than 14 mmHg. Sizing of the defect was achieved with transesophageal echocardiography.

**Results:** The device was implanted successfully in all 12 patients (5 with extracardiac conduit and 7 with intracardiac lateral tunnel) without significant protrusion into either the systemic or pulmonary venous side of the baffle. All the patients tolerated test occlusion and device occlusion of the fenestration without significant changes in arterial or venous pressures. Mean pressure in the Fontan circuit increased from 10.6 mmHg (7 to 13 mmHg) to 11.5 mmHg (9 to 14 mmHg) with an average increase of 0.83 mmHg (0 to 3%). All of them experienced significant improvement in oxygen saturation. Arterial partial oxygen pressure (paO<sub>2</sub>) increased from an average of 59.4 mmHg (40 to 72 mmHg) to 113 mmHg (77 to 164 mmHg). The immediate postimplantation angiogram revealed only trivial residual leaks in four patients. Subsequent follow-up echocardiography showed complete occlusion in all four patients. The position and integrity of the device was satisfactory and there were no complications during the follow-up period.

**Conclusions:** The Amplatzer Duct Occluder II device is a valuable tool that can be used successfully to occlude Fontan fenestration. It is a low profile device that allows for ease of placement without significant protrusion to either side of the circulation.



#### P-146 Transcatheter closure of atrial septal defects with transthoracic echocardiography

Şahin M. (1), Özkutlu S. (1), Karagöz T. (1), Yıldırım I. (1), Çeliker A. (2), Alehan D. (1), Özer S. (1)

Hacettepe University Department of Pediatrics, Section of Pediatric Cardiology, Ankara, Turkey (1); Acibadem Hospital, Maslak, İstanbul, Turkey (2)

**Objectives:** Transcatheter closure of atrial septal defects (ASD) is usually performed under general anesthesia with guidance of transesophageal echocardiography (TEE). However, in selected patients, transthoracic echocardiography (TTE) can be used instead of TEE. The aim of this study is to evaluate our clinical experience using the Amplatzer septal occluder (ASO) for catheter closure of secundum ASD under TTE guidance without general anesthesia.

**Methods:** Between November 2006 and April 2009 secundum ASD was closed with ASO in thirty-nine patients with TTE guidance, while TEE was used in 47 patients. The largest ASD

diameter measured in different views was selected as the reference ASD diameter. Balloon sizing was used in four patients. Reassessment was done at first month, 6th month and every year thereafter with transthoracic echocardiography and Holter monitoring.

**Results:** The mean age of these patients was  $7.8 \pm 3.8$  years (range 3–17 years). Patients weighed between 11–58 kg (mean weight  $27.5 \pm 12.4$  kg). The mean ASD diameter determined by TTE was  $10.9 \pm 3.2$  mm (range 4–18 mm). Total septal diameter was  $38.4 \pm 6.5$  mm (range 31 to 55 mm), the mean device diameter and the difference between device diameter and ASD diameter were  $12.8 \pm 4$  mm (range 4–22 mm) and  $1.8 \pm 1.3$  (range 0–6) respectively. The mean total procedure time was  $51.4 \pm 13.2$  minutes (range 30–90 minutes) and the mean fluoroscopy time was  $4.8 \pm 1.8$  minutes (range 2.2–9.2 minutes). There were no major complications like device fracture, embolization, or migration. The mean time of follow up was  $11.6 \pm 8.5$  (1–25) months.

**Conclusion:** In selected cases, in which the defects are small and the rims are adequate and TTE provides high image quality, TTE can be substituted with TEE. The fact that TTE imaging does not require general anesthesia makes it a more comfortable technique for the patient, renders the patient to less complication risks compared with general anesthesia and might decrease the procedure time. However, TTE should not be used when there is large or multiple defects, or the rims are thin and soft and the image resolution is inadequate.

#### P-147

##### Interventional palliation of a patient with isolated left pulmonary artery: clinical course and histopathology

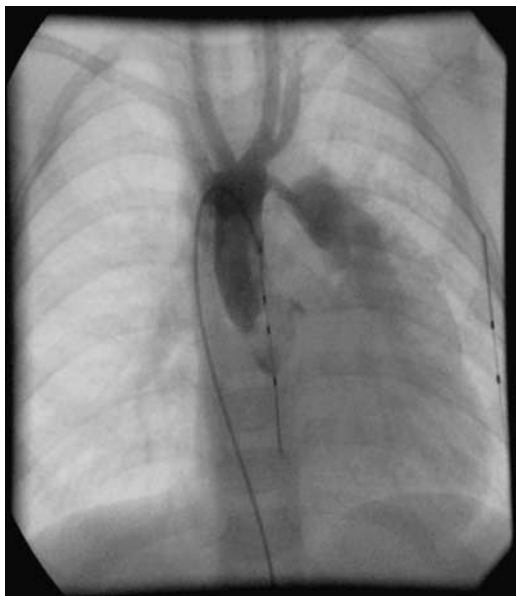
Koch A. (1), Cesnjevar R. (2), Dittrich S. (1), Breuer C. (1), Zink S. (1), Glöckler M. (1), Sigler M. (3)

Dep. of Pediatric Cardiology (1) and Dep. of Pediatric Cardiac Surgery (2), University of Erlangen-Nürnberg; Dep. of Pediatric Cardiology, University of Göttingen (3), Germany

**Introduction:** The absence of a main branch of the pulmonary artery is a rare congenital defect with duct-dependent pulmonary circulation. A gap between the main pulmonary artery and the hilar vessel complicates repair of this anomaly by surgery in neonates. Therefore, initial palliative treatment is often performed either by surgical creation of an aortopulmonary shunt or by stenting the arterial duct using coronary stents. However, progressive luminal narrowing due to intimal proliferation and peel formation is a considerable problem.

**Case report:** Successful interventional palliation in a 7-week-old girl with isolated left pulmonary artery was performed using Sirolimus-eluting stents. Echocardiography and angiography (Fig.) revealed a good perfusion of the left pulmonary artery during the follow-up. At the age of 2.5 years, surgical repair was performed by implantation of the left pulmonary artery into the pulmonary trunk. Macropathology of the surgically removed supplying vessel demonstrated an open inner lumen of the stented vessel without thrombus apposition at the wall and complete coverage of the stent struts by a thin layer of whitish tissue. Histology revealed an elastic artery with a comparatively narrow media and a normal adventitia without necrosis. The thickness of the neointima was up to 250 µm, but there was also a circumscribed thinning of the intima over a quarter of the circumference. Additionally, there was a moderate inflammatory reaction locally related to the struts with histiocytes and also some foreign body giant cells, but neither granulocytes or lymphocytes nor metallic particles were identified in the surrounding tissue.

**Conclusion:** The implantation of Sirolimus-eluting stents preserved the patency of the stented vessel for 29 months. Stents eluting antimitotic agents may allow a more sustainable palliation in patients with duct-dependent pulmonary blood flow.



#### P-148

##### **Long term follow-up of transcatheter closure of atrial septal defects for paradoxical embolism with the Helex Septal Occluder: a contrast-enhanced transcranial Doppler study**

Lunardini A. (1), Recla S. (1), Ait-Ali L. (1), Chiappino D. (2), Monteleone A. (2), De Lucia V. (1), Kristo I. (1), Spadoni I. (1) *Pediatric Cardiology and GUCH Unit (1); Department of Radiology (2); "G. Monasterio" Tuscan Foundation, "G. Pasquinucci" Heart Hospital, Massa, Italy*

**Objectives:** to evaluate the long term follow-up result of transcatheter occlusion of atrial septal defects with the Helex Septal Occluder (HSO).

**Patients and methods:** the clinical and echographic data of 43 pts (mean age  $44 \pm 13$  years) with atrial septal defects and paradoxical embolism, treated with HSO implantation in our Institution, and followed-up for at least 2 years, were prospectively collected and evaluated. Atrial septal anatomy was defined as simple (patent foramen ovale, PFO,  $\pm$  atrial septal aneurysm, ASA; ASA with one fenestration, F-ASA) or complex (F-ASA with PFO; ASA with multiple fenestrations  $\pm$  PFO). FU protocol consisted of: contrast-enhanced transthoracic echocardiography (ce-TTE) at discharge, 6 and 12 months, and contrast-enhanced transcranial Doppler (ce-TCD) at 1 year and every year in case of RS. The amount of ce-TCD RS was graded as: 0–10 microbubbles: no or trivial; >10: moderate; >50 or curtain pattern: severe. Moderate or severe RS were considered significant.

**Results:** the anatomy was simple in 26 pts and complex in 18. Mean FU duration was  $4,3 \pm 1,4$  years. A significant RS was present in 16 pts (37%) at discharge, 11 pts (26%) at 6 months, 9 pts (21%) at 1 year and in 5 pts (12%, 3 with complex and 2 with simple anatomy), at 2 years-FU. Among the 9 pts with a positive ce-TCD at 1 year FU, in 2 ce-TTE did not show RS. All the 5 pts with late RS were asymptomatic; 2 pts, with severe shunt and multiple defects, underwent implantation of a second device. In the remaining 3, the amount of RS was moderate.

**Conclusions:** the Helex Septal Occluder is safe and effective for treatment of atrial septal defects in pts with paradoxical embolism. Complete closure can be delayed after the first year; however, in case of severe RS and complex anatomy, an additional procedure may be needed. Ce-TCD is a high-sensitive tool for FU evaluation.

#### P-149

##### **Experience with hybrid pulmonary valve replacement in an acute pig model**

Schmitz C. (1), Thalmann R. (1), Merkel E. (2), Bombien R. (1), Akra B. (1), Aleksieva G. (1), Negaioe A. (1), Fano C. (3), Dauner M. (3), Sodian R. (1), Lang N. (2), Breuer J. (4), Neuss M. (5), Reichart B. (1), Kozlik-Feldmann R. (2)

*Department of Cardiac Surgery, University of Munich, Munich, Germany (1); Department of Pediatric Cardiology and Intensive Care Medicine, University of Munich, Munich, Germany (2); Institute for Textile Technology and Process Engineering Denkendorf, Germany (3); Department of Pediatric Cardiology, University of Bonn, Germany (4); Manemed, Bonn, Germany (5)*

**Introduction:** Interventional implantation of pulmonary valves via the femoral vein is often limited by tortuous vascular access or size restraints of the pulmonary valve. In these cases a hybrid procedure provides an alternative. But currently there is no commercial device available for this type of procedure.

**Methods:** We tested the feasibility of hybrid pulmonary valve implantation in an acute pig model ( $n = 10$ ). For the experiment we built custom-made stented pulmonary valves, consisting of either a balloon expandable bare metal stent or a self-expanding nitinol stent with different types of heart valves (bovine jugular vein ( $n = 3$ ), bovine pericardial valve ( $n = 4$ ), sprayed polyurethane ( $n = 3$ )). The chest was opened either via partial median sternotomy ( $n = 6$ ), median sternotomy ( $n = 2$ ), transverse sternotomy ( $n = 1$ ) or right anterior thoracotomy ( $n = 1$ ) or. A large sheet was introduced directly into the right ventricle through two Teflon-armed purse strings sutures. Finally, the stented pulmonary valve was placed under fluoroscopic and epicardial echocardiographic guidance.

**Results:** Experience of our first pulmonary valve implantation demonstrated the feasibility of the procedure in an acute pig model. We were able to implant both balloon expandable and stent expandable stented valves. No limitations regarding size of the stented valve were seen.

**Conclusions:** We describe our experience with hybrid pulmonary valve implantation in an acute pig model. We demonstrated the feasibility of the procedure regarding the surgical technique and perioperative management, and prepared the field for a chronic trial.

#### P-150

##### **Right ventricular outflow tract stent implantation as a primary palliation in patients with Tetralogy of Fallot**

Reinhardt Z., Stumper O, Miller P, Stickley J, Viswanathan S, De Giovanni J.V.

*The Heart Unit, Birmingham Children's Hospital, Birmingham, UK*

**Introduction:** Creation of a Blalock-Taussig shunt remains the initial palliation in neonates and infants with complex Tetralogy of Fallot (TOF). Although this is usually an effective procedure it carries a significant morbidity. In selected cases a trans-catheter approach with stenting of the right ventricular outflow tract (RVOT) may be a feasible and effective alternative.

**Aim:** To evaluate the effectiveness of RVOT stenting in symptomatic neonates and infants with complex TOF physiology.

**Method:** Retrospective data analysis of RVOT stent implantation as a primary palliation between 2005 and 2010.

**Results:** Twelve patients underwent primary RVOT stenting over the 5 year period. Indications were severe cyanosis and spells ( $n = 9$ ), TOF with complete atrioventricular septal defect ( $n = 3$ ). Six patients suffered from an associated genetic syndrome. Patient's age at stenting ranged from 19 to 406 days (median 66) and weight ranged from 1.75 to 12.2 kg (median 3.8). Pulmonary arteries were hypoplastic in all patients with a mean pulmonary valve diameter of 4.5 mm, RPA diameter of 3.5 mm and LPA of 3.0 mm.

