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Cardiology in the Young

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Session 1: Arrhythmias

O-1

"What you see is what you get": Integration of 3-D-MRI and CT data in electroanatomical mapping for ablation of complex arrhythmias in patients with congenital heart disease [CARTO MERGE]

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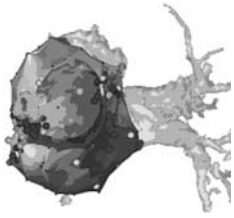
Introduction: Electroanatomical mapping has improved the results of ablation for reentrant tachycardias in pts. after surgical repair of CHD. Now 3-D-image integration is a newly available tool allowing real time navigation of ablation catheters in an individual 3-D-model of the target chamber. We report about our first experience with CARTO MERGE® in ablation of intraatrial reentrant tachycardias [IART] and scar related reentrant VT pts. after repair of CHD.
Methods:
Imaging – CT: 64 line Siemens, 0.5 mm slices/MRT: Siemens Sonata 1.5 T. Segmentation with software integrated in Carto Merge. Carto tachycardia-map and linear ablation after analysis of the reentrant circuit.
Patients – n = 9 (3 × Senning repair of dTGA, 4 × TOF (1 DORV), 1 × M. Ebstein with TK replacement and ASD patch, ×ASD II patch) with drug refractory reentrant tachycardias.
Results:
Imaging – In 7/8 pts. segmentation and integration of the imaging was easily achieved. In 5/7 the accuracy of the match with the CARTO-Map was adequate (defined as mean deviation <2.5 mm and absolute deviation in target area <5 mm). Limitations were mainly caused by reduced quality of contrast-imaging e.g. in the baffle area of Senning repair.
Arrhythmias – 15 IARTs (CL 287 ± 44 mm) in 8 pts. were induced. In 1/8 no tachycardia, but a substrate map was possible. Clinical VT in 1 pt. with TOF with CL 320 msec was induced and mapped.
Ablation – Successful ablation (clinical IART) in 7/8 with 14 ± 4 RF-applications. In 2/8 cases a biatrial approach was needed for successful ablation. VT was also ablated successfully.
Mean procedure time: 434 ± 78 min.



Conclusion: 3-D-Image integration in CARTO Merge® is feasible in pts. after surgical repair of congenital heart diseases. Optimized imaging remains a challenge for the radiologist. Individualized imaging protocols might improve the situation. The navigation of the ablation catheter in the individual 3-D-model might facilitate mapping and ablation in complex substrates.

O-2

The use of CT – derived data in mapping and ablation of tachycardias in complex congenital heart disease

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Introduction: Complex arrhythmias are a frequent indication for catheter ablation in patients with congenital heart disease, especially after palliative surgery. Previous catheter mapping systems had limitations in depicting the individual anatomy of these patients. This study shows the first results with a new mapping system that implements the individual anatomic dataset derived from multislice computer tomography (MSCT) into the electroanatomical mapping system.
Methods: The new system (Carto-Merge®) was used in 12 patients (pts.) with complex anatomy (6 pts. after Fontan operation, 4 pts. after Mustard operation, 2 pts. following palliative treatment of other heart malformation. The individual anatomical dataset was derived from a contrast-enhanced 64-row MSCT (Multislice-CT 64®). 3D-reconstruction and segmentation process of the cardiac

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chambers were performed, followed by implementation of the dataset into the electroanatomical mapping system. After determination of the anatomical landmarks a surface registration algorithm was performed to align the images. Catheter mapping and ablation was then performed using the fused anatomy.
Results and Conclusion: Using CT-derived anatomy incorporated into an electroanatomical mapping system during the EP study enables an exact depiction of the anatomy of patients with congenital malformations. As correlation between tachycardia and anatomical substrate is now comprehensible, planning and performance of the mapping and ablation procedure is facilitated and enhanced.

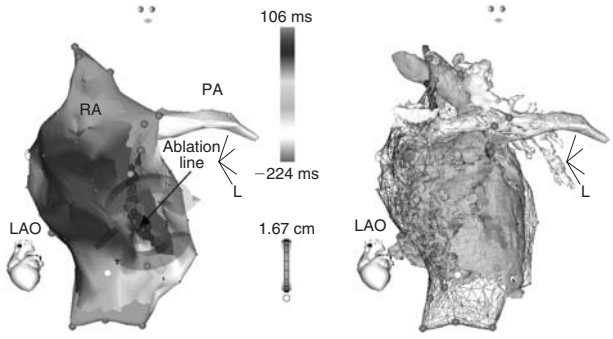

Figure. (Left) LAO view of a conventional map showing incisional flutter (arrows) after Fontan – OP. (Right) Corresponding LAO fused image, which delineates the exact anatomy.

O-3
NavX™ reduces fluoroscopy times in pediatric catheter ablation

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Introduction: Catheter ablation has become an established curative therapy for pediatric tachyarrhythmias. However, exposure to ionizing radiation from fluoroscopy (fluoro) during the procedure is of concern to patients, parents and caregivers.
Purpose: We sought to assess the impact of an impedance-based three-dimensional navigation system (NavX, Endocardial Solutions, Inc., St. Paul, MN) on pediatric catheter ablation procedures.
Methods: We retrospectively analyzed procedural data for catheter ablations of AV nodal reentrant tachycardia (AVNRT) and accessory pathways (APs), comparing procedure durations, total fluoro time, and ablation fluoro time (from insertion of ablation catheter until completion of procedure) between NavX and conventional (conv) mapping.
Results: 119 patients (aged 1–18 years, M:F 65:54) underwent ablation of AVNRT (n = 35), right-sided (n = 44), left-sided (n = 32), or multiple (n = 8) APs using either conv (n = 73) or NavX (n = 46) mapping. Overall success rate was 95%. NavX mapping significantly reduced ablation fluoro times (11.2 + 9.4 min vs 16.6 + 14.6 for conv, $p < .01$) with a trend towards a reduction in total fluoro times (23.6 + 12.3 min vs 27.1 + 16.0 for conv, $p = .09$). Total procedure duration was not different between the two mapping methods (217 + 55 min for NavX vs 210 + 66 min for conv). When analyzed by individual arrhythmia substrate (see Table) there were significant reductions in ablation fluoro times for left and right-sided APs and total fluoro times for right-sided APs, with trends towards reduced fluoro times in multiple accessory pathways.

Fluoroscopy Times (min) mean ± SD.

	AVNRT		Left AP	
	Ablation	Total	Ablation	Total
NavX™	8.6 + 6.5	18.6 + 9.9	9.7 + 8.7	24.7 + 11.6
Conv	8.6 + 5.5	17.6 + 5.3	15.5 + 8.0	28.4 + 9.5
p	NS	NS	.03	NS

	Right AP		Multiple AP	
	Ablation	Total	Ablation	Total
NavX™	10.6 + 8.7	21.3 + 11.9	22.7 + 13.1	39.4 + 8.7
Conv	22.7 + 19.4	32.8 + 21.0	31.0 + 17.8	44.5 + 18.1
p	.004	.01	NS	NS

Conclusions: NavX mapping reduced ablation fluoro times during pediatric catheter ablation, particularly in cases of accessory pathways.

O-4
Improvement in acute outcomes of cryoablation of right septal substrates in children: role of the learning curve and evidence for a dose-response effect

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Introduction: Cryoablation of septal tachyardia substrates in children has become the primary curative modality in selected centers. However, acute success rates from initial series were less than that of established radiofrequency ablation experience. We sought to identify whether acute success rates had improved, and what factor(s) were responsible.
Methods: Retrospective review of all cryoablations for AVNRT or septal accessory pathways (APs) at a single center, comparing procedures in the current calendar year against previous years. Summary statistics, Student's t-test, and Fisher's exact test for proportions were evaluated.
Results: A total of 65 cryoablations (39 for AVNRT, 26 for septal APs) were performed, 18 in the current year and 47 in the 3 years preceding. There was no difference in the proportion of AVNRT and APs between the two groups (11 AVNRT/7 AP current year vs 28 AVNRT/19 AP preceding, $p = 0.2$). Acute success for cryoablations in the current year was 18/18 (100%) compared with 37/47 (79%) in preceding years ($p = .03$). Comparisons between the current and preceding groups showed significant increases in numbers of cryomaps (12 ± 8 vs 8 ± 5, $p = .02$), numbers of cryoablations (4 ± 3 vs 2 ± 2, $p = .01$), insurance lesions (2 ± 2 vs 0 ± 1, $p = .002$), and the use of 6 mm tip catheters (8/18 vs 1/47, $p = .0001$).
Conclusions: This experience supports the premise that, over time, acute success rates of cryoablation for septal tachycardia substrates can reach that of radiofrequency ablation. This improvement appears to be associated with institutional and individual experience, more persistent cryomapping, and a larger cryoablation "dose".

O-5
Cardiac resynchronization therapy in congenital and pediatric heart disease: a retrospective European multicenter study

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Objectives: Data on cardiac resynchronization therapy (CRT) in congenital and pediatric heart disease are scarce. This study collected a multicenter European experience by addressing the members of the Association for European Pediatric Cardiology.
Methods: CRT was applied in 74 pts. aged 0.24–67.5 (median 16.9) yrs with congenital heart disease (N = 59), cardiomyopathy (N = 7), congenital complete AV block (N = 6) and other disease (N = 2) with systemic left (N = 49), right (N = 22) or single (N = 3) ventricular dysfunction and spontaneous (N = 16) or pacing induced (N = 58, median pacing duration 6.0 yrs) systemic ventricular desynchronization using transvenous (N = 28), thoracotomy (N = 21) or mixed (N = 25) lead systems. Concurrent cardiac surgery was performed in 11 (14.9%) pts. Follow-up ranged from 0.0–45.8 (median 8.1) months.
Results: QRS duration decreased from median 160 to 132 ms ($p < 0.001$), Z-score of the systemic ventricular enddiastolic dimension (indexed to a normal left ventricle) from median +3.28 to +1.65 ($p < 0.001$) and grade of systemic AV valve regurgitation from median 1.0 to 0.0 ($p < 0.001$). Shortening fraction of the systemic ventricle increased from mean 16 ± 9 to 23 ± 9% ($p < 0.001$) and ejection fraction or fractional area of change from 30 ± 14 to 41 ± 13% ($p < 0.001$). NYHA class decreased from median 2.0 to 1.5 ($p < 0.001$). Reverse remodeling was better for systemic LV than RV (decrease in the Z-score of the enddiastolic dimension by 2.1 vs 0.67, $p < 0.05$). A total of 9 pts. (12.2%) did not respond to CRT. CRT had to be discontinued in 6 (8.1%) pts. (lead failure = 4, infection = 1, refractory heart failure = 1), 3 pts. died (ventricular arrhythmia = 2, circulatory arrest = 1) and 3 of 8 pts. could be delisted as HTx candidates.
Conclusions: In congenital and pediatric heart disease CRT was more frequently indicated for conventional pacing induced than spontaneous ventricular desynchronization. Major beneficial midterm effects in terms of reverse systemic ventricular remodeling and functional improvement were observed. (RG supported by the Research Project of University Hospital Motol No 00064203/6301).