Seventeen stents were implanted in 12 patients. Five had 2 stents at the time of initial procedure due to instability of stent or inadequate cover of the entire length of the infundibulum. A further 3 required repeat stenting subsequently.

Stents implanted were: 13 premounted Liberte coronary balloon expandable stents (Boston Scientific), 3 peripheral vascular Jomed (Abbott) and 1 Genesis (Cordis).

RVOT stenting improved systemic oxygen saturations from a median of 75% (40–82) to 90% (81–97) [ $p < 0.05$ ].

There were 3 major complications: The first patient had a perforation and tamponade requiring emergency RVOT patch surgery. The second had transient pulmonary oedema. The third patient had perforation of main pulmonary artery, developed pericardial effusion and pneumothorax and despite successful implantation of stent had a cardiac arrest and died.

Nine patients had a subsequent complete repair, with 2 awaiting further management. Median time interval between the primary intervention and definitive surgery was 204 days.

**Conclusion:** Stenting of the RVOT is a feasible and effective palliation and offers a new treatment option in the initial management of patients with complex TOF physiology.

#### P-151

##### **A new device (Cardi-O-Fix muscular ventricular septal defect occluder) for percutaneous closure of congenital muscular ventricular septal defects: early results**

Akdeniz C., Santas T., Erdem A., Odemis E., Demir F., Demir H., Celebi A

Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Center, Department of Pediatric Cardiology, Istanbul, Turkey

**Background:** Muscular ventricular septal defects (mVSD) are an important and difficult surgical problem with increased morbidity and mortality. Device closure is becoming an attractive and reliable alternative to surgery. The device cost is a major problem in developing countries.

**Objective:** To assess our experience with percutaneous closure of mVSD, with the new Cardi-O-fix Muscular VSD Occluder devices (Starway Medical Technology Inc.).

**Methods:** We treated 11 patients with congenital mVSD with the new devices. All the patients had a significant hemodynamic shunt and all procedures were performed using the standard percutaneous technique.

**Results:** Between February 2008 and May 2009, device closure of mVSD was attempted in 11 patients age from 3 months to 34 year (median: 2 year) and in weight from 7 to 67 kg (median: 10 kg). The hemodynamic size of the VSD ranged from 2.8 to 5.9 mm (median: 4.6 mm) and the Qp/Qs ratio ranged from 1.4 to 2.8 (median: 1.9). All implants were well-succeeded and in all cases a considerable reduction in flow or complete closure was achieved. No major complications were encountered.

**Conclusion:** The device appears to be safe, effective with good early results and acceptable cost affectivity.

#### P-152

##### **Cheatham-Platinum Stents for Aortic Coarctation: Immediate and Early Results**

Erdem A., Celebi A., Akdeniz C., Santaş T., Erol N., Demir H., Yalcin Y.

Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Center, Department of Pediatric Cardiology Istanbul, Turkey

**Objective:** To present our institutional experience of endovascular Cheatham-Platinum (Numed™) stent implantation in child and adults with aortic coarctations.

**Method:** Between August 2007 and December 2009, 45 patients had aortic coarctation treated with 47 stent implantation. We preferred stent implantation primarily in adult patient with coarctation, in children it is preferred in cases of aneurysm, subaortic coarctation and association with patent ductus arteriosus or in recoarctations in patients more than five years-old.

**Results:** Sixteen covered and 31 bare totally 47 balloon expandable Cheatham Platinum stents were implanted in 45 patients. In two patients two stents were implanted in tandem to adequately cover the lesion. Seven patients were postoperative surgical repair, eight had primary balloon angioplasty, and four underwent both procedures, the other 26 patients were native coarctations. Four patients were subaortic coarctations, one had blind coarctation, four had patent ductus arteriosus and two had aneurysm. In two patients the lesions were on transverse aorta and bare stents were implanted without any complication in these patients.

The mean age  $12.2 \pm 5.9$  (4–33 years) and mean weight was  $42.6 \pm 18.5$  (18–80 kg). The systolic peak pressure gradient was decreased from  $45 \pm 23$  (20–150) to  $3 \pm 3.6$  (0–10) mm Hg ( $P < 0.000$ ) and ascending aortic pressure decreased from  $148 \pm 33$  (115–230) to  $135 \pm 16$  (100–170) (P: 0.006). The mean coarctation diameter increased from  $6.9 \pm 3.4$  mm to  $13.8 \pm 3.4$  mm ( $P < 0.000$ ). The mean follow up duration was  $11.1 \pm 7.1$ , median 10 month (1–28 month).

There was no procedure related death. One patient experienced an acute wall rupture at the distal end of the implanted bare stent, which was successfully managed by implanting a covered stent immediately at the same session. During the follow five patients remained hypertensive requiring antihypertensive treatment.

**Conclusion:** Cheatham Platinum Stent implantation is safe and effective in treating coarctation of the aorta. They are very effective in reducing coarctation gradient and increasing lesion diameter.

#### P-153

##### **Transcatheter closure of congenital ventricular septal defects (vsd) with various devices and coils; early and mid-term results**

Celebi A, Akdeniz C., Zeybek C., Erdem A., Santaş T, Demir H., Erol N., Imanov E., Yalcin Y

Department of Pediatric Cardiology, Dr Siyami Ersek Thoracic and Cardiovascular Surgery Center, Istanbul, Turkey

**Objectives:** Percutaneous transcatheter closure of VSD is one of the greatest challenges in interventional cardiology. We sought the safety, efficacy, and follow-up results of transcatheter closure of VSD using different types of devices and coils.

**Methods:** Totally 56 patients underwent to transcatheter closure of VSD during the last three years. Seven Patients with suitable muscular VSDs, considered too small for transvenous closure, underwent periventricular muscular VSD closure. In perimembranous VSDs, left side of the device was inserted either to left ventricular side of the septum or into the aneurysm if available to avoid AV block.

**Results:** The median age was 5.8 (0.2–55) years. The mean follow-up was  $19.6 \pm 8.5$  months. The mean Qp/Qs ratio was  $1.9 \pm 0.95$ , and the median VSD diameter was 8.8 mm (3.5–20 mm). Implantation was successful in 52 (93%) patients with 35 perimembranous, 17 muscular VSDs. Eighteen Amplatzer muscular VSD device, 6 membranous VSD occluders, 9 Nitt-occlud coil (PFM), 11 Cardiofix muscular VSD occluders, 6 Amplatzer duct occluders, 1 Amplatzer Septal occluder and 1 detachable coil (Cook) were used. There was no early or late implant embolisation. After the procedure complete closure or important reduction of the shunt was observed. Complete occlusion rate was 52% at completion of the procedure, rising to 67% the day after and 78% at sixth month during the follow-up. No major complication was encountered except transient complete AV block in two patients and hemolysis in one patient. During follow-up, there was no de novo aortic regurgitation except in one patient with perimembranous VSD closed by muscular VSD occluder. Patients that the device implanted into the aneurysm have high residual shunts during follow-up, comparing to patients that the left disc of the device inserted to the left ventricular side ( $p < 0,05$ ).

**Conclusions:** Transcatheter closure of VSD with different types of devices is an effective and safe procedure in selected patients. Device insertion into aneurysm is an alternative in perimembranous VSD thought to have decreased possibility of AV block but tend to have increased residual shunt in early and mid-term period.

#### P-154

##### **Lead removal in children and young adults with a congenital heart disease**

Zartner P., Wiebe W., Toussaint-Goetz N., Schneider M.  
*Deutsches Kinderherzzentrum, Sankt Augustin, Germany*

**Aims:** In young patients with a congenital heart disease (CHD) transvenous leads for pacemakers (PM) or implantable cardioverter defibrillators (ICD) can cause later vascular obstruction or infection. Removal of non-functional leads is controversial as it bears the risk of vascular disrapture and embolisations. We report the data of a single centre for paediatric cardiology on efficiency and safety of transvenous lead removal.

**Methods:** Between May 2005 and August 2009 in 22 patients with a mean age of 12.9 years [range: 3.6 to 29.5 years] removal of 28 transvenous leads (mean lead age: 5.1 years) was attempted. The main indications for removal were vascular obstruction, increased threshold and lead dislocation. Commercially available retraction tools were used, if necessary.

**Results:** 25 leads (89%) were retrieved with clinical success, of which 22 (79%) were removed with complete procedural success. In 3 leads the lead tips were retained, while 3 leads could not be retrieved. No major complications occurred. Additional interventions such as recanalisation, balloon dilation or stent implantation were performed as indicated. Procedure and X-ray times could be correlated to the implant age of the leads.

**Conclusion:** Using non-electrical techniques, transvenous lead removal can be performed with a success rate of 89% in young patients with a CHD. In the case of vessel obstructions, lead replacement combined with revascularisation should be performed early, as the older the lead, the more prolonged and more hazardous the extraction procedure becomes. The use of new leads and precautionary implantation techniques may facilitate later lead removal.

#### P-155

##### **Novel pre-mounted balloon expandable graft stent (Avanta V12) expands interventional options to treat cardio-vascular obstructions.**

Schranz D., Christian Jux, Karl-Jürgen Hagel, Jürgen Bauer, Klaus Valeske, Hakan Akintürk

*Pediatric Heart Center Giessen&Marburg and Frankfurt, Germany.*

Certain congenital or acquired cardio-vascular obstructions need surgical or interventional (re-) repair. Percutaneous or surgical-interventional hybrid graft stent implantation might be indicated when risks as bleeding, rupture or aneurysm formation are expected. Our initial experiences with a V12 graft stent (AdvantaR, Atrium) are reported.

**Patients and Method:** 8 patients, aging between 5 and 25 years were treated utilizing V12 stent. Two patients suffered (re-)aortic coarctation and a five-year-old girl with HLHS after hybrid stage III from re-obstructed distal aortic arch. In three patients obstructed left pulmonary artery was stented, in one as a hybrid approach during surgical replacement of a pulmonary homograft. Further 2 patients, one with an atretic bridging vein (V anomyma), and one with a long segment atretic right ventricular outflow tract (RVOT), were treated after HF-frequency perforation and gradual balloon dilation. The V12 implantations were performed in conscious sedation in all percutaneous approaches.  $12 \times 29/41$  mm V12 stents were placed by utilizing a 9Fr long sheath;  $14 \times 41$  and  $16 \times 41$  V12 graft stent were advanced through a 11F long sheath.

**Results:** V12 STENT placement was technically un-problematic in all, even as a hybrid approach or without covering by a long sheath. In a 15-year-old boy, who suffered from native aortic coarctation, significant recoiling after V12 stent placement needed an additional MaxLD stent for re-expansion. Hemodynamic improvement was achieved in all, even in the 25-year-old patient with a AP-shunt dependent pulmonary circulation, in whom an additional pulmonary flow was created by placement of a V12 graft within the RVOT to improve hypoxemia; fluoroscopy time of the seven patients treated in the cath-lab ranged between 6,7 and 46,1 min. In the follow-up between 2 and 9 months, no patient needed a re-intervention.

**Conclusion:** V12 expands the options to treat cardio-vascular obstructions; pre-mounted stent-graft design allows smaller delivery sheaths or no long sheath covering; graft design reduced the risk of bleeding and aneurysm formations; the open cell design has the advantage of “growing” from 12 and 20 (22) mm, higher stent flexibility with higher residual vessel compliance after stent placement has to weight against higher re-coiling risk in rigid vascular diseases.

#### P-156

##### **Up to 7-year Follow-up after Transcatheter Closure of Paravalvar Leaks**

Radtke W.A.K. (1), Bandisode V.M. (2) O’Murchu B (3)  
*Nemours Cardiac Center, Wilmington, DE, USA (1); Medical University of South Carolina, Charleston, SC, USA (2); Temple University, Philadelphia, PA, USA (3).*

Paravalvar leaks may cause significant hemolysis or congestive heart failure and surgical repair is difficult. Transcatheter device closure is a non-surgical alternative but information on late outcomes is limited. We have closed paravalvar leaks using Gianturco coils and Amplatzer Muscular VSD Occluders and report our medium-term outcome with this approach.

We performed 14 such procedures on 10 patients, aged 9 to 79 years (median 56 yrs). Patient weight ranged from 31 to 102 kg (median 62 kg). Significant co-morbidities were present in 7 patients. The leaks were para-aortic in 2 and para-mitral in 12 procedures. Hemolysis was present before 10 procedures. Left atrial and ventricular diastolic pressures were elevated in all patients. Single leaks were targeted in 8, and multiple leaks in 6 procedures. Leak diameters ranged from 1 to 10 mm. Para-mitral

leaks were approached antegrade from the right internal jugular and femoral vein after transeptal puncture. Ten Coils and 15 Amplatzer Muscular VSD Occluders were implanted in 10 patients (device diameter 4–10 mm). Fluoroscopy time ranged from 18 to 124 minutes (median 65 min). There were no procedure related late complications. Hemolysis improved or resolved in 4 patients but later recurred in 1. There was no interference with artificial valve function. Complete closure was achieved in 4 patients. However, despite initial clinical improvement 6 patients expired 1.5 to 18 months after the procedure: 2 from renal failure, 1 from multi-organ and 1 from heart failure, 1 from subdural hematoma and 1 from complications after surgical revision for persistent severe hemolysis. One patient is lost to follow-up. Three patients are well with resolved or insignificant residual hemolysis 4 and 7 years after the procedure. Transcatheter closure of paravalvar leaks appears safe but hemolysis often persists and the improvement in overall outcome appears limited.

#### P-157

##### **Percutaneous closure of atrial septal defects in children: what are the limits?**

Barre E. (1), Petit J. (1), Losay J. (1), Capderou A. (2), Piot J.D. (1), Stos B. (1), Legendre A. (1), Serraf A. (1), Lambert V. (1)  
Centre Chirurgical Marie-Lannelongue, Le Plessis Robinson, France (1); INSERM U-999, Le Plessis Robison, France (2)

**Introduction:** Percutaneous atrial septal defect (ASD) closure has become a safe and efficient alternative to surgery in adults. In paediatric population, the feasibility of this technique is still debated and may be limited, especially in case of large defect in young children. Our aim was to determine the anatomical factors that may predict the feasibility and the success of a percutaneous ASD closure.

**Methods:** Records of consecutive patients less than 12 years-old who underwent an attempt of a percutaneous ASD closure using Amplatzer septal occluder device were retrospectively reviewed. Anatomical analyze of ASD included transthoracic echocardiographic measures of ASD diameter, length of rims surrounding ASD, length of atrial septum, and balloon-stretched ASD diameter obtained during catheterization. Calculated ASD area was normalized to the body area of patients. The procedure was abandoned when the device placement failed after three attempts.

**Results:** 140 patients (mean age  $7.7 \pm 2.5$  years, mean weight  $25.3 \pm 9$  kg) were included. Mean echocardiographic ASD diameter was  $15.0 \pm 4.1$  mm, balloon-stretched diameter  $19.8 \pm 4.7$  mm, length of atrial septum  $34.8 \pm 5.9$  mm, diameter of implanted device  $18.6 \pm 4.3$  mm. In all patients, a rim measuring at least 3 mm surrounded ASD. Percutaneous ASD closure was achieved in 131 patients, with some difficulties to place the device in 5. Procedure was abandoned in 8 patients, an immediate device embolization into right atria occurred in one; these patients were referred to surgery without any complication. Factors associated with a successfully percutaneous closure included a smaller ASD echocardiographic and balloon-stretched diameters than in failed procedure (respectively  $14.6 \pm 3.8$  vs  $19.9 \pm 4.4$  mm,  $p < 0.001$  and  $19.0 \pm 0.6$  vs  $25.6 \pm 6.3$  mm,  $p < 0.001$ ), a smaller ASD area/body area ratio ( $338 \pm 139$  vs  $569 \pm 210$ ,  $p < 0.05$ ), a smaller device size ( $18.3 \pm 3.9$  vs  $22.4 \pm 5.7$  mm,  $p < 0.05$ ), and a smaller difference between balloon-stretched and implanted device diameters ( $1.0 \pm 0.6$  vs  $3.1 \pm 2.8$  mm).

**Conclusion:** Percutaneous ASD closure using Amplatzer device is effective and safe in most of children. Successfully procedure is associated with a small ASD and a small device but the temptation to undersize devices should be avoided. In large

ASD and/or in small patients, normalization of ASD area to body area may help to choice the best treatment.

#### P-158

##### **Haemoptysis after the Fontan procedure: Coil embolisation of aortopulmonary collaterals**

Ahmad.Z., Lim.Z., Kiesewetter.C., Veldtman.G., Vettukattil.J.J  
Congenital Cardiac Centre, Southampton University Hospital Trust, UK.

Haemoptysis is a serious complication in patients with Fontan circulation, leading to haemodynamic compromise. Aetiology includes aortopulmonary collaterals, pulmonary arteriovenous malformation and pulmonary embolism.

**Methods:** This is a retrospective study over 2 years. We identified 3 patients with haemoptysis and Fontan circulation. Notes were retrieved to determine baseline characteristics, catheterisation findings, intervention, results and short-term outcome.

**Results:** 3 patients, age range from 8 to 15 years, 2 had extracardiac total cavopulmonary connection (one had Fontan conversion) and one had atriopulmonary Fontan. All patients had Glenn procedure at childhood. All patients were found to have significant aorto pulmonary collaterals on cardiac catheterisation. Timely and appropriate transcatheter coil embolisation therapy of these collaterals was effective in all patients, with clinical and saturation improvement. Identified sources of bleeding collaterals are the internal mammary arteries, bronchial arteries, thyrocervical artery and lateral thoracic artery. Lower oxygen saturation is associated with earlier haemoptysis after Fontan procedure.

**Discussion:** Aortopulmonary collateral is a significant cause of haemoptysis in Fontan patients. Given that multiple collaterals often coexist it is crucial to identify the bleeding site. History, clinical signs, CXR, CT-chest, bronchoscopy and understanding of the established sites of collaterals vessels may help to identify sources prior to embolisation. This procedure is often uncomplicated but should be performed by an experienced operator given the potential haemodynamic consequences.

**Conclusion:** Aorto pulmonary collateral associated with Fontan circulation can lead to significant haemoptysis. Embolisation of collaterals is effective and low risk in experience hands.

#### P-159

##### **Transcatheter closure of ventricular septal defects with the Amplatzer Duct Occluder II (ADO II)**

Bauch A., Reinhardt Z., Miller P., De Giovanni J.  
The Heart Unit, Birmingham Children Hospital, Birmingham, UK

**Introduction:** We report our early experience of closing ventricular septal defects with the ADO II, which is a self-expanding, fabric-free, nitinol mesh device with dual articulations. It requires only a 4F or 5F delivery catheter and may be deployed from the arterial or venous side.

**Method:** Single centre observational study over 18 months.

**Results:** Nine patients were included (6 male). Median age: 9.2 years (3 months–18 years). Median weight: 35.6 kg (4.8 kg–71.1 kg). Five were perimembranous ventricular septal defects (PMVSD) and 4 were muscular ventricular septal defects (VSD). The diameter ranged from 1 to 6 mm. The indications for VSD closure were failure to thrive, decreased exercise tolerance, left ventricular volume overload, worsening aortic insufficiency, residual VSD after previous device closure and small asymptomatic VSD for a career choice. In 2 patients 2 devices were deployed 5/4 and 6/4 ADO II. In the remaining 5 patients 3/4, 3/6, 4/4, 4/6 and 5/6 devices were used. Seven devices were deployed from the arterial side and 2 from the

venous. Mean procedure time was 106 min (range: 21–151 min), mean fluoroscopy time was 39 min (range: 5–87 min), mean dose: 106 cGy/cm (range: 8.1–227cGy/cm). Immediate complete angiographic closure was seen in 6 patients (66%), while in the other 3 patients a small residual shunt remained. In one patient the device embolised to the left pulmonary artery by the following day. It was removed and replaced by a muscular device. In another patient the device was not released due to severe tricuspid regurgitation, but the VSD was then successfully closed with an ADO I. The mean follow up time was 13 months (range 6 weeks to 20 months). There was no evidence of new left ventricular outflow tract obstruction, mitral valve regurgitation or aortic insufficiency or clinically significant arrhythmias.

**Conclusion:** The new ADO II is a versatile, flexible and effective device for selected muscular and PMVSDs closure. It could be used as an alternative to the currently available devices but a larger series, longer follow-up, and comparative studies are required to further evaluate the safety and effectiveness of the new ADO II.

### P-160

#### Features and Concepts: Saudi Congenital Heart Defects Registry

Dessouky N

Saudi Congenital Heart Defects Registry, Department of Biostatistics, Epidemiology, Scientific, Computing, King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia

**Introduction:** True figures reflecting the rate of incidence and prevalence, disease classification and distribution, along with risk factors of Congenital Heart Diseases in Saudi Arabia are not yet well known. Congenital Heart Defects Registry started on a hospital based level in 1998 at King Faisal Specialist Hospital and Research Center Riyadh, Saudi Arabia. The registry is an on going process of collection, processing, and analysis of data on congenital heart defects patients. The registry is secured web-based program using the state-of-the art technology for the on-line software.

**Objectives include:** providing leadership in establishing and maintaining comprehensive congenital heart defects registration in collaboration with other national and international organizations, and supporting scientific and clinical research impacting on prevention, intervention and overall management care of congenital heart defects.

**Methods** All patients having congenital heart defects are eligible for inclusion in the registry. Events recorded include; Demographics, diagnostics, catheter-based interventions, cardiac surgery and follow up visits. European Pediatric Cardiology Coding System is utilized for coding.

**Results:** Around 18,500 patients have been registered since the inception of the registry. Data are reported on annual basis. Collaboration with regional and national hospitals is ongoing. Researchers can obtain an on line up to date information regarding counts and statistics. Many spin off projects and studies based on the registry data have been published.

**Conclusions:** Congenital Heart Defects Registry is now well-established. Available data will help in understanding the burden of Congenital Heart Disease in our area and hence better planning by health authorities to improve provided care.

### P-161

#### Cardiac Catheter Guiding Surgical Closure of Muscular VSD

Zunzunegui JL, Ballesteros F, Alvarez T, Centeno M, Camino M, Medrano C, Maroto E, Briaies C, Panadero E, Greco R  
Gregorio Marañon University Hospital. Madrid. Spain

**Introduction:** Muscular VSDs have been a challenge to the surgeons because the apical or anterior muscular VSDs are difficult to visualize intra-operatively and, hence, closure is either incomplete or not possible. We present a technique to mark percutaneously the defect immediately before the surgical procedure in order to facilitate the intraoperative identification and closure of muscular VSDs.

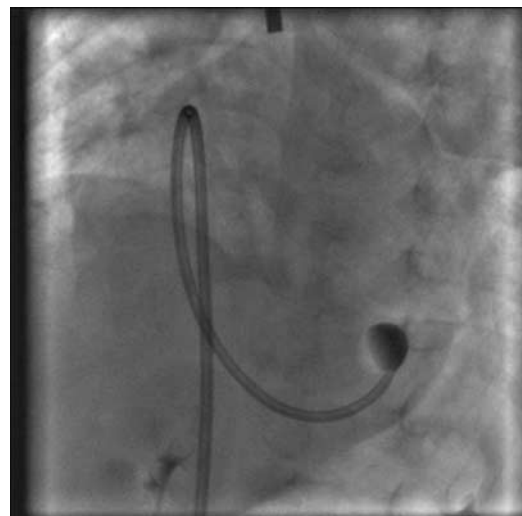
**Methods:** Retrospective review of patients with muscular VSD associated with other defects requiring open surgical repair in whom it was marked with a catheter.

**Technique;** access was obtained in the femoral artery and right jugular vein or femoral vein, ventriculography was performed to define the localization, size and numbers of VSDs, a arteriovenous 0.014" wire loop (from the aorta to the vein through the defect) was established. A 4 f. Bergman catheter was advanced over the wire from the jugular vein until the left ventricle, then the balloon was inflated with saline (75%) and contrast (25%) with slight pressure to stabilize the catheter tip into the ventricle chamber (*figure*), finally the wire was withdrawn and the patient was moved to the operating room from the cathlab. The surgeons identified the VSD through a minimum right ventriculotomy noting the point through which the catheter crossing the septum, and then the anaesthesiologist deflate and removed the catheter from the jugular introducer.

**Patients;** 5 Infants apical or anterior muscular VSDs associated with; CoA operated with Banding (2); DTGA (2); Multiple VSDs with a large apical defect (1). Weight 3.9–7.3 kg. Age 2–14 meses.

**Results:** There were not mayor complications during the catheterization, only some transient rhythm disturbances. There were only a slight residual shunt in the patients with multiple VSDs, in this case the catheter served like reference to close another small VSD around the main one.

**Conclusions:** This technique could reduce the cardiopulmonary bypass time and the incidence of residual shunting in complex congenital heart diseases associated with muscular VSDs. It is an alternative to the hybrid procedure avoiding some of its complications; device embolization, arrhythmia, valvular regurgitation, difficulties in crossing the septum with sheath, and device protrusion into small ventricle chambers.



### P-162

#### Midterm results of stented coarctation: A follow – up study of 90 patients

Fakler U, Eicken A., Genz Th., De Pasquale-Meyer G., Hess J.  
German Heart Center, Department of Pediatric Cardiology and Congenital Heart Disease, Technische Universität München, Munich, Germany



**Objective:** This retrospective single center study was performed to assess midterm results of stent implantation into the aorta for native and recurrent Coarctation (CoA) in children and adults.