O-6
Midterm effects of resynchronization therapy on left ventricular performance: a prospective study in 28 patients with congenital heart disease and dilatative cardiomyopathy

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Background: Experience with cardiac resynchronization therapy (CRT) in children and adults with congenital heart diseases is limited. The diagnosis of asynchrony in heterogeneous congenital cardiac malformations and the therapeutic response are still challenging. In this prospective institutional study indices of LV function were studied before CRT, immediately after initiation of CRT and during follow-up using tissue Doppler imaging and conventional Doppler parameters to evaluate the response to CRT.
Patients and Methods: Left ventricular asynchrony was detected in 28 patients (age 1.5–64 years) after surgical correction of congenital heart disease. The diagnosis was based on QRS width and evaluation of electromechanical delay using tissue Doppler derived strain and time to start of left and right cardiac ejection. Resynchronization and LV pacing were achieved via the coronary sinus in 19, using epicardial leads in 9, or both in 8 patients.
Results: Biventricular pacing (BV) resulted in a significant reduction of LV delay in all patients ($p = 0.01$). In contrast to RV pacing, BV and LV pacing was associated with improvement of LV filling time, flow velocity integral in LVOT, Tei index ($p < 0.01$) and increased systolic strain deformation in the LV lateral wall ($p = 0.025$) and diastolic wall velocity excursions in the LV lateral wall ($p < 0.01$). Follow-up 3, 6 and 12 months after the beginning of CRT showed reduction of QRS duration, EDD ($p = 0.04$), and improvement of LVEF, and NYHA Classification ($p = 0.005$). Four patients underwent heart transplantation 3.5 months after CRT (non-responder rate 15%).
Conclusion: CRT is a promising method for treatment of heart failure in patients with CHD independent of the morphology of the systemic ventricle and in pediatric patients with DCM. A prospective randomized multicenter study including a larger number of patients is needed to fully establish the value of CRT in this group of patients.

Session 2: Basic Science

O-7
NFkappaB is required for regulation of the hypoxia-inducible transcription factor HIF-1alpha by thrombin and oxidative stress in pulmonary artery smooth muscle cells

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Thrombotic activity, hypoxia and oxidative stress have been associated with pulmonary hypertension (PH) and vascular remodeling although the underlying mechanisms are not well understood. The hypoxia-inducible transcription factor HIF-1alpha has been shown to contribute to hypoxia-mediated PH, whereas the role of the oxidative stress-responsive transcription factor nuclear factor kappa B (NFkappaB) is less clear. We therefore investigated whether activation of HIF-1alpha and NFkappaB are linked in the response to thrombin and oxidative stress in pulmonary artery smooth muscle cells (PASMC).

Thrombin and oxidative stress, induced by exposure to H_2O_2, increased HIF-1alpha transcriptional activity, nuclear translocation as well as HIF-1alpha protein and mRNA levels and HIF-1alpha promoter activity, and antioxidants prevented these responses. Thrombin and H_2O_2 also stimulated NFkappaB transcriptional activity and nuclear translocation. Interestingly, inhibition of NFkappaB attenuated HIF-1alpha protein and mRNA upregulation as well as HIF-1alpha promoter activity by these stimuli whereas overexpression of the NFkappaB subunits p50 and p65 mimicked the response to thrombin and H_2O_2. Analysis of the

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HIF-1alpha promoter revealed a binding site to NFkappaB. Gel shift analysis demonstrated that NFkappaB can indeed bind to the consensus site in the HIF-1alpha promoter, whereas mutation of the NFkappaB binding site prevented the stimulation of HIF-1alpha promoter activity by thrombin and H_2O_2.

Taken together, these findings provide a novel mechanism whereby the redox-sensitive transcription factor NFkappaB is required for transcriptional upregulation of HIF-1alpha by thrombin and oxidative stress. Since inhibition of NFkappaB and HIF-1alpha reduced the expression of vascular endothelial growth factor (VEGF) and plasminogen activator inhibitor-1 (PAI-1) by thrombin and H_2O_2, induction of HIF-1alpha by NFkappaB may play an important role in promoting pulmonary remodeling processes and thrombosis in PH.

O-8
Human urotensin II upregulates matrix metalloproteinase-2: potential role in pulmonary vascular remodeling

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Vascular remodeling in pulmonary hypertension results from persistent vasoconstriction, smooth muscle cell growth and extracellular matrix (ECM) degradation of pulmonary arteries. The vasoactive peptide human urotensin II (hU-II) is the most potent vasoconstrictor known and has been recently associated with congenital heart disease and vascular remodeling although the mechanisms involved are not clear. Matrix metalloproteinases (MMPs) constitute a tightly regulated group of enzymes that degrade most components of the ECM and basement membrane. The gelatinases MMP2 and MMP9 have been recently implicated to contribute to vascular remodeling processes. Since reactive oxygen species (ROS) have been identified as important vascular signaling molecules we investigated whether hU-II can affect expression and activity of MMP2 and MMP9 in cultivated human pulmonary artery smooth muscle cells (PASMC) and human umbilical vein endothelial cells (HUVEC) via a ROS-dependent mechanism.

hU-II (100 nM) significantly enhanced activity and expression of MMP2 in a time-dependent manner in PASMC and HUVEC, as evaluated by performing zymograms and western blot analyses, respectively. In contrast, MMP9 activity was not detected in these primary vascular cells. Consistently, strong expression of MMP2, but not MMP9, was detected in the media and intima of small remodeled pulmonary arteries in lung tissue samples from patients with congenital heart disease. Furthermore, hU-II increased the generation of ROS by activating NADPH oxidases in PASMC and HUVEC. Inhibition of ROS generation by NADPH oxidases diminished hU-II-stimulated MMP2 activity in PASMC and HUVEC. Furthermore, hU-II rapidly activated p38 MAP kinase and protein kinase B (Akt) in a ROS-dependent manner, whereas inhibition of these kinases prevented hU-II-stimulated MMP2 activity in PASMC and HUVEC.

Thus, our findings demonstrate that hU-II increases MMP2 expression and activity in PASMC and HUVEC involving ROS generation by NADPH oxidases and activation of p38 MAP kinase and protein kinase B. Since MMP2 protein was present in small pulmonary arteries with signs of pulmonary vascular remodeling, this mechanism may play an important role in promoting pulmonary vascular remodeling processes.

O-9
Ascending aortic dimensions: a quantitative trait in first-degree relatives of individuals with congenital left ventricular outflow tract obstruction

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Background: Aortic dilation is frequently found in congenital left ventricular outflow tract obstructions (LVOTO). Dilation of these aortas does not correlate with the severity of the stenosis, raising the possibility that genetic factors are a major determinant of the elastic properties of the aortic wall in LVOTO patients and their family members. We sought to further explore this hypothesis by determining the incidence of aortic dilation and the heritability of aortic diameters.
Methods: We used a database to identify 16 families with at least two first-degree relatives with bicuspid aortic valve, aortic valve stenosis or coarctation of the aorta. We performed echocardiographic examination of the first-degree relatives of the index cases. We sought for any abnormality of the aortic valve and we measured the aortic root as well as the ascending aorta. Echocardiographic measurements were adjusted for height and sex and age. Heritability analysis of the aortic measures was carried out using variance components.
Results: None of the first-degree relatives of the probands were found to have LVOTO. Ten of these individuals (23%), 3 children and 7 adults from 7 families, had significant dilation of the aortic root (n = 2) or of the ascending aorta (n = 8), whereas 34 had normal echocardiographic parameters. Heritability (H^2) of the aortic root diameter in the entire cohort was estimated at 50%.
Conclusion: Incidence of isolated aortic dilation in first-degree relatives of patients with LVOTO is high. Heritability analysis is consistent with the hypothesis of strong quantitative genetic effects underlying the expression of aortic root dilation. These findings suggest that the pathogenesis favoring the development of LVOTO could also affect directly the elastic properties of the aortic wall.

O-10
Array-CGH a novel tool in genetic diagnosis for patients with congenital heart defects

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Array-CGH is a novel diagnostic tool for the detection of submicroscopic chromosomal imbalances. We report the results of array-CGH analysis in 60 patients with a congenital heart defect (CHD) who had, in addition, either developmental delay/mental retardation and/or additional major malformation(s) and/or dysmorphism. No etiological diagnosis could be reached by dysmorphological examination, routine karyotyping or additional genetic or non-genetic investigations.

Array-CGH was performed using a home-made 1 Mb array, with the 1 Mb BAC/PAC set from the Sanger institute. All detected anomalies were confirmed and parents were investigated.

New abnormalities were detected in 11/60 patients (18%). Mosaicism for monosomy 7 was detected in a patient with suspected diagnosis of Fanconi syndrome but normal DEB and mitomycin tests. Among the 10 others, there were 4 interstitial deletions (sizes between 2 and 14 Mb), 1 interstitial duplication (6 Mb), a paternally inherited duplication in chromosome 22q11.2, 1 terminal deletion

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5q (6 Mb), 1 unbalanced translocation (9q/20q) and two more complex intrachromosomal rearrangements involving deletions, duplications and inversions.

Besides these causative alterations, additional changes were detected where the interpretation is currently uncertain, including a maternally inherited duplication on the X-chromosome, and, in three patients, a duplication of chromosome 11qter, an anomaly which has not been described as a polymorphism before.

This study shows that array-CGH is able to detect previously undetectable submicroscopic alterations in a large proportion of patients with a congenital heart defect and a "chromosomal" phenotype. Nevertheless, the interpretation of some of the detected alterations remains difficult and requires further studies. A close collaboration between the cardiologist, clinical geneticist and cytogenetics laboratory will be essential for the successful introduction of this technology into clinical practice.

O-11

Recurrence of congenital heart disease among siblings of patients with Tetralogy of Fallot (TOF)

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Background: The recurrence of congenital heart disease among siblings of patients with Tetralogy of Fallot (TOF) has been estimated at 3%, with high concordance rates.
Methods: Retrospective chart review since 1986 at Sainte-Justine Hospital, Montreal. The study population included families with at least one sib with TOF and another sib with abnormal echocardiogram.
Results: From 601 patients with TOF, we identified 28 siblings with congenital heart disease. In 6 cases (1%), TOF was recurrent. In 22/28 cases, the malformations were discordant: ventricular septal defects (VSD, n = 10), atrial septal defects (ASD, n = 5), persistent ductus arteriosus (n = 3), aortic stenosis (AS, n = 2), aortic root dilatation (n = 2), pulmonary valve anomalies (n = 5) (multiple diagnosis possible). Both cases of aortic root dilatation were associated with atrial septal defect. One case each was encountered for interatrial septal aneurysm, dextrocardia, hypertrophic cardiomyopathy (HCM), supraventricular tachycardia and myocardial fibroma. One family with three affected children (TOF; ASD and aortic root dilatation; VSD) was identified. Two cases of leukemia in siblings were also found.
Conclusions: Most congenital heart disease in siblings of TOF patients is discordant. However, the most common abnormalities found on the echocardiograms correspond to individual features of TOF. Aortic root dilatation, frequently encountered in patients with TOF, can occur as a silent feature in siblings. The association of TOF with AS and HCM in sibs is a novel finding. Future genetic investigations of this patient cohort should provide a unique opportunity to study variable expressivity of mutations.

O-12

Demonstration of a bifactorial determination of congenital heart defects in a family

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Basis: In recent years, the role of several genes was evidenced in human Congenital Heart Defects (CHD): NKX2.5, GATA4, ZIC3 and NOTCH1. In all reported cases, the transmission mode is autosomal dominant or X-linked recessive (ZIC3) with variable expressivity and incomplete penetrance. The basis for this variability is not understood. In a series of 43 cases of transposition of the great arteries (TGA) that were screened for mutation in the ZIC3 gene, we found a mutation in a familial case and unravelled the second factor that explains the differential expressivity of this mutation.
Methods: 43 simple or complex, sporadic or familial TGA cases were proposed to participate to this study. Most cases were d-TGA. After signing an informed consent, DNA was obtained and ZIC3 was screened by dHPLC and/or sequencing.
Results: Out of 43 TGA, one boy with TGA and pulmonary atresia (PA) had a missense mutation that changed a highly conserved triptophan residue to Glycin in the first zinc finger domain (c.762T > G, p.255Trp > Gly). His younger sister who had TGA, PA and mitral atresia had the mutation which was also found in the normal mother. To understand why 2 heterozygous females with the same mutation were either normal or affected, we screened for mutation several other genes already implied in such CHD (LEFTY A and B, EGF-CFC, HINV and Nodal). No mutations were found. The different phenotypes were finally explained by skewed X-inactivation in the daughter who had inactivated in >95% of her cells the normal paternal X chromosome, whereas the mother had a balanced X-inactivation. No siblings of the mother neither the grand-mother had the mutation demonstrating that this was a de novo mutation appearing in the mother.
Conclusions: This is the first demonstration that a combination of 2 factors (gene mutation and skewed X-inactivation) explains adequately the occurrence of CHD in a familial case. It demonstrate the power of such approach in term of genetic counselling since the siblings of the mother had no risk of having children with CHD, neither the oldest daughter of the mother since she had not the mutation.