**Methods:** Since Feb. 1999, ninety patients with CoA (native CoA: 27; female: 26) were treated by stent implantation (median age 16.7 years, range 6.9–63 years; median weight 61.5 kg, range 19.9–116 kg). In native CoA full stent expansion was not intended and a second dilatation was scheduled 6–12 months after primary implantation. In recent years native coarctation was preferably treated with covered stents.

**Results:** The peak to peak systolic gradient between the ascending and the descending aorta was reduced significantly from a median value of 22 mmHg (range, 5–65 mmHg) to 1 mmHg (range, –6–24 mmHg) ( $p < 0.001$ ). The narrowed segment was widened significantly from a median value of 8.0 mm (range, 1–14 mm) to 12.4 mm (range, 7–19 mm) ( $p < 0.001$ ). One major complication occurred with operative treatment of a compartment syndrome. The stent had embolised into the leg and was unremovable. Preceding the intervention, all patients were hypertensive. On follow-up the median clinical gradient (right arm, right limb) was 5 mmHg (range, –20–35 mmHg). 63% of the patients required antihypertensive medication after stent implantation.

**Conclusion:** Stent implantation for selected patients with native or recurrent CoA is safe and it effectively reduces blood pressure gradients across the CoA site. We suggest to implant only stents dilatable to an adult size aorta. Scheduled Redilatation might be feasible in order to reduce major complication. However, arterial hypertension persists in a significant number of the patients requiring antihypertensive therapy.

#### P-163

##### **Interventional catheterization in the management of diffuse congenital pulmonary vein stenosis in the infants and small children.**

*kammache I., Fouilloux V., Quilici J., Cuisset T., Bonello B., Macé L., Kreitmann B., Fraisse A.*

*La Timone Hospital, Marseille, France*

**Introduction:** Diffuse pulmonary vein stenosis (PVS) is a profoundly debilitating disease, leading to right ventricular failure and death. Only heart-lung transplantation may provide long-term survival, although patients often die while being on the waiting list. We sought to determine whether an aggressive transcatheter interventional approach may improve patient's survival.

**Patients and Methods:** Since 2005, 4 children presented with PVS, congenital in 3 cases, including one associated with a complex double outlet right ventricle (DORV). One patient had acquired PVS after repair of total anomalous pulmonary venous connection. Three patients underwent initially surgical repair of the PVS, with 2, 3 and 4 operations, respectively, using mainly sutureless type of repair. All the 3 patient developed restenosis within 3 to 10 weeks (median: 5.5 weeks) after each operation. In one patient, transcatheter dilation and stenting of PVS was considered first because surgery was contraindicated in this former 26 weeks premature baby with severe bronchopulmonary dysplasia.

**Results:** Six interventional catheterization were performed in the 4 patients at a median age and weight of 18 (6 to 28) months and 7.7(4.6 to 9.3)kg, respectively. Eleven interventions were performed, including high pressure balloon dilation ( $n = 5$ ), bare stent implantation ( $n = 3$ ) and drug-eluting stent implantation ( $n = 3$ ). In addition, one patient underwent bare stent implantation

with coated-balloon dilation as a hybrid procedure. All the high pressure balloon dilations failed because of elastic recoil. Stent implantations were technically successful in all cases. One patient died a few days later from low cardiac output in the setting of severe right ventricular dysfunction. The 3 remaining patients improved their functional status. The 2 year-old girl with DORV underwent heart-lung transplantation 6 months after drug-eluting stent implantation. Intraoperative examination showed no intrastent restenosis. She subsequently died from postoperative infection. Two patients are currently alive and being well, with near-normal pulmonary pressure in one and half-systemic pulmonary pressure in the other, 2 and 14 months after interventional catheterization.

**Conclusion:** Diffuse PVS often remains fatal. Transcatheter management may prolong survival or can be successful as a bridge to heart-lung transplantation. This should be considered as an interesting alternative to surgical relief of PVS.

#### P-164

##### **Percutaneous closure of atrial septal defects in patients with right ventricular outflow tract (RVOT) obstruction: indications and results**

*Recla S., Ait-Ali L., Lunardini A., De Lucia V., Giusti S., Spadoni I. Pediatric Cardiology and GUCH Unit, Fondazione G. Monasterio, "G. Pasquinucci" Hospital, Massa, Italy*

**Objectives:** to review our experience of percutaneous closure of atrial septal defects (ASD) in patients with right ventricular outflow tract (RVOT) obstruction.

**Methods:** from 1999 to 2009, 16 pts with RVOT obstruction underwent percutaneous closure of ASD. Nine were children (mean age  $10.1 \pm 2.7$  years, mean weight  $35.5 \pm 14.5$  kg): 6 previous treated percutaneously, 2 surgically and one on natural history; 7 were adults (mean age  $47.6 \pm 14.4$  years, mean weight  $67.9 \pm 10.5$  kg): 6 with previous surgeries, 1 on natural history. All patients underwent cardiac catheterization and TEE monitoring. In 10 pts test occlusion of the ASD was performed. Atrial septal defect closure was performed with the standard technique. **Results:** all children (6 ASD, 3 PFO) were asymptomatic, except one with significant arterial oxygen desaturation and cerebral abscess. The mean arterial oxygen saturation was  $93.3 \pm 6\%$ . In 8 pts angiography showed a significant bidirectional shunt through the defect. Mean right atrial pressure and mean RVOT pressure gradient were  $8 \pm 2.2$  mmHg and  $12 \pm 11$  mmHg respectively. Test occlusion of the ASD (performed in 7 pts) showed no variations in systemic output and RA pressure. Associated procedures were pulmonary valvular dilation in one pt and LPA stent dilation in another.

The adult patients (5 PFO, 2 ASD) were all symptomatic: 5 had suffered embolic cerebral events, 2 presented significant cyanosis. The mean arterial oxygen saturation was  $93.5 \pm 2.5\%$ . In all patients a significant R/L shunt was shown by angiography. Before closure, balloon dilation of the pulmonary valve was performed in 2 pts. Mean RA pressure was  $10.3 \pm 4.7$  mmHg, mean RVOT pressure gradient was  $15.7 \pm 17.8$  mmHg. Test occlusion was performed in 3 patients with no variations in systemic output and RA pressure.

After a mean FU of  $3 \pm 3$  years, the mean arterial oxygen saturation is  $98.8 \pm 1.6\%$  among the children and  $98.6 \pm 1.4\%$  among the adult patients. All are asymptomatic.

**Conclusions:** closure of ASD in patients with RVOT obstruction is well tolerated, safe and effective and, because of the incidence of ischemic events and/or cyanosis in adulthood, it should be performed as a treatment strategy during childhood.

**P-165****Emergent use of extracorporeal membrane oxygenation during pediatric cardiac catheterization**

Ballesteros E, Zunzunegui JL, Biales C., Alvarez T, Maroto E., Brochet G., Greco R.  
Gregorio Marañón Hospital. Madrid. Spain.

**Objectives:** Extracorporeal membrane oxygenation (ECMO) can successfully support the circulation of pediatric cardiac patients in a variety of circumstances. We pretend to evaluate the utility of ECMO to resuscitate patients following critical cardiac events in the catheterization laboratory.

**Methods:** Retrospective review of medical records; study period from January 2006 to December 2009. Inclusion of all pediatric patients cannulated emergently for ECMO in the cardiac catheterization laboratory for hemodynamic deterioration due to low cardiac output syndrome or catheter induced complications.

**Results:** During the study period six patients were cannulated for ECMO, from a total of 1610 cardiac catheterization procedures performed in our institution (0,3% of all catheterizations). Median age was 37 months (range 0–156); median weight 12,1 Kg (3,4–30). Median duration of ECMO was 68 hrs (24–120). Indication for catheterization was aortic valve dilatation in three cases (50%). Indications for ECMO included low cardiac output syndrome (4), hypoxemia (1) and catheter related complication (1). During cannulation 4 patients (66%) were receiving chest compressions. Median duration of cardiopulmonary resuscitation was 42 minutes (17–61). All six patients were successfully decannulated. Four patients (66%) survived to discharge, and one of them (25% of survivors) sustained neurologic injury.

**Conclusions:** ECMO is a technically feasible and successful tool in the resuscitation of pediatric patients following critical events in the cardiac catheterization laboratory.

**P-166****The Amplatzer duct occluder type II (ADO II): a versatile device – especially outside the duct**

Jux C. (1), Mebus S. (2), Mazhari N. (1), Hagel K (1), Schranz D. (1) Pediatric Heart Centre, Justus Liebig University, Giessen (1), and Dept. of Pediatric Cardiology and Congenital Heart Disease, German Heart Centre Munich (2), Germany.

**Introduction:** The Amplatzer duct occluder type II (ADO II, AGA Medical) for transcatheter closure of PDA has recently been introduced into clinical practice. It differs from the established ADO I by its design, consisting of a thinner, tighter woven nitinol wire mesh without a polyester-fabric filling and can be delivered through braided proprietary 4 or 5 F delivery catheters. While several reports have summarized the initial experience with the ADO II in percutaneous ductal closure, information regarding the use of the ADO II for closure procedures outside the duct is scarce.

**Patients and Methods:** Between 7/2008 and 12/2009 a total of 16 ADO II devices were implanted in 15 patients outside the ductal region at our institution. The mean patient age was 6.4 years (range 0.56 to 21.54) and mean body weight 20.9 kg (range 5.6 to 56.0). ADO II were implanted for closure of VSD (5× perim., 1× musc./swiss cheese), MAPCAs (1×), coronary artery fistula (1×), TCPC fenestration (3x, including fenestration stent), ductal stent (1×) and veno-venous collaterals (4×) for either the femoral vein (8×), femoral artery (4×) or subclavian vein (4×) through a 4 or 5F sheath.

**Results:** Device implantation was possible without problems or complications in all patients. The immediate complete closure rate was 44% (7/16) and increased to 75% (12/16) after a mean

follow-up of 4 months. Residual shunts were trivial (3×) or mild (1×) and occurred in patients with VSD (3×) and TCPC fenestration (1×). In one patient (TCPC fenestration) the ADO II was removed percutaneously and uneventfully 4 months after implantation because of recurrent pleural effusions.

**Discussion:** This report is meant to increase the awareness of the applicability and usefulness of the ADO II for several extra-ductal applications. The ADO II is a soft, flexible device that can be delivered through small sheath allowing for implantation in small children and from the arterial side. These device characteristics also allow for easier retrieval after release/implantation if needed. Although our study cohort and follow-up is limited, the early complete closure rate of this “bare metal” device seems to be lower compared to Amplatzer devices containing polyester-fabric.

**P-167****Percutaneous right outflow tract valve implantation: when is right moment for pre-stenting?**

Brugada G. (1), Kammache I. (2), Iserin L. (3), Fraise A. (2), Boudjemline Y. (1)  
(1) Necker for Sick Kids, Paris, France; (2) Hôpital La Timone, Marseille, France; (3) George Pompidou Hospital, Paris, France

**Introduction:** Percutaneous pulmonary valve insertion has been recently introduced in clinical setting. Patient selection is widely accepted. Initial results demonstrated early and differed stent fractures that make consider pre-stenting as a previous step for the procedure. To date, differed or intra-procedure pre-stenting are both accepted techniques.

**Patients and methods:** We reviewed patients included over the last 6 months in the prospective study (REVALV) for patients undergoing RVOT intervention for severe stenosis and/or insufficiency. Only valved stent group is analyzed here. All patients undergoing valved stent implantation are previously pre-stented with a bare metal stent according to present recommendations. Thirty-seven patients were included, distributed in two groups according moment of pre-stenting: differed pre-stenting (bare metal stent implantation several days before valved stent implantation -20 patients-) and same procedure pre-stenting (bare metal stent implantation at the same procedure of valved stent implantation-17 patients-). For analytical purposes, we considered RVOT anatomy (homograft, synthetic tube, patch-extended RVOT or native outflow tract).

**Results:** Overall, no differences were found regarding mean procedure times (77,35 vs 96,88, p = NS) and time of hospitalization (2,95 vs 3,63, p = NS). Mean delay time from pre-stenting to valvulation was 196,5 ± 68 days. Rv to Ao ratio improvement from basal to valvulation was significantly better in intra-procedure pre-stenting group (0,172 vs 0,373, p = 0,009). Concerning complications, bare metal stent mobilization happened just after implantation while trying to place valved stent delivery gain. Two pelvic hematomas were observed (one of each group).

**Conclusions:** Intra-procedure pre-stenting influences final result when considering RV-to-Ao ratio improvement, probably related to increase radial strength. The risk, however, remains higher as freshly implanted bare metal stent can mobilize, especially in native RVOT. Stratification of patient should be considered while choosing candidates for valved stent implantation.

**P-168****Percutaneous right outflow tract valve implantation: substrate matters.**

Brugada G. (1), Kammache I. (2), Iserin L. (3), Fraise (2), Boudjemline Y. (1)

(1) Necker for Sick Kids, Paris, France; (2) Hôpital La Timone, Marseille, France; (3) George Pompidou Hospital, Paris, France

**Introduction:** Percutaneous pulmonary valve insertion has been recently introduced in clinical setting. Patient selection is widely accepted. These candidates are however heterogeneous, in regard of heart defects, and type of surgical right ventricular outflow tract (RVOT) reconstruction. It is presently unclear in the literature if type of surgical reconstruction matters for the success of the pulmonary valve insertion. Our goal was to compare hemodynamic results of percutaneous pulmonary valve in patients with homografts, prosthetic conduit or RVOT reconstructed with patch.

**Patients and methods:** We reviewed patients included over the last 6 months in the prospective study (REVALV) for patients undergoing RVOT intervention for severe stenosis and/or insufficiency. Only valved stent group is analyzed here. All patients undergoing valved stent implantation are previously pre-stented with bare metal stent according to present recommendations. Thirty-seven patients were included, distributed in three groups according to type of RVOT reconstruction (homograft  $n = 10$ ; prosthetic conduit,  $n = 20$ ; RVOT enlargement by patch,  $n = 7$ ). **Results:** Overall, all groups were similar in RV to AP gradient improvement (after pre-stenting mean 30,79 vs 28  $p = NS$ ; final result mean 23,71 vs 28,17,  $p = NS$ ), RV to aorta pressures ratio (after pre-stenting 0,187 vs 0,3117  $p = NS$ ; final result man 0,315 vs 0,317,  $p = NS$ ). If considering non-extensible synthetic tubes we observe that RV-to-AP improvement is significantly worse to the rest of the group (mean 7,07 vs 0,17,  $p = 0,005$ ). When focusing on outflow tract diameter, results did not differ in homograft group and patch group. In contrast, diameter did play a role in those patients having a synthetic tube, with a cut-off at 20 mm diameter. Below 20 mm, relieve of outflow tract gradient was significantly worse than for bigger conduits.

**Discussion:** Pulmonary valve insertion is efficient in all type of RVOT reconstruction at least in short term. The diameter of the conduits did not play a role in RVOT obstruction relief as long as surgical substrates are homografts or patch enlargement. In patients with prosthetic conduits, size matters. In non-extensible synthetic tubes results are worst. Reduced distensibility and progressive diameter reduction may lead to not consider these patients as good candidates for this procedure.

#### P-169

##### **Novel Technique to reduce the Size of a Fontan Diabolo Stent Fenestration**

Anderson B.W., Bhole V. Desai T. Mehta C. Stumper O.  
The Birmingham Children's Hospital, Birmingham, United Kingdom

**Objectives:** To develop an effective catheter technique to reduce the size of a Diabolo stent fenestration in the failing Fontan circulation.

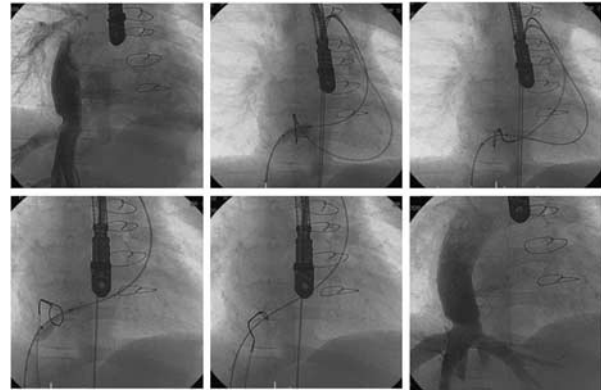
**Background:** Diabolo stent fenestration is employed by many centers in the treatment of the failing Fontan patient. With subsequent recovery exercise tolerance may be impaired by significant desaturations secondary to the right to left shunt across the fenestration. Complete fenestration closure carries the risk of recurrence of the initial symptoms and, hence, reduction of the size of fenestration should be the preferred technique.

**Methods:** 28 patients with a failing Fontan circulation (16 early, 12 late) underwent Diabolo stent fenestration for relief of symptoms. Five of these patients remained very limited by severe desaturation, even at rest, after complete recovery from symptoms. Further cardiac catheterization with crimping/reduction of the size of

the waist of the stent was carried out using a technique whereby a snare catheter was placed over the waist of the stent aided by an arterio-venous guidewire loop and a balloon catheter placed within the stent.

**Results:** All five patients had successful stent reduction with improvement in saturations, whilst still maintaining a small residual fenestration. Oxygen saturations rose from 70–82% [mean 76%] to 81–90% [mean 86%] ( $p < 0.05$ ). No significant changes in the pressures were observed. No complications were encountered.

**Conclusion:** This novel technique of reduction of a diabolo stent fenestration in a failing Fontan circulation avoids implanting further devices in the circulation and maintains the option to re-dilate the stent should symptoms recur.



#### P-170

##### **Surgical Correction of HOCM in Patients with Extreme Hypertrophy and Septal Myocardial Fibrosis.**

Borisov K.V.

Medical Center of the Executive Office of the President of Russian Federation, Moscow, Russia

**Objective:** The mechanism of sudden death in HOCM is ventricular tachycardia/fibrillation emanating from areas of fibrosis. The classic Morrow technique for HOCM in patients with simultaneous LVOT midventricular and RVOT obstruction combined with extreme left ventricular hypertrophy is not effective. A new technique of HOCM surgical correction in patients with severe hypertrophy and myocardial fibrosis was proposed.

**Methods:** The excision of the asymmetrical hypertrophied area of the interventricular septum (IVS) causing LVOT and RVOT obstruction simultaneously was performed from the conal part of the RV corresponding to the zone obstruction of LV. This excision was carried out on the right side of the IVS anterior of the Lancisi muscle and not trough the whole IVS thickness. The presented excision of IVS allows to avoid the damage to the right branch of the His bundle. The areas of septal myocardial fibrosis were removed corresponding to the zone of delayed enhancement (DE) imaging.

Septal myocardial fibrosis was detected by cardiovascular magnetic resonance (CMR) with DE imaging after gadolinium infusion.

Three patients with biventricular obstruction, extreme hypertrophy (NYHA Class 3) and episodes of ventricular tachycardia (VT) underwent this procedure. VT was assessed using Holter monitoring. Ages ranged from 15 to 23 years. The follow-up period was 28  $\pm$  6 months.

**Results:** Two patients were free of symptoms (NYHA class 1) and one patient had only mild limitations. The mean echocardiographic

LVOT gradient decreased from 86,0 ± 8,0 to 9,8 ± 2,1 mmHg, the mean value of gradient in RVOT reduced 43,3 ± 5,5 versus 4,1 ± 1,5 mmHg. Echocardiographically determined septal thickness was reduced 34,0 ± 3,0 versus 17,0 ± 1,0 mm. Sinus rhythm without block of His bundle right branch was noted in all patients after surgery. VT was not registered. The macroscopic examination of the myocardial specimens showed massive scarring. Microscopy investigation showed areas of disarray and scars.

**Conclusion:** This technique of HOCM surgical correction provides the effective elimination of LVOT and RVOT obstruction simultaneously in patients with extreme hypertrophy as well as the reduction of IVS thickness and precise removal of the areas of septal fibrosis.

#### P-171

##### **Operative Treatment Analysis of Patients with Anomaly Ebstein and Determination of Unfavorable Predictors for Surgical Repair.**

*Savitskaya N.V., Matushevich I.D.  
RSPC "Cardiology" CCSC*

**Introduction:** There are 3 concepts of surgical treatment of Ebstein's anomaly (AE): single-ventricle repair, double-ventricle repair, one-and-a-half-ventricle repair.

The choice of surgical tactics should depend on the adequate assessment of tricuspid valve anatomic structure (TV) and right ventricle assessment (RV) functionality.

**Materials and methods:** Today there are 84 patients with AE followed up at CCSC. 34 among them underwent surgical operation from 1997 till 2009. Totally 48 operations were made, 5 of them were repeated. All operated patients were divided into 3 groups: 1 group—mild form—13 patients, 2 group—moderate severe form—15 patients, 3 group—form of AE combined with other CHD—6 patients. The following aspects were assessed: clinical data, age, gender, form of AE, cardiothoracic ratio, ECG, ECHO.

The results of the performed operations were considered and analyzed. The predictors of outcome were determined.

**Study results:**

1. Patients' functional class improved: after operation 70,2% of patients were classified as 2 FC according to NYHA.
2. 71% of patients have lower tricuspid valve insufficiency.
3. Improved RV and LV functioning—73%.
4. High death rate among the patients with moderate and severe forms—46%.
5. High death rate among the patients of the postneonatal age—50%, older than 18 years—55%.
6. High death rate among the patients with visual cyanosis (saturation lower than 90%)—5%.
7. High death rate among the patients with cardiothoracic ratio above 65%—53%.

The following echocardiographic and clinical predictors were determined:

**Echocardiographic predictors:**

1. distal attachment of anterosuperior TV
2. displasia of RV (thinning of anterior wall of RV: twice smaller than normal, autopsy fibrosis of RV wall)
3. LV compression by dilated RV
4. RA+aRV>RV+LV+LA findings, death risk growing with rate over 1,5.
5. RV, LV dysfunction.
6. RV outflow tract obstruction

**Clinical predictors:**

1. cyanosis
2. dyspnoea
3. hepatomegalia
4. hemodynamically significant arrhythmia
5. paradoxical embolism, cerebral abscess

**Conclusions:** Future studies and long-term patient follow-up with AE will facilitate the search of new surgical approaches for such cases. Repair of AE still remains a very disputable issue in surgery. Each case is unique and much depends on surgeon's experience and competence.

#### P-172

##### **Critical aortic valve stenosis : prognosis and influence of first intervention on outcomes**

*Di Filippo S. (1), D'Agata B. (1), Henaine R. (2), Sassolas F. (1), Ducreux C. (1), Ninet J. (2), Bozio A. (1)  
Pediatric Cardiology, Cardiovascular Hospital Louis Pradel, Lyon, France (1); Cardiac Surgery, Cardiovascular Hospital Louis Pradel, Lyon, France (2).*

The aim of the study was to assess outcomes of neonates with critical aortic valve stenosis, and compare percutaneous interventional valvuloplasty to surgical valvulotomy.

**Material and methods:** Demographic, clinical and echocardiographic data and outcomes of patients diagnosed with critical AVS who underwent intervention within the 1st month of life from 1997 to 2008, were reviewed.

**Results:** 32 neonates (8 females) aged 3.5 ± 4 days (median 3) at diagnosis (6 antenatal diagnosis: 18.7%), underwent surgical valvulotomy (group I: 27) or percutaneous valvuloplasty (group II: 5), at median age 17 days, median weight 3.8 kg. Fifteen presented with heart failure (46.8%), 10 received PGE (31.2%, diagnosed prenatally: 66.7%, p = 0.03), 5 received inotropes (15.6%) and 11 were on mechanical ventilation (34.3%). Aortic valve was : tricuspid (7), bicuspid (12), unicuspid (5) or undetermined (5). Preoperative median SF was 28% and mean LV-aorta gradient 51.5 mmHg. Severe heart failure and 3-cusp aortic valve were more frequent in group II (respectively 60% vs 25% : p = 0.1, and 80% vs 11% : p = 0.006). Age at intervention was lower in cases with antenatal diagnosis (5.2 vs 27.5 days, p = 0.01) and similar between groups I and II. CICU stay and hospital stay were respectively 24 vs 5 days (p = 0.17) and 32.5 vs 11.7 days (p = 0.25) in groups I and II. Mean LV to aorta gradient decreased to 17 in group I vs 36 in group II (p = 0.01) and SF increased to 35% (no difference between groups). Aortic regurgitation was grade 0–1 in group II patients, grade 2–3 in 50% of group I, residual mean gradient 17 mmHg in II vs 37 mmHg in I (p = 0.01). Reintervention occurred in 5 cases, death in 5 (15.6%) at 8 to 423 days after first intervention (median 45 days). There was no difference for death or reintervention between groups I and II. lower in patients who received PGE (p = 0.006).

**Conclusion:** Our experience with aortic valve stenosis in neonates shows low rates of mortality and median-term reintervention, whereas death and residual stenosis are more frequent with percutaneous valvuloplasty performed in the most severely ill patients.

#### P-173

##### **Early measurement of plasma neutrophil gelatinase associated lipocalin (NGAL) to screen for acute renal injury after cardiac surgery**

*Koch A. (1), Glöckler M. (1), Cesnjevar R. (2), Zink S. (1), Dittrich S. (1)*

*Dep. of Pediatric Cardiology (1), Dep. of Pediatric Cardiac Surgery (2), University of Erlangen-Nürnberg, Germany*

**Objective:** Neutrophil gelatinase associated lipocalin (NGAL) is a new biomarker for acute renal injury. The aim of this study was to evaluate the diagnostic impact of plasma NGAL measurement in pediatric patients after cardiac surgery in daily clinical practice using a new commercially available point-of-care-test.