Session 3: General Paediatric Cardiology

O-13

Ebstein's anomaly: long-term results after modified reconstruction of the tricuspid valve without ventricle plication

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Introduction: Ebstein's anomaly (EA) is a rare congenital heart defect in which the hinges of the septal and/or posterior leaflets are displaced downward to the right ventricle. The anterior leaflet is usually not displaced but is enlarged and sail-like and valve closure is likewise displaced downwards.
Methods: Since 1988 we have operated 37 patients with EA using a modified repair technique of the tricuspid valve. This technique reconstructures the valve mechanism at the level of the true annulus by using the most mobile leaflet for valve closure without plication of the atrialized chamber.

We evaluated our long-term results with regard to functional capacity (NYHA functional class), tricuspid valve function, rhythm disturbances and re-operation rate. We quantified the right ventricular function by measuring flow velocity integral of the pulmonary artery (VTI_{PA}).
Results: All patients survived the operation. There were two hospital deaths (8%) and the late mortality was 2.7%. The mean follow-up period was 59 months (range, 6 months to 13 years). So far no re-operation has been necessary.

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Preoperatively, the majority of all patients were in NYHA classes III and IV (89%). After the first postoperative follow-up examination (2.9 months), 72% were in NYHA class II. Long-term follow-up examinations showed an additional improvement of 43% patients to NYHA class I. Echocardiographic studies demonstrated a significant improvement of tricuspid valve function. No tricuspid valve stenosis was observed. Significant improvement of VTI_{PA} was observed.

Analysis of the postoperative deaths demonstrated that all patients were cyanotic, older than 50 years and were classified as NYHA class III or IV with a severe form of EA.

Conclusions: We conclude that reconstruction of the tricuspid valve without ventricle plication not only achieves good functional results immediately after the operation but that follow-up examinations demonstrate stable or improved functional capacity in the long term. We postulate that incorporation of the atrialized chamber into the right ventricle may contribute to right ventricular contraction and thereby account for the improved functional capacity of the patients.

O-14

Hancock xenograft for right ventricular outflow tract reconstruction – 28 years surgical experience

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Objective: Choosing the right conduit to reconstruct the right ventricular outflow tract (RVOT) remains a difficult surgical task and randomized studies are rarely performed in this field. We retrospectively reviewed our experience using the Hancock xenograft in pulmonary position.

Method: Between September 1971 and February 2005 fifty-two porcine Hancock conduits were implanted (22 females, 30 males), Mean age at implantation was 8.9 ± 11.6 years (median 5.1 years). Diagnoses were: TOF-20, PA/VSD-12, PA/IVS-1, Truncus arteriosus-11, TGA/VSD/PS-8. The main end-points are survival, conduit dysfunction and replacement.

Results: There were 6 early deaths (3 of them before 1980), in-hospital mortality for the last 25 years is 6.8%. Mean follow-up was 11.2 years (maximum 28 years), for 80% completed. Actuarial survival at 15 years was 78.5% ± 2.7%. Univariate analysis identified functional NYHA class and early era of operation as risk factors for hospital death ($p < 0.01$). Mean systolic RV pressure before conduit implantation was 96 ± 19 mmHg. Reoperations for conduit replacement were performed in 15 patients. Mean duration of conduits implanted before 1990 is 14.5 years with a freedom from reoperation for conduit failure at 15 years of 45% ± 3.1%. Independent risk factors for conduit dysfunction are at univariate analysis diagnosis of truncus and younger age at operation ($p < 0.001$). No severe right ventricular dilatation was observed, neither conduit insufficiency even for patients with pulmonary hypertension. Oversizing the tube is not a predictive factor for longer conduit duration. 86% patients are in NYHA I-II at follow-up.

Conclusions: Hancock xenograft conduit can be safely implanted in reconstruction of the RVOT with low early mortality and prolonged durability. Small patients operated for truncus arteriosus are still at risk for conduit failure. Characteristic pattern of valve failure explains the long-term RV function maintenance and the good quality of life for these patients. In this study, Hancock xenograft proved its advantages that are still under debate for other types of conduits.

O-15

Impact of pulmonary regurgitation and right ventricular dysfunction on oxygen uptake recovery kinetics in repaired Tetralogy of Fallot

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Background: Patients with repaired Tetralogy of Fallot (ToF) featuring severe pulmonary regurgitation (PR) and/or right ventricular (RV) dysfunction have reduced exercise tolerance.

Aims: To assess the impact of PR and of RV function on the ability to recover from exercise in ToF patients.

Methods: 61 consecutive patients aged 23.1 ± 12.1 years underwent maximal cardiopulmonary exercise test (CPX), transthoracic echocardiography and magnetic resonance imaging. This data was compared to those of 153 matched healthy subjects.

Results: 19 patients (31%) had severe PR. RV dysfunction was noted in 19 patients (31%). Nine patients (15%) had both severe PR and RV dysfunction. Patients had lower peak oxygen uptake (VO_2), VO_2 slope, carbon dioxide production (VCO_2) slope and O_2 pulse slope ($p < 0.0001$), especially those with severe PR and RV dysfunction ($p < 0.0001$). Heart rate slope was similar between groups. No patient with severe PR and RV dysfunction had a predicted peak $VO_2 > 40\%$. CPX had a high sensitivity and specificity to identify patients with severe PR and RV dysfunction.

Conclusions: In ToF patients, severe PR and RV dysfunction lead to delayed recovery from exercise. CPX can identify patients with severe PR and RV dysfunction and may be useful to guide the pulmonary valve replacement decision-making process.

O-16

Selection of patients for percutaneous pulmonary valve implantation: morphological suitability

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Introduction: Significant pulmonary valve disease can sometimes be treated with percutaneous pulmonary valve implantation (PPVI). The selection of subjects for this novel procedure depends on the anatomy of the right ventricular outflow tract (RVOT). The purpose of this work was to identify morphological sub-groups of RVOT anatomy that may relate to suitability for PPVI and to document the prevalence of these anatomies in our patient population.

Methods: 83 consecutive patients with RVOT dysfunction (54% male, 14 years [5–41], 76% Tetralogy of Fallot or sub-type) who were referred to our centre for magnetic resonance (MR) imaging between March 2003 and February 2005 were studied. The MR datasets were imported into image processing software and reconstruction of the three-dimensional artery volume was performed. A morphological classification was created and correlated with surgical history, subsequent selection for the procedure and outcome.

Results: RVOT anatomy was heterogenous. Five types of RVOT morphology were identified (Figure). Type I was the most common shape (42%) and related to the presence of a transannular patch whilst other Types were more commonly seen in those with circumferential conduits. PPVI was performed in 11 patients. No patient with Type I morphology underwent PPVI. PPVI was successful in 2 patients with Type II morphology, one with Type III, three with

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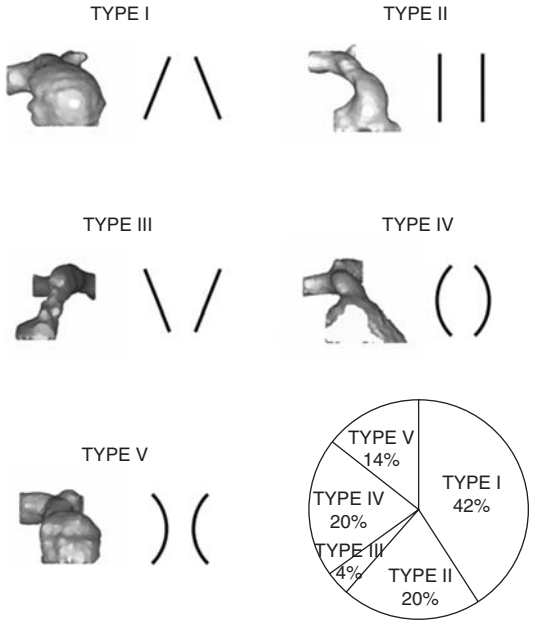
Type IV and four with Type V. PPVI was possible but sub-optimal in one patient with Type III morphology.



Conclusions: RVOT morphology, which can be derived from MR imaging and is often related to previous surgical intervention, is an important aspect of patient selection for PPVI. Only 58% of those presenting with RVOT dysfunction to our institution were morphologically suitable (Types II to V) for PPVI. In addition, patients are unsuitable for this approach if the dimensions of the proposed implantation site are greater than 22 mm due to current device design.

O-17

Balloon expandable right ventricular to pulmonary artery conduit: an animal study

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Objective: Assess the feasibility of using a new vascular stent graft as an expandable valved conduit for right ventricular outflow tract (RVOT) reconstruction in sheep.
Methods: Conduits were prepared by sewing an 18-mm valved conduit (Contegra, Medtronic) inside a vascular stent (CP, Numed). Crimped to 16-mm, it was implanted surgically under extracorporeal circulation (ECC) in 7 sheep (Group A) and without in 5 (Group B). Conduits were then dilated intraluminally using balloon catheters one and two months after the initial procedure. Prior to animal sacrifice, a 22-mm valved stent was electively implanted percutaneously into dilatable conduit.
Results: Five animals from Group A had a complicated postoperative course and needed slow weaned of ECC and inotropic support. Only one recovered. Despite low trans-prosthetic gradient, a balloon dilatation was attempted in 2 trying to reduce the afterload of the failing RV. The remaining 2 animals recovered normally. In animals from Group B, conduits were inserted in an extra-anatomic position. One died acutely due to kinking of the conduit. Balloon dilatations were successfully performed in all surviving animals (n = 7). Diameter increased to 18 and 20-mm after the sequential dilatation. Transcatheter valve implantation was attempted and successful in all animals. At sacrifice, no bleeding was found around the surgically implanted device.
Conclusions: We designed a biological valved conduit for RVOT reconstruction that can be dilated sequentially to follow animal growth. This new device can have tremendous applications in children with congenital hear diseases involving the RVOT.

O-18

Transapical implantation of Valved Stents – or how to prevent reoperation for postoperative pulmonary regurgitation

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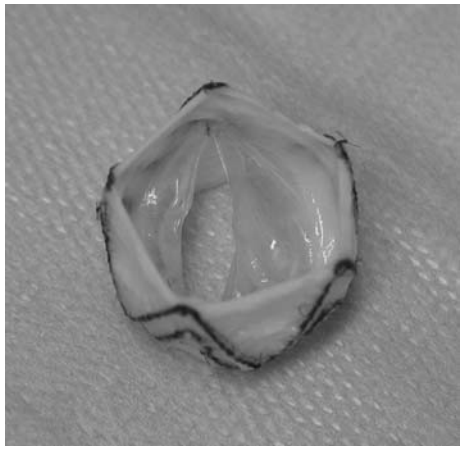
Background: Pulmonary insufficiency remains one of the leading causes for reoperations in congenital cardiac surgery. Recently a new treatment modality, based on the principle of a Valved Stents became available. The currently performed percutaneous approach is limited by the often very small diameter of the access vessel. We propose an alternative off-pump approach via the right ventricular apex to allow for implantation of increased-size self-expanding Valved Stents.
Methods: Valved Stents were implanted off-pump in 6 pigs (48.5 ± 6.0 kg) via the Direct Access transapical approach using a 4 cm subxyphoidal incision and continuous intracardiac echographic and fluoroscopic guidance. Acute Valved Stent function was studied with intravascular and two dimensional intracardiac ultrasound. The invasive valve gradient was assessed with a pull-back pressure catheter. All Valved Stents were tested in vitro before insertion. Macroscopic analysis was performed at necropsy.
Results: Procedural success was 100% (6 out of 6 pigs) at 1st attempt. Mean procedure time was 120 ± 20 minutes. All Valved Stents were delivered to the target site over the native pulmonary valve with good acute valve function. No Valved Stents dislodged into the either the right ventricle or the pulmonary trunk. No animal had significant regurgitation or a paravalvular leak following implantation on intracardiac echo. Planimetric valve orifice was 285 ± 32.0 mm^2. No damage to the pulmonary artery or structural defects of the Valved Stent were found at necropsy.