**Methods:** From May 2009 to December 2009, plasma NGAL concentration was measured by fluorescence immunoassay (Triage NGAL assay, Inverness®) in all patients with congenital heart defects about 2 hours after termination of cardiopulmonary bypass. 135 consecutive patients (74 males) aged 3 days to 37 years (median age 1.1 years) were included. NGAL levels were compared to perioperative data retrospectively.

**Results:** Plasma NGAL levels were between 60 and 525 pg/ml (median NGAL 139 pg/ml, interquartile range [IQR] 92–207 pg/ml). There was no correlation between NGAL and age, sex, time of extracorporeal circulation or aortic cross-clamping, and peak serum lactate concentration. According to age-specific normal values increased serum creatinine levels occurred in 44/135 patients within 3 days after surgery, predominantly in newborns (16/20), and in total 47/135 patients showed an increase of serum creatinine of more than 50%. However, no patient required dialysis. Beyond the neonatal period, patients who developed elevated serum creatinine in the postoperative course (n = 28) had significantly higher NGAL levels immediately after cardiopulmonary bypass (median NGAL 194 [IQR 132–248] vs. 131 [IQR 96–202] pg/ml; p = 0.03). NGAL was significantly correlated to peak serum creatinine (r = 0.327, p = 0.003; patients aged 4 weeks to 10 years), but the older the patients were the weaker the correlation was (r = 0.283, p = 0.006 in patients <15 years; no significant correlation in patients older than 15 years). In neonates, no significant correlation between NGAL and serum creatinine was found.

**Conclusion:** Beyond the neonatal period, measurement of plasma NGAL can help to identify infants and children with acute renal injury early after cardiopulmonary bypass. However, acute renal failure rarely occurs in pediatric cardiac surgery nowadays. Therefore, the diagnostic impact of plasma NGAL measurement routinely performed after cardiac surgery in children is limited.

#### P-174

##### **Brachial arterio-venous fistula in high risk patients for Fontan's operation**

*Quarti A., Oggianu A., Cetrano E., Colaneri M., Baldinelli A., Pozzi M. Ospedali Riuniti, Ancona, Italy*

**Introduction or Basis or Objectives:** Fontan operation is the gold standard in functional univentricular heart. In patients unsuitable for this surgical approach due to poor ventricular function, the brachial arterio-venous fistula can be a valid option to increase pulmonary blood flow and oxygen saturation. This surgical option can lead to an improvement in ventricular function and consequently to Fontan operation suitability. Herein we report our experience with four patients treated with brachial arterio-venous fistula.

**Methods:** Four patients considered unsuitable for a Fontan completion because of poor ventricular function, underwent brachial arterio-venous fistula. The fistula was performed in relation to the handedness of each patient. The brachial artery and basilica vein were accessed just beneath the biceps muscle and a 5 mm Goretex prosthesis was interposed between the vessels.

The choice of the goretex prosthesis was aimed at having a fixed flow through the fistula. Furthermore the goretex shunt is very easy to take down with minimum risk of vessels injury and bleeding.

**Results:** Following the creation of the fistula the mean oxygen saturation increased from 77% to 83,3%, haemoglobin and haematocrit reduced from 20,13 gr/dl to 18,36 gr/dl and from 60,3% to 54,7% respectively. Mean ejection fraction and shortening fraction increased from 38% to 49,5% and from 22,5% to 28,2% respectively. The patients were scheduled for the take down of the fistula and extracardiac Fontan completion. A Gore-tex conduit was used and in two cases a 5 mm fenestration was performed. None of the patients died or experienced a major complication. At a mean follow-up of 14 months they are all clinically in good conditions.

**Conclusions:** Among the original Fontan "commandments", ventricle function is still nowadays a key point for a successful Fontan operation. In patients with poor ventricular function, the fistula may play a role at different levels. It produces an increase in oxygen saturation and in ventricular preload and a reduction in blood viscosity and pulmonary arteriovenous malformations. These factors, not only reduces symptoms of cyanosis, but improve ventricular function as well. This might represent an intermediate step to rescue patients previously considered unsuitable or at high risk for Fontan operation.

#### P-175

##### **Secondary pulmonary valve implantation after repair of Tetralogy of Fallot. Factors influencing the incidence and timing of the reoperation**

*Nagy Z. (1), Wiegand G. (2), Sieverding L. (2), Hofbeck M. (2), Ziemer G. (1) Department of Pediatric Cardiac Surgery, University Hospital Tübingen, Germany (1); Department of Pediatric Cardiology, University Hospital Tübingen, Germany*

**Objectives:** Our aim was to identify demographic, anatomical and operative data associated with the need for secondary pulmonary valve replacement (PVR) after operative repair of TOF.

**Methods:** Follow-up study of 114 patients operated with the diagnosis of uncomplicated TOF between 12.1994 and 12.2008. There were three perioperative deaths. From the 111 survivors n = 60 (54.1%) were operated either via a classical transpulmonary /transanular (TAP), n = 7 (6.3%) via pure transventricular and n = 44 (39.6%) through a pure transatrial approach. The patients operated by the two latter approaches were defined as the non-TAP group (n = 51). Intraoperatively the pulmonary valve was either resected during the transanular repair, or progressively torn with Hegar dilators. Seven patients were palliated preoperatively, either with a shunt (n = 3) or a palliative balloon pulmonary valvuloplasty (n = 4).

**Results:** The non-TAP patients were slightly older than those operated by a TAP approach (12.77 vs. 7.07 months p = 0.177). Over an average follow-up of 72.33 months (SD = 52.94) 23 patients (20.7%) required secondary PVR, 21 homografts and 2 xenografts. The incidence of the PVR was not significantly different between the two groups 16/61 in the TAP vs. 7/50 in the non-TAP group (p = 0.158). However, PVR was required earlier postoperatively in the transanular group than in the non-transanular patients (80.5 vs. 127.5 months p = 0.015). Age alone (independent of the surgical approach) at the primary operation did not predict the need for PVR but infants operated under three months of age were more prone to require the valve implantation earlier (70.6 vs. 109 months, p = 0.031).

5/13 patients who needed postoperative interventions because of peripheral pulmonary stenoses required later a valve implantation which was significantly higher ( $p = 0.018$ ) than the rest of the group. The implantation in these patients was needed also significantly earlier (54 vs. 104.8 months,  $p = 0.013$ ).

**Conclusions:** Secondary pulmonary valve replacement is required in a significant proportion of surgically corrected TOF. Transannular patch repair, as well as age at primary repair under 3 months determined an earlier need for PVR. The presence of postoperative peripheral pulmonary stenosis increases the incidence of PVR, also reducing the age of secondary valve implantation.

#### P-176

##### **Telemetric Pulmonary Artery Banding Helps Growth of Hypoplastic Aortic Annulus in Children with Parallel Circulation**

Enodien B. (1), Comber M. (1), Dave H. (1), Balmer C. (2), Prêtre R. (1)

Department of Congenital Cardiac Surgery (1); Department of Cardiology (2)

**Introduction:** To analyze the impact of telemetric pulmonary artery banding (FlowWatch) on the growth of the Hypoplastic aortic annulus and thereby its potential to influence the type of definitive repair.

**Methods:** Between 2003 to 2009, 10 pts (median age: 7.5 days, range 2–66) with a mix of simple (5) to complex (complex TGA:3; DILV:1; DORV:1) intracardiac malformations, having a Cono-arterial VSD with or without additional VSDs, hypoplastic Aortic annulus and Aortic Coarctation underwent FlowWatch (FW) pulmonary artery banding (PAB) along with Coarctation repair. Median aortic valve diameter and the Z value were 0.65(0.53 to 0.75)cm and  $-2.44(-4.25$  to  $-0.74)$  respectively. Follow-up was performed with serial echocardiography. Median FW implantation duration was 8.5(3–11) months.

**Results:** At 6 months post implantation, aortic annulus increased from 0.65 to 1.1(0.84–1.4)cm with a corresponding Z value from  $-2.44$  to 0.95( $-1.5$  to 3.74) ( $p < 0.01$ ). The eventual aortic annulus diameter surpassed that of controlled matched population (Fig 1). 7 patients underwent a biventricular repair, 2 patients with functional Univentricular heart reached Fontan completion without a need for DKS and 1 patient settled for a 1&1/2 ventricular repair. None of the patients developed sub aortic obstruction upto the time of FW explantation. One patient with DILV underwent sub aortic tissue resection with myectomy during Fontan completion 32 months post FW explantation. The pulmonary valve retained normal function after FW explantation.

**Conclusions:** Telemetric PAB promotes growth of a borderline Hypoplastic Aortic valve annulus thus enabling a physiological correction. Telemetric adjustment allows gradual tightening, thus optimising haemodynamics and avoiding excessive ventricular-arterial gradients. The pulmonary valve is not subjected to extreme turbulence and hence remains pliable; and the pulmonary trunk requires no reconstruction following explantation of the device.

#### P-177

##### **Mid-term results of valve sparing aortic root replacement in children with Marfan or Loeys-Dietz syndrome.**

Rozendaal L. (1), ten Harkel A.D.J. (1), Ottenkamp J. (1,2), Koolbergen D.R. (3), Hazekamp M.G. (3)

Department of Paediatric Cardiology, Leiden University Medical Center, Leiden, the Netherlands (1); Department of Paediatric Cardiology,

Academic Medical Center, Amsterdam, the Netherlands (2); Department of Cardiothoracic Surgery, Leiden University Medical Center, Leiden, the Netherlands (3)

**Objectives:** To examine the results of valve sparing aortic root replacement in children with Marfan syndrome or other connective tissue disease.

**Methods:** Between 2004 and 2009, 8 children below 18 years underwent an elective valve-sparing aortic root replacement. Six of them had Marfan syndrome, the other 2 had Loeys-Dietz syndrome. Preservation of the aortic valve was performed using the reimplantation technique as described by David.

**Results:** Surgery was performed at a median age of 13.5 years (range 0,9 to 17,9 years), two patients were below 5 years of age. Six (75%) were male. The David operation was performed in all patients using dePaulis Valsalva vascular protheses of 20–28 mm. Associated procedures were aortic valve repair in 2, mitral valve repair in 6 (prosthetic ring annuloplasty in 5, anterior leaflet prolaps correction using neo-chordae in 3), and tricuspid valve repair in 3 (ring annuloplasty in 2, de Vega annuloplasty in one). Median follow-up was complete at 1,1 years (range 1 month to 4,1 years). There were no early deaths, 2 patients required re-sternotomy for postoperative bleeding. An intra-aortic balloon pump was temporarily needed in one. Discharge from hospital was at 12,9  $\pm$  8,2 days postoperatively. No patients had thromboembolic complications. A 17 year old patient died 1,3 years after operation due to ventricular fibrillation. Aortic insufficiency is absent or trivial in all at last follow-up.

**Conclusions:** Valve sparing aortic root replacement in children with Marfan or Loeys-Dietz syndrome shows good early results and can also be performed at a very young age. Adult sized aortic root prostheses could be implanted in all. Associated repair of aortic, mitral and tricuspid valves is usual.

#### P-178

##### **Inhaled nitric oxide for postoperative elevated pulmonary arterial pressure after Fontan-type operations**

Georgiev S., Lacheva A., Pilosoff V.

Department of Pediatric Cardiology, National Heart Hospital – Sofia, Bulgaria

**Introduction:** Elevated cavopulmonary pressure after Fontan-type operations results in systemic congestion and hypoxemia, which are challenging to manage. Inhaled nitric oxide (iNO) has been proven effective for treating postoperative pulmonary arterial hypertension in patients with biventricular circulations. Its hemodynamic effects after cavopulmonary operations are not so clear.

**Methods:** All patients treated with iNO after cavopulmonary operations for a 6 year period in a single tertiary pediatric cardiac surgery center were identified. Preoperative catheterization data, cardiopulmonary bypass time and postoperative hemodynamic data were compared between two groups of patients: those treated with iNO and a control group with the same number of patients with comparable diagnoses and interventions but without iNO. Effects of iNO on the cavopulmonary pressure, the transpulmonary gradient, the oxygen saturation and the arterial pressure were examined. All data are presented as medians with range.

**Results:** Fourteen patients were treated with iNO after cavopulmonary operations. Six of them underwent a BDG and 8 patients – TCPC. The mean used dose of iNO was 18 ppm. Compared to the control group the iNO group had higher preoperative pulmonary arterial pressures (PAP) – 17(11–30) vs.