Conclusions: Six pigs underwent Direct Access Valve Replacement of the pulmonary valve with deployment of a Valved Stent into the native pulmonary annulus. All valves observed for an average of a six hour period showed good, post-implantation valve function.

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Session 4: Fetal Cardiology

O-19

Outcome following diagnosis of isolated tricuspid valve malformations in the fetus

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Introduction: Tricuspid valve displacement or dysplasia has a guarded prognosis despite advances in neonatal and surgical care. Although prenatal diagnosis of tricuspid valve malformations (TVM) is well established there are conflicting reports regarding the prognostic significance of echocardiographic factors.
Methods: From our prospective database, 44 fetuses with isolated TVM were identified between 1995 and 2004 inclusive. The echocardiograms were reviewed by a blinded investigator (RA), and correlated with outcome in 43 fetuses for whom data was available. Fetuses with TVM as part of more complex abnormalities, twin/twin transfusion recipients and those with physiological tricuspid regurgitation were excluded.
Results: Primary referral reason was suspected congenital heart disease in 43 and raised nuchal translucency in 1. Median referral gestation was 23 weeks (range 13 to 37), and number of scans varied between 1 and 4. Tricuspid valve dysplasia was diagnosed in 22 fetuses, Ebstein's anomaly in 21, and unguarded tricuspid valve orifice in 1. Diagnosis was altered following postnatal echocardiogram or postmortem in 4. Atrial tachyarrhythmias were seen in 3 fetuses, who all died. Altogether there were 19 terminations, 9 intrauterine deaths, 5 neonatal deaths and 10 survivors beyond 1 month. Survival was 35% at birth and 23% at 1 month, or 63% and 42% respectively on an intention-to-treat basis. There was no significant change to termination or overall survival rates over time; but there was a significant improvement in survival for live-born babies in the second 5 years compared to the first ($p = 0.02$). Factors significantly associated with increased mortality included cardiothoracic ratio >70% ($p < 0.001$), Celermajer index >1 ($p = 0.001$), reduced antegrade pulmonary valve flow/retrograde duct flow ($p = 0.001$), and RV:LV ratio >1.5 ($p = 0.03$). Diagnosis, referral gestation, LV output, foramen ovale diameter:atrial septal length ratio, functional TV opening:annulus ratio, and degree of TV displacement were not. Five of 6 fetuses with hydrops died, but numbers were inadequate to demonstrate significance.
Conclusions: Despite differences in the pattern of fetal circulation, our findings suggest that the prenatal echocardiographic factors which are of most prognostic significance in TVM are the same as those used postnatally. If adverse echocardiographic factors are present, this should be reflected when counselling parents.

O-20

Prenatal atrioventricular septal defect: outcome and prognostic factors of fetuses without chromosomal abnormalities

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Prognosis of atrioventricular septal defect (AVSD) detected in the prenatal period is known to be poorer than the ones detected in the postnatal period. The purpose of this multicentric prenatal retrospective Doppler-echocardiographic study was to emphasize on the prognostic factors and outcome of AVSD in fetuses without chromosomal abnormalities. Regarding AVSD, the ones with abnormal chromosomes are known to have less severe morphological features.

The population was 141 AVSD. Sixty one (43%) were excluded because they had an abnormal karyotype. Most of them had trisomy 21 (80%). A third of the fetuses with abnormal karyotype had associated cardiac malformations and 20% extra-cardiac malformations. The 80 AVSD fetuses with normal chromosomes were diagnosed at 24.8 GA (±5.4 SD) and maternal age was 29.3 years (±5.3 SD). The cardiac morphological features of AVSD with normal chromosomes were: 80% of complete forms of AVSD, 60% of unbalanced ventricles with a majority of right dominance, mitral regurgitation was detected in 34%, great vessels were normally positioned in 65%; other cardiac malformations were detected in 55% with 13 left heart obstructions and 12 right obstructions. Extra-cardiac malformations were found in 58% of the cases (27 were heterotaxia syndrome, 1 CHARGE association, 2 VACTERL, 1 Ellis-van Creveld and 1 Fanconi syndrome).

Pregnancies were terminated in 54%. Among the continuing pregnancies 16% died in utero. Birth occurred in 36/80 (45%) of the AVSD. Infant death occurred in 6 (16%), 3 died post op (8%). The mortality rate of the initial population was 73% which represents an overall mortality rate of 42% in the non terminated pregnancies. Disparity of the ventricular size was the only statistically significant independent variable ($p < 0.01$) associated with a poor prognosis.

Despite the data implied by surgical series, this study shows that AVSD with normal karyotype is a severe cardiac disease, especially when detected in the prenatal period. Unbalanced ventricles is a significant factor for outcome, which should be taken into account when counselling parents as well as extra-cardiac and cardiac malformations.

O-21

Fetal critical aortic stenosis – ventricular growth and function with and without prenatal valvuloplasty

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In critical aortic stenosis (AS), fetal aortic valvuloplasty has been shown to be feasible. The purpose of this study was to longitudinally assess parameters of ventricular growth and function in these fetuses with and without prenatal intervention.
Patients and Methods: Between 12/2001 and 4/2005 10 fetuses (gestational age: median 24 (17–32) weeks) were diagnosed with critical AS (dilated, poorly contracting LV, signs of EFE, retrograde aortic arch flow). In five fetuses aortic valvuloplasty was attempted (median age: 27 (22–33) weeks and was successful in 3. One of them died in-utero, 2 went to term (34 and 37 weeks), both underwent a successful Ross-Konno procedure postnatally and are alive and well. From the 2 unsuccessful cases 1 died in-utero, the other went to term and received a successful Stage I Norwood palliation due to hypoplastic left heart syndrome (HLHS). Five mothers declined a prenatal intervention, 1 opted for abortion, 4 fetuses were liveborn, 1 died on day 2 of life (prematurity) and 3 underwent successful Norwood palliation due to HLHS. Parameters of ventricular growth and function of surviving fetuses ($N = 6$) were measured until birth.
Results: Group 1 ($n = 4$: no aortic dilation) The Ratio of LV/RV long axis significantly declined from a median of 1.21 (1–1.67) at

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the time of diagnosis to 0.51 (0.48–0.77) before birth and the ratio of mitral valve to tricuspid valve diameter from 0.68 (0.61–0.9) to 0.49 (0.41–0.69). LV area remained unchanged or increased only minimally, as did aortic valve and ascending aorta. LV shortening fraction remained less than 10% in all.

Group 2 (n = 2: successful aortic dilation) In both fetuses antegrade flow in the ascending aorta was established and LV shortening fraction increased significantly from 10 to 22%. In one case with marked LV enlargement and ascites, LV diameter and area decreased until term and hydrops disappeared. In the other case LV dimensions increased.

Conclusions: In fetuses with critical AS there was progressive failure of left ventricular growth and function resulting in HLHS postnatally. Successful prenatal aortic valve dilation resulted in beneficial changes of LV dimensions and function and a two-ventricle circulation postnatally.

O-22
Hypoplastic left heart syndrome: an analysis of factors affecting parental decision-making following prenatal diagnosis

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Background: Hypoplastic left heart syndrome (HLHS) may be diagnosed accurately by fetal echocardiography. Following prenatal diagnosis, options include palliative surgery, non-intervention postnatally and termination of pregnancy. There is little data on factors which influence parental decision-making.

Study Design: All parents attending our tertiary unit have a prospective record made of parental religion, obstetric history, family history, referral indication and attending fetal cardiologist. These factors were analysed to observe which influenced parental decision-making. The period between January 2000 and December 2004 was included. 8147 fetuses were evaluated during this period with 798 cases of congenital heart disease (CHD). Of these, 111 fetuses had HLHS (14% of all CHD). 21 of these cases were excluded because the initial diagnosis was made elsewhere. 90 pregnancies were included in the study. 53 (58%) parents elected to terminate the pregnancy (Group A) and the remainder requested active surgical intervention postnatally (Group B). 90% of referrals were made because of suspected CHD with the remainder because of increased nuchal translucency, fetal arrhythmia or extracardiac abnormality. Rates of TOP did not differ according to referral indication. There was no significant difference in maternal age ($p = 0.87$), gestation at diagnosis ($p = 0.64$) or primigravidas ($p = 0.82$) between Group A and Group B. 14/40 (35%) mothers with poor obstetric history opted for TOP versus 39/51 (78%) without such history ($p = 0.11$). Parent religions included protestant Christian, Roman Catholic, Islam and Hindu, and parents electing for TOP were represented in all religious groups. 90/91 patients were assessed by one of two fetal cardiologists with no difference in TOP rates between them ($p = 0.9$).

Conclusions: There is a trend for parents with a difficult obstetric history to continue with pregnancies affected with HLHS. None of the other factors studied had a systematic impact on parental decision-making. An assumption that some religious groups will not consider termination is not supported by this study. There was no difference in parental decision-making related to the attending fetal cardiologist. Our results emphasise that parental decisions are complex and are not predictable by demographic factors and further studies are clearly indicated.

O-23
Assessment of fetal cardiac function by Tissue Doppler Echocardiography

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Introduction: Despite the progress in the prenatal diagnosis of cardiac disease, the early detection of fetal heart failure remains challenging. In adult cardiology, Tissue Doppler Echocardiography (TDE) is a helpful tool for the quantification of cardiac function and has proven to be sensitive in early stages of systolic and diastolic heart failure. So far, experiences with this method are very limited in the fetal setting.

Methods: In an ongoing prospective study, we examined fetuses with morphologically normal hearts in mothers with non-complicated pregnancies by quantitative color Doppler TDE. Four-chamber views were recorded in cine-loop with a VIVID 7 Dimension scanner (GE), with care being taken to align the examined segment to the ultrasound beam. Systolic and diastolic peak myocardial velocities and isovolumic acceleration (IVA) of septum, right or left free ventricular wall were calculated off-line using Echopac-software (GE). For artefact minimization, the desired position of the sample volume – immediately below the AV-valve annulus – was manually corrected for 3 consecutive cycles, and data of these 3 cycles were averaged. A time limit for raw data acquisition was set at 15 min.

Results: 65 patients (gestational ages 19 to 33 weeks) were so far enrolled in the study. In 83% acquisition of TDE raw data was successful within the time limit. Data analysis was possible in 75 to 92%, depending on the localization studied. Identification of peak velocities corresponding to the different phases of the cardiac cycle was feasible in the ejection phase (S) in 100%, pre-ejection phase (IVA) in 57%, early and late filling phase (E, A) in 90%. Intra-observer variability was acceptable for peak velocity measurements in ejection and filling phase, but high for IVA. Mean velocities were lower than in neonates and increased from second to third trimester. E was lower than A, but increased more than A during gestation.

Conclusions: Quantitative Color TDE is feasible in fetal hearts. However, the method requires ideal conditions and experienced examiners. It is time-consuming, and at the current state of technology, limited spatial resolution and passive myocardial movements considerably affect its reproducibility.

O-24
Right aortic arch in the fetus

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Aim: To examine our experience of the detection of a right aortic arch in the fetus over an eight year period.

Methods: Between February 1998 and December 2005, all patients prospectively diagnosed with a right aortic arch were identified from our database and the videotape reviewed. In addition, the videotapes of 300 normal, 105 abnormal fetal echocardiograms, 122 cases of tetralogy of Fallot and 8 of a common arterial trunk were reviewed. Data such as indication for fetal echocardiography, gestational age at diagnosis, karyotype, nuchal measurement, and outcome were collected.

Results: A right aortic arch was diagnosed prospectively in 53 fetuses and a further 19 on retrospective videotape review. There were 21 examples of isolated aortic arch and 3 thought to have a double arch. A right arch was found in association with additional intracardiac malformations in 45 cases. The detection rate of a

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right aortic arch increased over the study period. The majority of patients were referred for a suspicion of congenital heart disease on obstetric scanning. Mean gestational age at diagnosis was 21 weeks. The karyotype including 22q11 status was known in 38/72 cases. There were 20 karyotypic anomalies, 11 of which were 22q11 deletions, occurring in 2/24 of the isolated group and 9/45 of the complex group. A further 2 cases were suspected but not proven to have 22q11 deletion. There were 27 pregnancy interruptions, 4 intrauterine deaths, 23 live-births, 2 neonatal deaths and 2 patients lost to follow-up. The remaining 13 pregnancies are continuing. Of the 3 with suspected double arch, 2 were confirmed postnatally and 1 was only a right arch.
Conclusion: The diagnosis of a right-sided aortic arch can be made by fetal echocardiography, either as an isolated lesion or in association with other cardiac malformations, from as early as 12 weeks gestation. It can be difficult to distinguish from a double arch. It's increasing incidence in our series probably indicates that the diagnosis was previously overlooked. Karyotyping in the absence of other abnormal findings may be unnecessary, but establishment of 22q11 microdeletion status in those cases with other anomalies is important.

Session 5: Pulmonary Hypertension

O-25
Serum markers in pediatric pulmonary arterial hypertension

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Background: Appropriate serum markers are needed for the non-invasive follow-up of children with pulmonary arterial hypertension. Both N-terminal pro-BNP and uric acid (UA) are known to be increased in adults with heart failure. They might therefore play a role in the monitoring of pediatric patients with right heart failure due to pulmonary arterial hypertension.
Methods: Serum NT-proBNP and UA levels were assessed and correlated with invasive hemodynamic data and follow-up in 26 pediatric patients with pulmonary arterial hypertension (17 with primary pulmonary hypertension and 9 with secondary pulmonary hypertension due to a congenital heart defect) that visited the Beatrix Children's Hospital in Groningen between 1999 and 2004.
Results: Patient characteristics were as follows: 35% male, mean age 9.7 years (range: 1.0–18.3 years). Mean follow-up was 17 months (range: 3 days–7.4 years). Mean NT-pro BNP was 1130 ± 391 pg/ml and mean serum UA was 0.31 ± 0.02 mmol/l. A trend towards a negative relation between NT-proBNP and age existed ($r = 0.37$, $p = 0.08$). Serum UA increased with age ($r = 0.483$, $p = 0.02$) and was higher in females ($p = 0.03$). Serum NT-proBNP and UA did not differ between diagnosis groups. No relations could be established between NT-proBNP or uric acid and 6-minute walking distance. NT-proBNP levels did not correlate with WHO-class, whereas serum UA did ($r = 0.44$, $p = 0.04$). NT-proBNP levels did not correlate with invasive hemodynamic data as pulmonary arterial pressure, pulmonary vascular resistance or right atrial pressure. A positive correlation was found between serum UA and mean pulmonary arterial pressure ($r = 0.572$, $p = 0.016$) and between serum UA and pulmonary vascular resistance ($r = 0.598$, $p = 0.011$). The strongest correlation existed with the inverse of the cardiac index and serum UA ($r = 0.618$, $p < 0.01$). This relation remained highly significant when corrected for age and sex. High serum NT-proBNP and high serum UA predicted decreased survival (logrank-test $p = 0.04$ and $p = 0.02$ respectively).
Conclusions: Serum UA correlated with invasive hemodynamics in pediatric PAH. Both NT-proBNP and UA predicted survival in these patients.

O-26
Decreased left ventricular function of isolated and perfused hearts from adult rats treated with monocrotaline

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Introduction: Pulmonary hypertension may cause a decrease in left ventricular function. This has been variously attributed to alterations in left ventricular geometry, release of trophic substances from the right ventricle, interstitial edema, decreased left ventricular preload, or changes in intracellular calcium. Several animal investigations have examined cardiac function following induction of pulmonary hypertension with the toxin monocrotaline (MCT). Although it has been assumed that MCT has no direct effect on the heart we previously reported that MCT administration results in myocardial inflammation, necrosis, fibrosis and in a diffuse coronary arteriolar angiopathy. We describe here the effects of MCT in the rat on left ventricular myocardial contractile function in the isolated and perfused heart.
Methods: Ten adult Sprague-Dawley male rats (90-days old) received a single injection of MCT (50 mg/kg s.c.). Twenty-one days later the animals were anaesthetized (pentobarbital 40 mg/kg i.p.) and underwent echocardiography (Acuson 128 XP, 7.5 MHZ probe). Their hearts were then excised and perfused in the constant pressure Langendorff mode. Left ventricular pressure (LVP), peak instantaneous rate of pressure increase (+dP/dT) and peak instantaneous rate of pressure decrease (−dP/dT) were recorded. Ten 110-day old rats that did not receive MCT acted as controls.
Results: Pulmonary artery pressure (PAP) by echocardiography in the controls was 13 ± 6 mmHg (SEM). In the MCT group PAP was <20 mmHg in 35%, 20–60 mmHg in 25% and >60 mmHg in 45%. There was no difference in heart rate between the MCT and control groups. LVP, +dP/dT, and −dP/dT were all significantly less in the MCT group compared to controls (LVP 71 ± 10 vs. 106 ± 8 mmHg, +dP/dT 2397 ± 249 vs. 3753 ± 449 mmHg/s, −dP/dT −1751 ± 290 vs. −2778 ± 333 mmHg/s $P < 0.001$). There was no correlation between these variables and PAP.
Conclusions: Our findings indicate that MCT has a significant effect on left ventricular systolic and diastolic function. This left ventricular myocardial impairment likely resulted from our previously described myocardial and coronary arteriolar effects of MCT. The investigation of the left ventricular effects of pulmonary hypertension using MCT must be called into question.

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O-27
Remodeling effects of bosentan in experimental flow-associated pulmonary hypertension

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Background: Pulmonary arterial hypertension (PAH) may be a complication in children with congenital heart disease and left-to-right

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shunts. Beneficial effects of bosentan, a dual endothelin-receptor antagonist, have been demonstrated in patients with PAH. Its exact therapeutic mechanism is not clear.
Methods: Flow-associated PAH was created in 28 adult male Wistar rats by the injection of monocrotaline (60 mg/kg) combined with an abdominal aortocaval shunt one week later. Rats were randomized to treatment with bosentan in rat chow (300 mg/kg/day; n = 14, PAH+) or normal chow (n = 14, PAH−) for three weeks.
Results: Treatment with bosentan improved hemodynamics (systolic pulmonary arterial pressure 48 ± 3 mmHg in PAH+ vs. 59 ± 2 in PAH−, p = 0.003). Right ventricular hypertrophy, expressed as right to left ventricular plus septal weight ratio, decreased (0.37 ± 0.02 in PAH+ vs. 0.44 ± 0.02 in PAH−, p = 0.02) and right ventricular contractility improved (81.4 ± 2.3 s^{-1} in PAH+ vs. 71.5 ± 1.6 in PAH−, p = 0.02). Intra-acinar pulmonary vascular remodeling was partly reversed by bosentan treatment (pulmonary vascular occlusion score: 10.1 ± 1.4 in PAH+ vs. 16.7 ± 1.3 in PAH−, p < 0.001), while myocardial capillary density increased after treatment (2438 ± 65 capillaries/mm^2 in PAH+ vs. 2100 ± 63 in PAH−, p < 0.001).
Conclusion: In this rat model of flow-associated pulmonary hypertension, bosentan treatment beneficially affected cardiac and pulmonary vascular remodeling, leading to improved pulmonary hemodynamics and improved right ventricular function.

O-28
Atrasentan treatment of pulmonary vascular disease in piglets with increased pulmonary blood flow

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Objective: Endothelin-1 (ET-1) plays an important role in the pathophysiology of pulmonary hypertension in patients with left-to-right shunt. ET-1 exerts vasoconstrictive and mitogenic activities mediated by the ETA-receptor on vascular smooth muscle cells. We aimed to study the effect of a selective ETA-receptor antagonist (Atrasentan) in piglets with high pulmonary blood flow and pulmonary vascular disease.
Methods: 17 domestic farm piglets (German Landrace), 4 weeks of age and with a body weight of 14 [10–16] kg (median [range]) were used. After left thoracotomy the left subclavian artery was connected to the main pulmonary artery (BT-shunt). Flow across the BT-shunt was 760 [190–1520] ml/min (Median [range]). The animals were then randomized to two groups: Group I: placebo-treatment (water) (n = 7). Group II: treatment with Atrasentan (2 mg/kgBW/die) (n = 10).
Results: After three months of treatment significant left-to-right shunt was equally present in both groups (Placebo: Qp/Qs = 1.7 [1.0–2.9]; Atrasentan: Qp/Qs = 1.6 [1–2.1]). Mean pulmonary artery pressure (PAP) in the Atrasentan-treatment group was significantly lower (14 [10–17] versus 19 [16–20] mmHg; p = 0.008 (ANOVA on ranks)). Percent medial thickness of pulmonary arteries (D: 50–150 μm) was lower in the Atrasentan group (13.7 [11.7–17.2] versus 17.5 [12.6–25.5] percent wall thickness; (p = 0.034)). Plasma cGMP, nitrate and ET-1 were not different between groups. In lung tissue, quantitative real time PCR for ET-1-, ETA-/ETB-receptor- and Endothelin converting enzyme-1 (ECE-1) – mRNA did not differ between groups. However immunostaining for VEGF as well as quantitative mRNA for VEGF was significantly lower in the Atrasentan treatment group (706 [464–1375] versus 1231 [1185–1520] percent of 18S expression; p = 0.017).
Conclusion: Atrasentan treatment effectively reduces medial hypertrophy in piglets with chronic pulmonary hyperperfusion. This is associated with a decrease in expression of VEGF-mRNA and VEGF immunostaining. Chronic ETA-receptor blockade by Atrasentan may interfere with the expression of VEGF.