12(10–25)mmHg,  $p = 0.10$ , higher pulmonary vascular resistance (PVR) – 2.04(0.27–6.94) vs. 1.02(0.49–5.20)Wood units,  $p = 0.37$ , longer bypass times – 154(41–218) vs. 91(15–276)min,  $p = 0.13$ . Compared to those from the control group the patients from the iNO group postoperatively had higher cavopulmonary pressures – 19(14–23) vs. 15 (12–16)mmHg,  $p = 0.00$  and transpulmonary gradients – 9(6–17) vs. 6(5–6)mmHg,  $p = 0.00$ , lower arterial oxygen saturations – 79 (69–88) vs. 95 (79–100)%,  $p = 0.00$  and arterial blood pressures – 80 (55–110) vs. 87 (76–100)mmHg,  $p = 0.11$ . Linear correlation analysis showed tendencies for reduction of the cavopulmonary pressure ( $p = 0.13$ ), reduction of the cavopulmonary gradient ( $p = 0.02$ ), elevation of the oxygen saturations ( $p = 0.10$ ), and elevation of the arterial pressure ( $p = 0.00$ ).

**Conclusions:** Inhaled nitric oxide was effective for lowering high cavopulmonary pressure and high transpulmonary gradient and for elevating oxygen saturation and arterial pressure after cavopulmonary operations in our small cohort of patients. According to our data higher risk for elevated cavopulmonary pressures postoperative have patients with preoperative PAP > 17 mmHG and PVR > 2Wood units, and those with longer bypass times (>154 min).

#### P-179

##### **Neurodevelopmental outcome after hybrid procedure in children with functional hypoplastic left heart syndrome**

Liamlahi R. (1), Knirsch W. (1), Bürki C. (2), Prêtre R. (3), Bernet V. (4), Latal B. F. (5), Kretschmar O. (1)

Division of Cardiology (1), Department of Anaesthesia (2), Division of Congenital Cardiovascular Surgery (3), Intensive Care Unit (4) and Child Development Center (5); University Children's Hospital, Zurich, Switzerland

**Objective:** To compare the neurodevelopmental outcome between children with functional hypoplastic left heart syndrome who were operated with the Norwood procedure and children who underwent the hybrid procedure (bilateral pulmonary artery banding and arterial duct stenting), supposed to be a more neuroprotective therapeutic option by delaying open heart surgery.

**Methods:** Twenty-nine children (17 males) born between April 2004 and October 2008 were consecutively enrolled. Twenty (69%) children survived until age one year and were examined with the Bayley Scales of Infant Development II. A standardized neurological examination was performed.

**Results:** Thirteen children (45%) were operated with the hybrid and 16 (55%) with the Norwood procedure. Median age at first open-heart surgery was 5 (0–7) days versus 5.5 (2–52) days ( $p = 0.27$ ). Mortality was 31% in both groups. These children died during early ( $n = 4$ ) respectively late ( $n = 3$ ) postoperative ICU course due to heart failure, or at home ( $n = 2$ ) interstage. In the survivors ( $n = 20$ ) complete duration of hospital stay during hybrid and Norwood procedures was similar in both groups (median 48 (37–204) days in the hybrid group versus 48 (26–90) days in the Norwood group;  $p = 0.37$ ). Complete duration of intensive care unit stay was shorter in the hybrid group compared to the Norwood group (median 14 (6–25) versus 19 (8–43) days;  $p = 0.175$ ). In both groups the median PDI (Psychomotor Development Index) at age one year was below the normal range (<85), but there was a trend towards better motor outcome in the hybrid group compared to the Norwood group (Median PDI 65 (50–99) versus 56.5 (49–81);  $p = 0.18$ ). Cognitive and neurological outcome, measured as MDI (Mental Development Index), were encouraging in both groups and did not differ from each other significantly (Median MDI 88 (71–102) versus 93

(65–109);  $p = 1.0$ ; median neuroscore 1 (0–1) versus 1 (0–1);  $p = 0.88$ ).

**Conclusions:** These preliminary data show a potential benefit in neurodevelopmental outcome after one year for children treated with the hybrid procedure, particularly for motor outcome. The results of the cognitive development were satisfactory and comparable in both treatment groups. Larger cohorts and long-term neurological follow up is needed to define whether this trend persists.

#### P-180

##### **Biomechanical properties and application of glutaraldehyde treated human pericardium in congenital heart surgery**

Ozolins V, Smits L., Sikora N., Teivane E., Bergmane I., Kasyanov V., Ozolanta I., Lācis A.

Rīga Stradins University, Rīga, Latvia

**Introduction:** Autologous pericardium can be used in its fresh state, or after chemical fixation with glutaraldehyde solution as a biomaterial for surgical repair of congenital heart anomalies. The aim of experimental section of this study was to investigate changes of biomechanical properties of human pericardium, which was used as patch material for congenital heart surgery, after harvesting and treatment with glutaraldehyde. The aim of retrospective clinical study section is to follow up patients after congenital heart defect correction to find out calcification, aneurysm formation, resorption or septic events associated with patch material.

**Methods:** The protocol of the study was approved by ethics committee of University Children's Hospital. After taken out of a pericardium at the time of operation it was fixed with 0.2% solution of glutaraldehyde for 6 minutes. Age of patients was from 1 month to 3 years. Thickness of the samples was measured by cathetometer KM-6 LOMO. For investigation of biomechanical properties uniaxial tensile tests were performed with testing machine Zwick-Roell Z010 and data processing software Testexpert 11.02. There were set up 18 pieces of fresh human pericardium and 11 pieces of glutaraldehyde-treated human pericardium with size 3,5 mm x 25 mm. Force – elongation curves were recorded at rate 5 mm/min. For clinical study single centre operation and ECHO records from year 1997–2009 was used.

**Results:** There is difference ( $p < 0.05$ ) of ultimate strain ( $\epsilon_{\max}$ ) between fresh pericardium  $\epsilon_{\max} = 0.119 \pm 0.037$  and glutaraldehyde treated human pericardium  $\epsilon_{\max} = 0.098 \pm 0.015$ , respectively. There is difference ( $p < 0.05$ ) of ultimate stress ( $\sigma_{\max}$ ) between non-treated pericardium  $\sigma_{\max} = 3.14 \pm 0.70$  MPa and glutaraldehyde treated human pericardium  $\sigma_{\max} = 6.20 \pm 1.47$  MPa. There is difference ( $p < 0.05$ ) of tangential modulus of elasticity (E) at strain level of 30% between non-treated and glutaraldehyde treated human pericardium:  $98.67 \pm 20.75$  MPa and  $50.25 \pm 16.04$  MPa, respectively. Together in 248 operations treated autologous pericardium was used. No signs of calcification aneurysm formation, resorption or septic events were recognize in follow up one to twelve years after correction.

**Conclusions:** This study shows that human pericardium after treatment with glutaraldehyde became stiffer and stronger. There are no significant complications related with pericardial patch treatment of glutaraldehyde in long term follow up.

#### P-181

##### **Valve insertion for a dysfunctional pulmonary valve secondary to a Primary Congenital Heart Repair**

Dave H (1), Comber M (1), Balmer C (1), Enodien B (1), Nikolaou G (1), Trigotindade P (2), Pretre R (1,2)

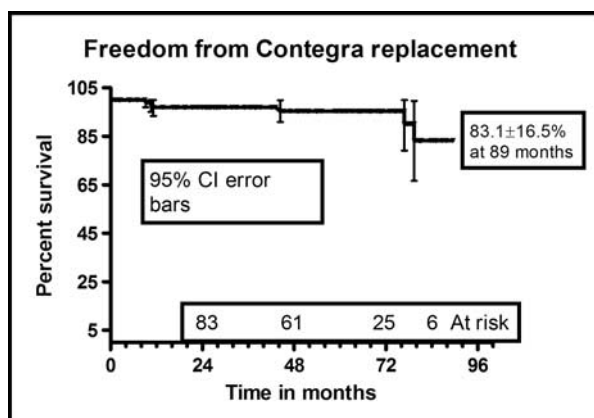
1) University Children's Hospital, Zurich, Switzerland 2) University Hospital, Zurich, Switzerland

**Objective:** To describe our results of valve insertion for a dysfunctional pulmonary valve secondary to primary correction of congenital heart defect.

**Methods:** 110 patients (10–97 kg) having a degenerate pulmonary valve following correction of TOF/DORV/Pulmonary Atresia (90), Truncus arteriosus (9), TGA\_VSD\_PS (6), and Others (5) were reoperated for insertion of a Contegra valved conduit. While central cannulation was used for smaller patients, groin cannulation was used for CPB in older patients (multiple sternal re-entries). Minimum dissection was performed to separate the cardiac mass from underneath the sternum. All prior patch/conduit tissue was excised. Contegra was sutured distally end to end using continuous/interrupted prolene; the proximal anastomosis was performed in a latero-lateral fashion. 29 patients received branch PA reconstructions. Cardioplegic arrest was used in 23 patients to correct residual defects. Conduit placement was performed on beating heart. Online TEE guarded against air embolism. Median CPB time was 133 mins.

**Results:** There were 2 early (severe RV failure in a case of Pulmonary Atresia, second due to Candida mediastinitis) and 2 late deaths, none related to Contegra. There was one major sternal re-entry related bleeding (without cardio-neurological consequence) and no cannulation site complication. One patient needed early re-exploration for haematoma evacuation. All the conduits showed good pulmonary valve function. KM freedom from replacement was  $83.1 \pm 16.5\%$  at 89 months (Figure). At a median follow-up of 54 (0.2–90) months, 6 Contegra were replaced. Three (12, 14, 14 mm) implanted for severe TOF/Pulmonary Atresia were replaced within one year for anastomotic site stenosis. One (18 mm) was replaced after 80 months pre-emptively (needed operation on aortic valve). Two (18, 20 mm) were replaced with a Melody valve at 44 and 77 months for Valvar stenosis. 3 Contegra (14, 16, 22 mm) stenosis were relieved transcatheterally. 17 patients have a mean gradient of median 24 (21–36) mm Hg across the Contegra graft. Two patients needed intervention for iliac artery stenosis at follow-up.

**Conclusions:** Pulmonary valve insertion after a primary intracardiac repair can be performed with minimal morbidity and mortality. These results are reassuring for timely restoration of pulmonary valve competence and correction of residual defects which cannot be addressed percutaneously. These results support the strategy of early re-intervention for residual defects after primary repair.



## P-182

### Risk factors for early morbidity after extracardiac Fontan operation

Ovroutski S., Miera O., Alexi-Meskishvili V., Hübler M., Ewert P., Hetzer R., Berger F.  
German Heart Institute Berlin

**Objectives:** We analyzed risk factors for severe morbidity early after extracardiac Fontan operation (ECFO).

**Methods:** Between 11/95 and 12/09, 130 patients (median age 4.0 years) underwent ECFO. We assumed as preoperative risks systemic right ventricle (RV, n = 47), heterotaxia (Hx, n = 22), pulmonary artery index  $< 200 \text{ mm}^2/\text{m}^2$  (PAI, n = 45), mean PAP  $> 13 \text{ mmHg}$  (n = 25), systemic ventricular EDP  $> 10 \text{ mmHg}$  (n = 20), oxygen saturation  $< 70\%$  (SaO<sub>2</sub>, n = 24), adult age ( $> 16$  years, n = 13) and very low age ( $< 2$  years, n = 50) at surgery. Bypass time (Bt)  $> 60$  min. (n = 33) and cardioplegia (n = 23) were analyzed as intraoperative risks.

**Results:** Hx showed a correlation with early mortality (n = 6 of 8 deaths, 4.7%,  $p < 0.001$ ), as did adult age (3 of 13 adults,  $p = 0.001$ ). Low cardiac output (n = 25 of 130) was developed more frequently by pts. with RV (17 of 47 RV, vs. 8 of 83 left ventricle (LV),  $p < 0.006$ ), pts. with Hx (11 of 22 Hx vs. 13 of 108  $p = 0.001$ ), with low SaO<sub>2</sub> (6 of 11 vs. 18 of 119,  $p = 0.028$ ) and adults (n = 6,  $p = 0.035$ ). Renal failure (n = 17) was observed prevalently in pts. with RV (n = 13,  $p < 0.001$ ), with low SaO<sub>2</sub> (n = 6,  $p = 0.007$ ), with Hx (n = 6,  $p = 0.009$ ) and in adults (n = 5,  $p = 0.001$ ). Mechanical ventilation  $> 24$ h (n = 49) was necessary in pts. with RV (n = 28,  $p < 0.001$ ), with Hx (n = 16,  $p = 0.011$ ) and with low PAI (28 of 45,  $p = 0.005$ ). Prolonged ( $> 10$  days, n = 48) pleural effusions or ascites occurred more often in pts. with low SaO<sub>2</sub> (n = 9,  $p = 0.027$ ) and with Hx (n = 15,  $p = 0.048$ ). Tachyarrhythmia (n = 23, 20%) occurred more frequently in pts. with RV (n = 18,  $p < 0.001$ ), with Hx (n = 9,  $p < 0.002$ ), with low PAI (n = 12,  $p = 0.026$ ) and with Bt  $> 60$  min (20 of 82,  $p = 0.027$ ).