O-29
Combined intravenous tezosentan and inhalative iloprost improves pulmonary function after meconium aspiration

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Objective: Meconium aspiration induces a pulmonary lesion similar to the adult respiratory distress syndrome. Iloprost has been shown to improve pulmonary function after meconium aspiration. Since pulmonary hypertension plays a key role in the meconium aspiration syndrome, endothelin receptor blockade associated vasodilation may be effective in this setting. This study was designed to evaluate the effects of iloprost or the endothelin receptor blocker tezosentan alone versus a combination of tezosentan *and* iloprost on pulmonary gas exchange.
Methods: Lung injury was induced in 24 anesthetized pigs by instillation of human pooled meconium. Six animals received intravenous tezosentan 5 mg · kg, another six animals received inhalative iloprost 5 μg · kg^{-1}, six more animals received inhalative iloprost *and* intravenous tezosentan (combination), while six animals served as controls. Circulatory variables and arterial blood and inert gas samples were recorded or taken at 60 and at 180 minutes after meconium instillation.
Results: At the 180 minute measurement, oxyhemoglobin saturation (SaO_2) was higher in all treatment groups when compared to control (tezosentan: 87 ± 6, ilomedin: 84 ± 12, combination: 89 ± 5, control: 70 ± 17%). No intergroup differences between the treatment groups could be observed. Arterial partial pressure of oxygen (PaO_2) was highest in those treated with the combination when compared to control (70 ± 6 vs. 55 ± 5 Torr, $P = 0.04$). This improvement in oxygenation with the combination occurred in the presence of significantly lower pulmonary blood flow heterogeneity (LogSD$\dot{Q}$, 0.77 ± 0.1 vs. 1.46 ± 0.6, $P = 0.04$, vs. control).
Conclusion: The combination of inhaled iloprost *and* intravenous tezosentan but not neither iloprost nor tezosentan alone resulted in improved PaO_2 and SaO_2.

O-30
BREATHE-5: bosentan improves exercise capacity and haemodynamics in Eisenmenger syndrome

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Introduction: Eisenmenger syndrome (ES) is characterized by the development of pulmonary arterial hypertension (PAH) and consequent intracardiac right to left shunt and hypoxemia in

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patients with pre-existing congenital heart disease. No evidence-based treatment approach currently exists for patients with ES. Theoretically, some treatments may worsen the shunt and increase hypoxemia. Since ES is associated with increased endothelin expression, it has been hypothesized that patients may benefit from endothelin antagonism. BREATHE-5 is the first placebo-controlled trial to investigate the effect of bosentan in patients with ES.
Methods: This multi-center, randomized, double-blind, placebo-controlled study evaluated the effect of the dual endothelin receptor antagonist, bosentan, on oxygen saturation (main primary objective, non-inferiority) and haemodynamics (second primary objective, superiority) in patients with WHO functional class III. Secondary objectives included exercise capacity, safety, and tolerability.
Results: Fifty-four patients were randomized 2:1 to bosentan (n = 37) or placebo (n = 17). Similar distribution of ventricular and atrial septal defects was observed in both treatment groups (65%, 22%, bosentan; 71%, 29%, placebo). In addition, 13% of patients in the bosentan but none in the placebo group had combined defects. Mean time from ES diagnosis was 23.7 (13.6) years (SD) in the bosentan and 20.5 (13.0) in the placebo group.

After 16 weeks, the treatment effect on SpO_2 was 1.0% (95% CI = [−0.7, 2.8] > −5, non-inferiority margin), demonstrating that bosentan did not worsen oxygen saturation. As this primary safety endpoint was reached, efficacy analyses were conducted. In comparison with placebo, PVRi was significantly reduced (treatment effect 472 dyn.sec.cm^{-5}; p = 0.04) and exercise capacity, measured by the 6-minute walk test was significantly increased (treatment effect + 53 m; p = 0.008). Four patients discontinued due to adverse events: two (5.4%) in the bosentan group and two (11.8%) in the placebo group. The safety profile of bosentan was comparable to that observed in previous clinical trials in PAH.
Conclusion: In this first placebo-controlled trial in patients with ES, bosentan was well tolerated, improved exercise capacity and haemodynamics without compromising peripheral oxygen saturation. These results suggest that bosentan may be an important new treatment option for patients with ES.

Session 6: General Paediatric Cardiology

O-31
Myocardial scarring determined by delayed enhancement MRI at long-term follow-up after operation of left coronary artery from the pulmonary artery (ALCAPA)

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Background: Patients with an anomalous left coronary artery from the pulmonary artery (ALCAPA) improve their decreased left ventricular (LV) function postoperatively. LV functional studies have been carried out using angiography, echocardiography, and scintigraphy for short to mid-term follow-up. However, little is known about the distribution of myocardial scarring. In particular, the distribution of myocardial scarring in long-term follow-up remains unknown. Delayed enhancement magnetic resonance imaging (DE-MRI) is a modern imaging method and is accepted as the goldstandard for detection of LV myocardial scarring. Therefore the aim of this study was to describe the distribution of LV myocardial scarring after successful operation of patients with ALCAPA after long-term follow-up.
Methods: Since 1974 n = 63 patients with ALCAPA have been operated at our center. Because compliance during a MRI study is assured in patients older than 14 years, we included only patients older than 14 years to the study (n = 33). LV ejection fraction (LVEF) and LV enddiastolic volume (LVEDV) was determined by MRI. LV myocardial scarring was determined by DE-MRI.
Results: Interim results are available for n = 14 patients. No scarring was found in n = 4 patients. These patients all had pre-operative evidence of scarring by Thallium 201 SPECT szintigraphy. Transmural scarring was found in n = 3 patients, diffuse or endocardial scarring in n = 7 patients. No specific pattern of scarring could be determined in these patients. The presence of scarring could not be correlated to the operation method, the age at operation, or time after operation. LVEF was 55 ± 9%. LVEDV was 131 ± 22 ml.
Conclusion: Infant neonatal myocardial scarring may resolve in long-term follow-up. Because the size of scar tissue was relatively small, neonatal myocardial remodeling mechanisms may be very different than adult myocardial remodeling mechanisms. Additionally, the individual coronary collateralization seems to be the major determinant factor for the pattern and the size of myocardial scarring.

O-32
The role of Lipoprotein (a) in pediatric heart transplant recipients

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Objective: Lipid disorders may represent a risk factor for coronary artery disease after heart transplantation. The aim of this study was to assess lipoprotein (a) (Lpa) and lipoproteins blood levels changes after heart transplantation (HT) in children, to identify recipients at increased risk of dyslipidemia.
Methods: Since 1992, Lpa, total cholesterol (TC), HDL-cholesterol (HDL), LDL-cholesterol (LDL) and triglycerides (TG) levels were recorded at HT and every 2 months after HT. Twenty-seven cyclosporine-based immunosuppressed patients more than 1-year post-transplant, were evaluated and classified according to basal Lpa level, in groups: A (<0.3 g/l: 17 patients) and B (≥0.3 g/l: 7 patients). Age at HT was 9.0 ± 6.1 years.
Results: There was no difference between the 2 groups regarding age at HT, previous cardiac disease, length of follow-up, gender, weight, steroid doses, cyclosporine blood levels and basal TC, HDL, LDL, and TG levels. Lpa levels did not vary significantly. Baseline TC (4.2 ± 0.9 mmole/l) increased to 5.1 ± 1.1 at 3 months, 5.2 ± 1.2 mmole/l at 1 year (p = 0.0012) and stabilized beyond 1st year; levels ranged above normal value for age in group B and were higher than in A. HDL increased from baseline 1.06 ± 0.36 to 3rd month 1.44 ± 0.36 mmole/l (p = 0.0019); HDL decrease was observed only in group B (3rd month 1.63 ± 0.28 to 6th month 1.34 ± 0.61 mmole/l, p = 0.05). HDL ranged at the lower limit for age in group B. LDL increased from baseline (2.55 ± 0.76 mmole/l) to 3rd month (3.18 ± 0.93 mmole/l) (p = 0.02) and stabilized thereafter. LDL levels ranged above normal value for age in group B and were higher than in A. TG increased from baseline (1.21 ± 0.55 mmole/l) to 3rd month (3.18 ± 0.93 mmole/l); TG further turned to normal range and did not differ between groups A and B.
Conclusion: Heart-transplant children with basal Lpa > 0.3 g/l display more frequent post-transplant atherogenic lipid profile and might carry an increased risk of graft coronary artery disease. These results would have to be confirmed by larger prospective studies.

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O-33

Efficacy and tolerability of long-term administration of Carvedilol in pediatric patients with chronic heart failure

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Since long term experiences with Carvedilol in children are not reported, we present our clinical experiences with Carvedilol over the last eight years (1997–2005).
Patients and Methods: 31 pediatric patients (17 days up to 19 years) with congestive heart failure due to congenital heart disease ($n = 10$) or dilative cardiomyopathy ($n = 21$) were treated with slowly increasing doses of oral Carvedilol (mean start: 0.09 mg/kg/ day; mean end dose 0.64 mg/kg/day). The mean duration of therapy was 2.6 years (1 month up to 6.5 years). From 1997 until 2005 shortening fraction (FS), clinical symptoms (Ross Score) and natriuretic peptides (BNP or N-BNP) were evaluated.
Results: In 19/31 patients the therapy was determined as effective. In 10/31 patients therapy was stopped permanently after a mean of 2.1 years due to a stable improvement of the clinical status. Two of 31 (6.5%) patients withdrew from therapy due to intolerability. Ten patients died during the additional therapy with Carvedilol. Nine patients are still being treated (mean 5.6 years) with Carvedilol. In 5/31 patients the therapy was additionally installed for bridge-therapy to operation or heart transplantation. Cardiac function (FS: 18 to 23%; $n = 25$; $p < 0.05$) and neurohumoral stimulation (BNP: 513 to 304 pg/ml; $n = 10$ and N-BNP 683 to 554 fmol/ml; $n = 10$; $p < 0.05$) improved under Carvedilol. Clinical symptoms decreased under therapy from 5.5 to 2.8 points (Ross Score; $n = 22$; $p < 0.05$).
Conclusion: Our first clinical data of the long-term administration of Carvedilol indicate that oral carvedilol constitutes an effective and tolerable treatment in addition to standard therapy in pediatric patients with congestive heart failure.

O-34

Left ventricular force frequency relationships are abnormal after orthotopic heart transplantation

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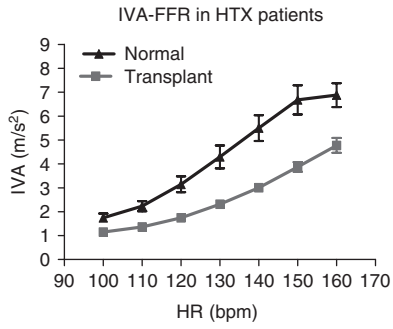
Non-invasive evaluation of myocardial performance in post heart transplantation in pediatric population needs to be established. Despite apparently normal resting cardiac function using standard assessment techniques, patients have reduced exercise capacity post-orthotopic heart transplantation (HTX). We hypothesize this may partly be due to an impaired myocardial force-frequency response (FFR). FFR was studied in patients post-HTX using isovolumic acceleration (IVA) at the time of biopsy. Unlike other conventional 2-D echocardiographic measurements, IVA obtained with Tissue Doppler Imaging (TDI) has been shown to be relatively load-independent index of myocardial performance, which could describe FFR non-invasively. And IVA has been shown to detect rejection in heart transplant.
Methods: 56 studies were done on 43 post-heart transplant patients (aged 7.0 ± 4.7 y at biopsy, and 3.3 ± 2.5 y post-HTX). Patients underwent TDI at a time of endomyocardial biopsy with transvenous atrial pacing at 10 bpm increments up to a maximum of 200 bpm or Wenckebach. IVA, IV velocity (IVV) and systolic wall velocity (SWV) were measured at the LV basal lateral free wall. Data were correlated with rejection grade, basal diagnosis, coronary artery disease, and graft ischemia time.
Results: All patients had normal systolic function at rest. With pacing, TDI systolic indices were depressed in patients compared to normal controls (IVA; $p < 0.0001$, IVV; $p < 0.0001$, and SWV; $p < 0.0001$, 2-way ANOVA). Patients who had multiple studies and significant rejection had depressed FFR at the time of rejection. Graft ischemia time >300 min was found to be associated with depressed FFR.
Conclusion: IVA derived FFR can be assessed during usual surveillance in children after orthotropic heart transplantation. The ventricular myocardial FFR is impaired in post HTX by comparison with normal controls.

Causes of impaired FFR may include prolonged graft ischemia and acute cellular rejection, and IVA FFR may be useful in long term follow up.



O-35

Cardiorespiratory capacity in patients with hypoplastic left heart syndrome (HLHS) after total cavopulmonary connection (TCPC)

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Aim: The study was performed to obtain parameters of cardiorespiratory capacity in children with HLHS after TCPC.
Methods: 26 patients who underwent their surgical treatment from 1996 to 2001 were examined at a median age of 5.5 (3.7–9.3) years at least 1 year after TCPC. The exercise testing was performed on standardized incremental treadmill protocol. We analyzed maximal heart rate, maximal oxygen pulse, oxygen consumption at the anaerobic threshold (VO_2-AT) and maximal tidal volume and compared these results with reference values obtained by Dubowy et al.
Results: We observed no complications of exercise testing. The maximal heart rate of patients was significantly ($p < 0.05$) below normal values (18%). The chronotropic response was significantly lower (68 vs 96 bpm) in the patient group. The maximal oxygen pulse was found significantly decreased (−1.1 ml/beat) as well as the VO_2-AT (−0.2 l/min). The maximal tidal volume during exercise of patients with HLHS was in the lower range of reference values, but not significantly decreased. During rest, 56% of the patients showed a restrictive lung function.
Conclusion: Patients with HLHS have a decreased ability to increase their cardiac output with heart rate and oxygen pulse. They reach their anaerobic threshold earlier than healthy children. Respiratory impairment may contribute to the limited cardiorespiratory

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capacity. However, as the limitations are moderate and no patient showed complications of exercise testing, we feel that participation in recreational sports can be recommended.

O-36
Developmental outcome after three stage palliation for hypoplastic left heart syndrome

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Introduction and Methods: 35 children born between 1996 and 2001 with hypoplastic left heart syndrome were evaluated after their three staged palliation (Norwood-, Hemifontan-, Fontan operation) with the help of standardized tests for their cognitive (K-ABC) and visual motor integration (VMI) developmental stage and were compared with a control group of healthy children who were matched for their age and gender. At the same time we used a parent questionnaire to get information about the quality of life (Kindl test) and behaviour problems (CBCL).

Possible influence factors, for example the duration of the total cardiac arrest time, the pulmonary bypass time were checked of significant correlations with the test data.

Results: The scores of the intelligence quotient from the "scale of intellectual abilities" and "scale of proficiency" (K-ABC) were with 79.0 ± 3.1 respectively 78.3 ± 15.6 significantly ($p < 0.01$) lower as for the control group (105.1 ± 8.3; 105.3 ± 10.4).

The children with hypoplastic left heart syndrome showed in the area of visual motor integration also significantly lower scores (77.5 ± 19.6, $p < 0.01$; control group 106.9 ± 10.6).

Particularly the influence factor cardiac arrest time showed a negative significance for both areas.

Both groups had equal good quality of life and no behaviour problems.

Conclusion: The first successfully operated children with hypoplastic left heart syndrome show after their stage palliation a worse cognitive and visual motor integration developmental stage than the control group.

Fortunately the children with hypoplastic left heart syndrome have also a good quality of life.

Session 7: General Paediatric Cardiology

O-37
N-terminal-pro-B-type natriuretic peptide differentiates sepsis from acute left ventricular dysfunction in critically ill children

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Background: N-terminal-pro-B-type natriuretic peptide (NT-proBNP) has been shown to be a marker for cardiac dysfunction. The peptide is also elevated in patients with sepsis. The purpose of this study was to assess whether NT-proBNP levels can differentiate pediatric patients with sepsis from patients with acute left ventricular dysfunction (LVD).

Methods: Pediatric patients admitted to intensive care units with sepsis or acute LVD were evaluated clinically and the grade of the hemodynamic aspects of systemic inflammatory response syndrome (SIRS) was determined. Echocardiogram was performed, and levels of NT-proBNP were measured. NT-proBNP level was also measured in patients with simple febrile illness.

Results: There were 10 patients with sepsis and 10 with acute LVD. The age of the patients was similar. SIRS grading was not different (sepsis 2.8 ± 0.4, acute LVD 2.6 ± 0.7, $p = 0.49$). NT-proBNP levels were elevated in patients with sepsis (median 6064 pg/ml, range 495–60417 pg/ml), but were significantly higher in patients with acute LVD (median 65630 pg/ml, range 15125–288000, $p = 0.01$). The area under the receiver operating characteristics (ROC) curve for the diagnosis of acute LVD was 0.9 (95% CI 0.77–1.0). NT-proBNP of 20 patients with simple febrile illness was significantly lower (median 280 pg/ml, range 12–1970 pg/ml ($p = 0.002$ vs sepsis).

Conclusions: NT-proBNP levels are elevated in pediatric patients with sepsis, but are significantly higher in patients with acute LVD. Excessive elevation in NT-proBNP levels suggests cardiac etiology for acute hemodynamic deterioration in infants and children.

O-38
Arginine-vasopressin in neonates with vasodilatory shock after cardiopulmonary bypass

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Introduction: Severe systemic vasodilation can occur due to systemic inflammatory response after cardiopulmonary bypass. Successful therapy of vasodilatory shock after cardiopulmonary bypass in adults and children with arginine-vasopressin (AVP) has been reported previously. Data on the use of vasopressin in neonates with vasodilatory shock is limited, indications and dosing have not been established.

Methods and Results: From March 2003 through December 2005 172 neonates underwent open heart surgery, 17 developed severe vasopressor- resistant hypotension and were treated with AVP. The study group included 11 males and 6 females with a median age of 6 days (3 to 12 days). 2 patients had an arterial switch operation, 12 patients a Norwood I- palliation, 2 a Ross-Operation and one patient with single ventricle had an aortic arch reconstruction and pulmonary artery banding. All patients were receiving multiple traditional inotropes and vasopressors prior to administration of AVP. All had adequate cardiac function as assessed by echocardiography. AVP was initiated 0–24 (median 14.5) hours postoperatively. The vasopressin infusion was started at 0.00005–0.001 (median 0.0001) $U\,kg^{-1}\,min^{-1}$ and titrated up to a maximum of 0.0001–0.001 (median 0.0003) $U\,kg^{-1}\,min^{-1}$. AVP led to a significant increase in blood pressure (from baseline 49 ± 8 mmHg to 69 ± 7 mmHg) and a significant increase in urine output (from baseline 0.87 ml/kg/h to 1.68 ml/kg/h). AVP-infusion was discontinued after 48 (range 9–85) hours. There was no change in serum sodium level, no peripheral vasoconstriction and no other side effects observed. 13 of these profoundly ill neonates survived to discharge, 4 patients, 3 with hypoplastic left heart and 1 patient with single ventricle after arch repair and PA-banding died. In 2 patients death occurred due to additional complications 6 days after AVP was discontinued. One patient, who was still on AVP died 42 hours postoperative after prolonged hypoxemia not responding to inhaled NO. 1 patient arrested on postoperative day 3 when AVP was almost weaned.

Conclusion: In critically ill neonates with vasodilatory shock after cardiopulmonary bypass AVP is a potent agent to maintain blood pressure and cardiac output when traditional vasopressors are failing.

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O-39

Off-pump pulmonary valve replacement using the Shelhigh Injectable Stented Pulmonic Valve

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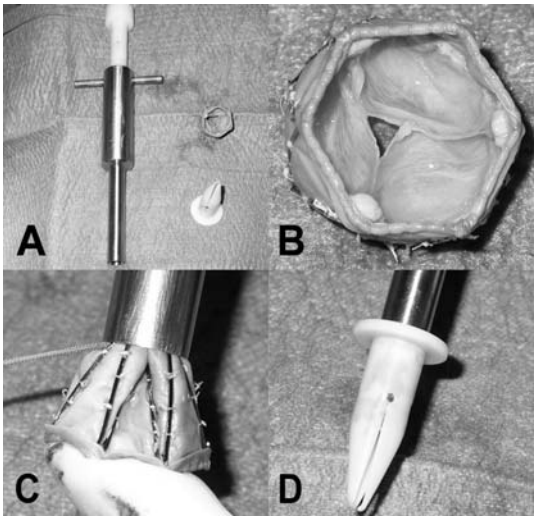
Objectives: Significant pulmonary regurgitation (PR) is common after surgical or percutaneous treatment of pulmonary stenosis and Tetralogy of Fallot (TOF) leading to significant late morbidity and mortality. Optimal timing of pulmonary valve replacement (PVR) and procedures minimizing surgical trauma are therefore important. The new Shelhigh Injectable Stented Pulmonic Valve NR4000-PA MIS (Shelhigh Inc, New Jersey, USA) offers the possibility of PVR without cardiopulmonary bypass (CPB). Here we report our early experience with the first clinical use of this new device.
Methods: 4 symptomatic patients aged (median) 14.8 (11.2–40) years, with severe pulmonary regurgitation and progressive RV dilatation with dysfunction after TOF repair (n = 3) or correction of congenital pulmonary stenosis (n = 1) were treated with the new Shelhigh Injectable Stented Pulmonic Valve.



Results: Valve sizes 23 (n = 2), 25 and 29 were used. Median operation time was 165 (150–280) min. Valve implantation was primarily successful with easy insertion, delivery and placement in all patients. Echocardiographic and invasive hemodynamic assessment showed no regurgitation in all patients, a median peak systolic gradient of 6.25 (4.7–11) mmHg and a median mean systolic gradient of 2.15 (2–4.9) mmHg. Before valve insertion, one patient required additional reduction plasty of the severely enlarged MPA (33 mm). Early recovery was uneventful and all patients discharged home after a median length of stay of 7.5 (7–8) days. One patient required reoperation with replacement by a conventional conduit 2 months later due to valve migration with severe MPA obstruction and recurrent PR. This patient had a dilated and conical shaped MPA (35 mm) not having been reduced at the time of valve insertion. Echocardiographic follow-up after 4, 9, 12 and 18 (median 12.2, range 4.3–18.2) months showed good results in the remainder 3 patients with low gradients in all and moderate paravalvular PR in only one. RV function has recovered all but the one patient with moderate PR to subnormal values.
Conclusions: The new Shelhigh Injectable Stented Pulmonic Valve allows for easy pulmonary valve replacement without cardiopulmonary bypass. Reduction plasty in the presence of a dilated main pulmonary artery or right ventricular outflow tract and external fixation are crucial to ensure proper positioning and excellent durable performance.

O-40

Implantation of a prosthesis mounted inside a self-expandable stent in the pulmonary valvar area without use of cardiopulmonary bypass

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Objectives: Pulmonary regurgitation is the predominant problem in the long-term follow-up of TOF patients after primary repair. Apart from standard homograft implantation, a percutaneous approach has been described recently. A RVOT diameter of greater than 22 mm, however, precludes this approach to date. We describe a novel technique with a transventricular implantation of a stented bio-prosthesis without cardiopulmonary bypass that allows implantation of prosthesis with diameters up to 31 mm.
Methods: All five patients (14, 15, 16, and 27 years of age), presented with severe pulmonary regurgitation (median MRI pulmonary regurgitation was 50%; range 45–64%) and hugely dilated right ventricles (median MRI RVEDD/m^2 was 141 ml/m^2; range 118–174 ml/m^2). The initial repair included a transannular RVOT patch. After repeat sternotomy, a No-React® treated porcine valve mounted inside a self-expandable stent (size 24, 25, 27, and 31 mm, Shelhigh, Model NR-4000 MIS), was injected just beneath the RVOT without use of cardiopulmonary bypass. Externally, sutures were placed at the proximal and distal site of the valve to ensure fixation.
Results: The implantations were uneventful, with the patients hemodynamically stable throughout the procedure. Echocardiographic assessment confirmed the adequate position and function of the valve. In one patient with hugely dilated RVOT (34 mm), a homograft was implanted after 2 days due to paravalvular leakage. After a mean follow-up of 4.4 months (30 days – 9 months) right ventricular function improved with satisfactory valvar function.
Conclusions: This newly available device in combination with the wide range of prosthesis sizes offers yet another treatment option. Cardiopulmonary bypass can be avoided in selected patients. In the future, technical refinements may allow either changed modes of valve delivery, different securing methods of the stented valve, or even also a closed chest approach.