Multivariate analysis revealed Hx as a risk for early mortality. RV, Hx and low SaO<sub>2</sub> were found to be risk factors for low cardiac output. Pts. with RV and low SaO<sub>2</sub> were at risk for renal failure. **Conclusions:** Patients with heterotaxia, systemic right ventricle and very low preoperative oxygen saturation are at high risk for and severe morbidity after ECFO. Appropriate preoperative patient selection and special perioperative and postoperative management are essential for optimal outcome.

## P-183

### Medium-term outcome following Ross/Ross-Konno operation in very young children

Maiya S., Naqvi N., Damda F., Chubb H., Moledina S., Sethia B., Shore D., Slavik Z.  
Royal Brompton & Harefield Foundation NHS Trust, London, UK.

**Introduction:** Ross or Ross-Konno operation has been used widely for surgical correction of left ventricular outflow obstruction in older children and adults for several decades now. Experience with the use of these procedures and their outcome in very young children remains limited.

**Methods:** We reviewed retrospectively hospital records of 16 patients younger than 5 years of age who underwent Ross or Ross-Konno operation in a single tertiary paediatric cardiac centre between June 2002 and November 2007.

**Results:** Age of the patients ranged between 1.2–53.5 months, median 19.7 months (7 patients younger than 12 months) at the time of their operation. Indications for surgical intervention



included aortic valve stenosis in 7 patients, aortic regurgitation in 5 patients, and mixed aortic valve disease in 4 patients. Fourteen patients (88%) had preceding treatment to their left ventricular outflow (balloon valvuloplasty in 10 patients, aortic valve surgery in 4 patients) 10 months (median) before the Ross or Ross-Konno operation. Ross operation was performed in 11 patients (69%) and Ross-Konno operation in the remaining 5 patients. Following Ross-Konno operation there were 2 early postoperative deaths (12% of entire cohort), 2 patients required permanent pace-maker insertion, and 2 have clinically important pulmonary hypertension at most recent follow-up. None of these sequelae occurred following Ross procedure. Re-operation on aortic and pulmonary valves was performed in only 1 patient (68 months after initial procedure). Median postoperative follow up was 27 months, range 0.3–78 months (one patient was lost to follow up following hospital discharge). Actuarial survival was 87% (CI 57–97%) at 5 years. Echocardiographically detectable aortic valve incompetence was present in 6 patients (46% – only 2 with moderate degree of incompetence) out of 13 patients free of re-intervention. Majority of the surviving patients are in NYHA class I (71%) with the remaining patients in NYHA class II.

**Conclusions:** Our results show that Ross operation can be performed safely even in the very young patients with excellent medium-term outcome. Complex left ventricular anatomy requiring Ross-Konno operation is associated with much higher risk of postoperative death and/or complications.

#### P-184

##### The Use of Arginine Vasopressin in Postoperative Norwood Patients

Burton G., Goot B., da Cruz E., Kaufman J.

University of Colorado Health Sciences Center Denver, CO, USA

**Introduction:** Neonates undergoing Norwood palliation may experience postoperative hemodynamic instability. Low cardiac output syndrome, pulmonary overcirculation, and systemic vascular dysregulation may all necessitate an escalation of inotropic and vasoactive support. Arginine Vasopressin (AVP), an endogenous peptide with vasoactive properties, may be useful in supporting these patients with hemodynamic instability refractory to conventional vasodilator and inotropic therapy.

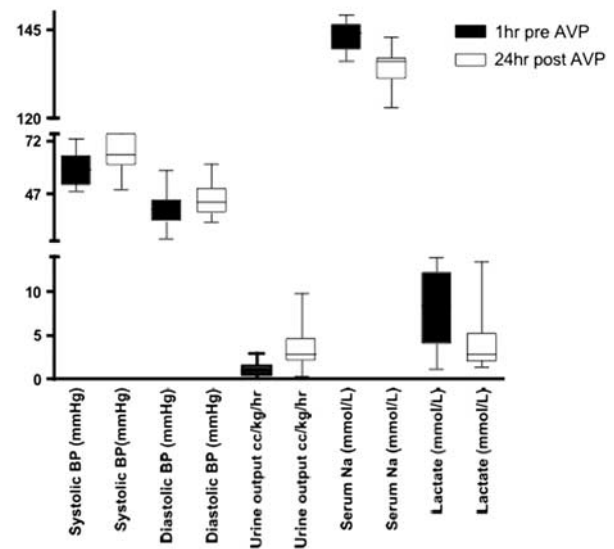
**Methods:** A retrospective evaluation of all neonates at this institution between 2007 and 2009 who underwent a Norwood procedure and received AVP postoperatively. Patients were evaluated for 24 hours after the initiation of AVP with respect to changes in inotrope score, heart rate, blood pressure, SpO<sub>2</sub>, serum pH, serum lactate, serum sodium, urine output, and regional oxygenation.

**Results:** 33 children underwent the Norwood procedure, 23 (70%) of whom also received AVP for a minimum of 12 hours (91% in excess of 24 hours). The initiation of AVP was at the discretion of the attending physician; general indications included refractory hypotension or increasing serum lactate. The mean dose of AVP over 24hrs was  $0.0005 \pm 0.0003$  units/kg/min. There was no difference one hour before AVP and 24 hours after AVP in terms of heart rate, inotrope score, or SpO<sub>2</sub>. However, systolic blood pressure increased from  $58 \pm 8$  mmHg to  $68 \pm 11$  mmHg,  $p = 0.002$  and diastolic blood pressure increased from  $39 \pm 7$  mmHg to  $44 \pm 7$  mmHg,  $p = 0.018$ . Serum lactate decreased from  $7.9 \pm 4.3$  mmol/L to  $3.7 \pm 2.8$  mmol/L,  $p = 0.0039$ . Arterial pH increased from  $7.36 \pm 0.07$  to  $7.46 \pm 0.06$ ,  $p < 0.0001$ . The urine output increased from  $1 \pm 0.8$  cc/kg/hr to  $3.6 \pm 2.3$  cc/kg/hr,  $p < 0.0001$  while the serum sodium level decreased from  $143 \pm 4$  mmol/L to  $134 \pm 5$  mmol/L,

$p < 0.0001$ . 65% of patients also had regional saturation data assessed by NIRS; within 6 hrs of initiating AVP cerebral NIRS increased from  $43 \pm 6\%$  to  $46 \pm 7\%$   $p = 0.0005$ , and regional NIRS increased from  $36\% \pm 11$  to  $43\% \pm 15$ ,  $p = 0.0005$ .

**Conclusion:** Following Norwood Palliation, neonates in our institution have an improvement in blood pressure and markers of end-organ perfusion that is temporally associated with AVP administration. Further study is required to ascertain the efficacy of AVP in this population.

Hemodynamic and Serum Markers 1hr before and 24hrs after AVP administration



#### P-185

##### Early postoperative outcomes following surgical repair of complete atrio-ventricular septal defects (CAVSD): Is trisomy 21 a risk factor?

Desai A., Maiya S., Comitis G., Vyas D., Slavik Z., La Rovere J.  
Royal Brompton Hospital NHS Foundation Trust London,  
United Kingdom.

**Introduction:** Trisomy 21 was shown to reduce risk of reoperation due to lower incidence of dysplastic atrio-ventricular valves in CAVSD. Little is known about the effect of trisomy 21 or atrioventricular valve dysplasia on early postoperative outcome following CAVSD repair.

**Methods:** Retrospective review of data of all children undergoing surgical repair of CAVSD in a single tertiary paediatric cardiac centre between January 2004–December 2009.

**Results:** Sixty seven patients with trisomy 21 (63% – Group 1) and 40 patients without trisomy 21 (Group 2) were operated on. The median age (months) at operation for Group 1 and 2 was  $4.2$  (IQR 3.5, 7.7) vs  $5.75$  (IQR 3.9, 9.3), respectively ( $p = 0.14$ ). Preoperative echocardiogram showed similar incidence of significant left and right AV valve dysplasia and/or regurgitation between Group 1 and 2 (Table). Cardiopulmonary bypass time, inotrope score on admission and at 24 hours, and incidence of low cardiac output state or postoperative chylothorax were not different between Group 1 and 2. On admission and at 24 hours, there was no difference between the groups in arterial pH, lactate, and base excess values. Length of ventilation (hrs) and ICU stay (days) in Group 1 and Group 2 were  $41$  (IQR 19.5, 61) vs  $27.5$  (IQR 15, 62) ( $p = 0.2$ ) and  $2$  (IQR 1.3, 3) vs  $2$  (IQR 1, 3) ( $p = 0.9$ ) respectively. No difference was found in the number of patients requiring epicardial pacing on admission ( $p = 0.6$ ) and at

24 hrs ( $p = 0.4$ ) postoperatively. Early reoperation rate was similar in Group 1 (2.9%) and Group 2 (5%). There was no early postoperative mortality.

**Conclusions:** Trisomy 21 status was not a risk factor for increased early postoperative complications. Contrary to previously published data, we did not confirm higher incidence of dysplastic atrioventricular valves in children with normal chromosomes. In the current era, surgical repair of CAVSD can be achieved with very low mortality and morbidity.

	Group 1 (Trisomy 21)	Group 2 (Non-Trisomy 21)
Left AV valve dysplasia	5	5
Right AV valve dysplasia	1	0
Left AV valve regurgitation	Mild = 15 Moderate = 5 Severe = 2	Mild = 8 Moderate = 2 Severe = 0
Right AV valve regurgitation	Mild = 19 Moderate = 1 Severe = 1	Mild = 19 Moderate = 0 Severe = 0

### P-186

#### Tetralogy of Fallot: changing practice in perioperative care

Desai A., Sainath R., Davies L., Slavik Z., Halley G.  
Royal Brompton Hospital NHS Foundation Trust, London, United Kingdom.

**Introduction:** Many paediatric cardiac surgical procedures have low mortality and significant changes in postoperative outcome can only be measured by analysis of morbidity. We analysed early postoperative data for complete repair of tetralogy of Fallot (TOF) in children <2 years of age (2002–2009) to evaluate whether changes introduced following a benchmarking exercise in December 2004 have been sustained over the subsequent 5 year period.

**Methods:** Group I data (Jan'02–Dec'04) was used for benchmarking against data from other centres as well as published literature. On presentation five changes to perioperative practice were proposed (below). The first 4 were specifically targeted at

the TOF perioperative pathway. The reduced use of post-operative human albumin was proposed for all elective surgical cases. 1. ↑ milrinone intraoperatively (MILR) 2. ↓ high dose adrenaline >0.1 mcg/kg/min (ADREN) 3. ↓ high dose dopamine and dobutamine >10 mcg/kg/min (DOPA-DOB), 4. ↑ intraoperative modified ultrafiltration (MUF) and 5. ↓ post-operative human albumin solution (HAS) as volume expander. We demonstrated improved outcomes in the 2 years post audit (Jan'05–Dec'06). We analysed the subsequent 3 years (to Dec'09) to assess whether change in practice and improvements in outcome have been sustained.

**Results:** There was significant change in all five targeted areas which was sustained on analysis over a five year period. In 2009 the use of Early postoperative outcomes improved as illustrated in Table 1. There was no significant change in patient age at operation. Improvements have been seen year on year and in 2009 the use of MILR and MUF increased to 100%, and the use of ADREN, DOP-DOB and HAS fell to zero.

**Conclusions:** The data demonstrates that by targeting specific areas in the perioperative management of TOF repair we were able to create an effective and long lasting change in practice, with associated benefits in early postoperative outcome. By focusing on the details of a lesion specific perioperative pathway and having a targeted approach it is possible to shorten ICU length of stay and associated cost per patient.

Table 1. Values in Median (IQR) or number (percentage)

	Group I (n = 64)	Group II (n = 56)	P value (Group I and II)	Group III (n = 75)	P value (Group II and III)
MILR	6 (9%)	28 (50%)	0.0001	56 (75%)	<0.0001
MUF	None	31 (55%)		68 (90%)	<0.0001
ADREN	9 (14%)	2 (5%)	0.05	1 (1%)	0.6
DOPA-DOB	15 (24%)	2 (3%)	0.002	None	0.2
HAS	54 (84%)	19 (40%)	<0.0001	13 (17%)	0.03
Age (years)					
Hospital mortality	0	0		0	
ICU Stay (days)	3 (2, 6)	2 (1, 4)	<0.0001	1.5 (1, 3.5)	0.8
Paced for > 24hrs	17 (26.5%)	12 (28%)	0.6	10 (13%)	0.2
Junctional ectopic tachycardia	16 (25%)	8 (12%)	0.17	10 (13%)	1