O-41

Quality of life after corrective surgery for congenital heart disease

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After corrective surgery for congenital heart defects (CHD) many patients suffer from residual defects, some with reduced cardiorespiratory capacity and possible impairment of their quality of life (QoL). The aim of our study is to evaluate, how children after

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surgery for CHD rate their QoL. A standardised questionnaire developed for self-rating in children between 8 and 18 years and dealing with seven different domains concerning the QoL (friends, family, physical functioning, cognition, body image, emotional function and autonomy) was independently answered from patients and their parents throughout Germany during an outpatient visit at their pediatric cardiologist.

173 patients (40% female, 60% male, mean age 11.6 years) were interviewed; the mean time interval after surgery was 9.8 ± 3.4 years. Patient were subdivided according to the type of CHD (VSD n = 52, TOF n = 52, univentricular heart n = 27, TGA n = 42). The results were compared with those of an age-matched control group (n = 551).

Patients with CHD reported a better QoL than the controls for all items ($p < 0.01$). Especially the body image was rated as superior by pubertal and postpubertal patients. Patients older than 15 years were rated less positive in the domains family and emotion compared to younger patients. Comparing the four categories of CHDs there was no difference except for the physical function, which was reported to be reduced for patients with TGA. With regard to the mode of surgery only patients after atrial switch operation rated their physical function and cognition reduced compared to other patients. The impact of the number of interventions and the time interval between the last intervention and the interview influenced only the domain "social function with friends" negatively. Parents assessed their children significantly worse in three domains (social function, body image and emotion; $p < 0.01$ vs. patients).

We conclude that children after surgery for CHD surprisingly view their QoL very positively and significantly better than healthy controls for all domains studied. There are few differences between the various groups of CHD. Parents rate their children's QoL worse in three domains. Further investigation should demonstrate the relationship between QoL assessment and objective cardiopulmonary function.

O-42
Long-term neurodevelopmental outcome after open heart surgery

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Background: Constantly decreasing mortality rates after open heart surgery enhances the importance of assessing long-term neurodevelopmental outcome in children with congenital heart disease (CHD). The range and the severity of impairments as well as the predicting factors still need to be determined.
Objectives: To assess intellectual, motor and neurological outcome in school-aged children who have undergone open-heart surgery and to identify peri- and intraoperative risk factors for adverse outcome.
Methods: Between 1995 and 1998, 155 children with CHD (40% cyanotic) underwent open-heart surgery in our institution. Neurodevelopmental assessment was performed in 120 (79%) at a mean age of 10.4 years. Children with identified chromosomal anomalies were excluded. Intellectual outcome was assessed with the Raven Standard Progressive Matrices, motor performance with the Zurich Neuromotor Assessment and a standardized neurological examination was performed.
Results: Mean age at operation was 2.3 years (range 0–8.7 years), mean ECC time was 95 min (5–206) and circulatory arrest was performed in 9% of the children. Mean IQ was significantly below the norm (89 (SD 16.4); $p < 0.001$); Mild mental delay (IQ 70–84) was diagnosed in 35% and severe mental delay (IQ < 70) in 8.4% (both $p < 0.001$), with 13% of all children requiring special schooling. Cerebral palsy (CP) was diagnosed in 12% (normal population prevalence 0.2%). Motor impairment in non-CP children was observed predominantly in the pure motor and static balance task (z score −0.30, $p = 0.01$; −0.56, $p < 0.001$, respectively). Socioeconomic status was the only significant risk factor for later IQ impairment. Postoperative EEG changes or focal neurological abnormalities were predictive of CP ($p = 0.001$); no other perioperative variable was independently associated with CP. Motor performance was related to the number of open-heart surgeries ($p = 0.01$).
Conclusion: Children undergoing congenital-heart surgery are at risk for neurodevelopmental impairments at school age. Socioeconomic status is a significant determinant of later intellectual performance, whereas perioperative factors seem to play a minor role in predicting adverse neurocognitive outcome. Careful serial neurodevelopmental evaluation of these patients and early initiation of therapeutic interventions is recommended for these children.

Session 8: General Paediatric Cardiology

O-43
Myocardial velocities measured with MRI: comparison between patients with corrected tetralogy of Fallot (TOF) and patients after atrial switch of d-transposition of the great ateries (d-TGA)

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Introduction: MRI allows quantification of myocardial velocities and gives a refined statement about cardiac function. Aim of this investigation was to find out differences between myocardial velocities in different congenital heart diseases and how those differ from normal velocities found in healthy subjects.
Methods: We examined 48 patients (6–61 years) three to 24 years after corrective surgery of TOF and 27 patients with d-TGA between (9–26 years) 18 ± 4 years after atrial switch procedure. Furthermore, we examined 13 normal control subjects.

Examination was performed on a 1.5 T MRI scanner. Results were compared between the two patient groups and with those of the control subjects. Myocardial velocities were measured in a four chamber view for each of the subjects using 3D-phase contrast imaging in three directions (longitudinal, radial, circumferential). Three intramyocardial ROIs (LV, RV, interventricular septum = IVS) in the myocardium were defined for each data set and myocardial velocity-time curves were generated.
Results: In longitudinal and radial direction we found velocity-time curves, which showed a systolic (s-wave), an early (e-wave) and late diastolic (a-wave) motion for each intramyocardial ROI.

In longitudinal direction the RV showed lower mean velocities in d-TGA- than in TOF-patients: systole (5.6 vs. 6.7 cm/s, $p = 0.02$), in early (5.5 vs. 6.6 cm/s, $p = 0.01$) and late diastole (3.3 vs. 5.4 cm/s, $p < 0.01$).

IVS showed an unphysiologic motion in radial direction in half of the d-TGA-patients and in one third of the TOF-patients. This paradoxical septal motion leads to a significantly lower LV-EF of those TOF-patients as compared to TOF-patients with normal septal motion (mean LV-EF = 51% vs. 57%, $p = 0.03$).

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In circumferential direction lower septal and RV-velocities were measured in d-TGA-patients than in TOF-patients ($p \leq 0.03$).
Conclusions: Like in echocardiographic tissue Doppler, MRI can describe regional systolic and diastolic myocardial function in more detail as compared to the simple calculation of global ejection fraction. In contrast to tissue Doppler, velocities in all three directions can be measured. This approach might provide a better diagnostic parameter than EF alone in patients with congenital heart disease and showed significant differences in the different diseases.

O-44
Reduced left ventricular deformation in children with hypertrophic cardiomyopathy correlates with wall thickness and is associated with reduced exercise capacity

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Background: Clinical presentation and consequences of hypertrophic cardiomyopathy (HCM) are highly variable. Recently, it has been shown that myocardial velocities are good predictors of exercise capacity and arrhythmia in children with HCM. As HCM is a disease with marked regional differences in myocardial function, the use of parameters that reflect regional myocardial function may be more appropriate. Strain (S) and Strain Rate (SR) imaging are tissue Doppler modalities that allow quantitative non-invasive assessment of regional myocardial function.
Aim: To determine the relationship between SR and S with exercise capacity and wall thickness in children with HCM.
Methods: We examined 19 children with asymmetric HCM, (mean age 14.6 ± 2.7 yrs). Standard echocardiograms and tissue Doppler imaging were performed. End diastolic wall thickness was measured at the basal septum. Regional LV deformation parameters of peak systolic SR and end-systolic S were calculated at: (1) Basal septum at largest wall thickness; (2) Mid-apical septum without significant hypertrophy. Exercise capacity was evaluated by measuring percentage of predicted peak oxygen uptake (VO_2).
Results: In five patients a peak gradient >20 mmHg at the LV outflow tract was seen. Basal septal end diastolic wall thickness was 5.1 ± 2.6 Z scores. Fractional shortening was: 39.0 ± 10.2%. Peak systolic SR and S were significantly lower at the basal septum compared to mid septum, see Table. Moreover, systolic lengthening was seen in seven patients. Peak systolic SR and S at the basal septum significantly correlated with exercise capacity (r: 0.74, $p < 0.01$; and r: 0.68, $p < 0.01$ respectively). Peak systolic SR and S inversely correlated with wall thickness (r: 0.82, $p < 0.001$; r: 0.85, $p < 0.82$, $p < 0.01$ respectively).
Conclusions: In children with HCM the development of hypertrophy is associated with reduced deformation. These decreased LV deformation correlates significantly with impaired maximal aerobic exercise capacity. Strain rate imaging provides the clinician with sensitive quantitative measures of LV function that may be serially used for follow-up to facilitate strategies designed to preserve long term ventricular function and exercise performance.

Table.

	HCM basal septum	HCM mid septum	P value
Strain rate (1/s)	−0.75 ± 0.51	−1.27 ± 0.81	0.02
Strain (%)	−8.46 ± 9.29	−16.90 ± 12.28	0.02

O-45
A new simple method to quantify pulmonary regurgitation by transthoracic echocardiography: comparison with magnetic resonance imaging

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Introduction and Aim: Pulmonary regurgitation (PR) is a common sequela and an important cause of morbidity after surgical repair of Tetralogy of Fallot or other right ventricle outflow tract anomalies. PR quantification by Trans-Thoracic Echocardiography (TTE) has major limitations, whereas Cardiovascular Magnetic Resonance Imaging (CV-MRI) by phase velocity contrast acquisition is the established gold standard. the aim is to validate a novel method to quantify PR by TTE based on systo-diastolic right pulmonary artery excursion
Methods: 43 patients with repaired right ventricle outflow tract, without significant pulmonary artery branches stenosis or aneurysm, or previous pulmonary artery stent implantation, were randomly submitted to TTE and CV-MRI. In one, the TTE study could not be completed. In the remaining 42 patients (aged 18 ± 9 years) the ratio between systolic/diastolic right pulmonary artery diameter (RPA excursion index) was evaluated by TTE and correlated with PR fraction blindly assessed by phase velocity mapping CV MRI.
Results: By TTE, the overall feasibility of RPA excursion index was 97.5%, with an extra-imaging and analysis time <2 min per patient. PR fraction by CV-MRI ranged from absent (0%, n = 4) to severe (>45%, n = 10). RPA index ranged from 1.03 (no excursion) to 1.96 (marked excursion). The RPA excursion index closely correlated with the PR fraction ($r = 0.80$, $P < 0.001$) (fig. 1).
Conclusions: the RPA excursion index is easily calculated by TTE and could be a reliable method to quantify PR.

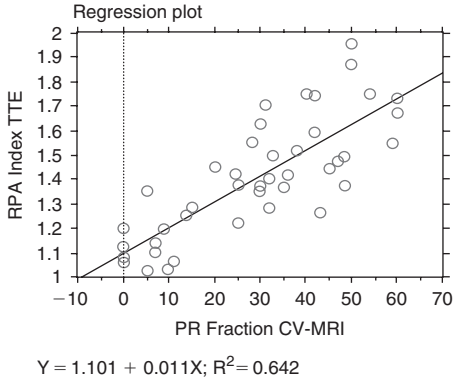

Figure 1. Correlation RPA excursion index (TTE)/PR Fraction (CV-MRI).

O-46
Oral everolimus inhibits degeneration in prosthetic pulmonary valves in pigs

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Introduction: Growth factor induced cell proliferation can lead to prosthetic heart valve stenosis and the degeneration of valve

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substitutes. We studied whether the administration of oral everolimus (Certican, Novartis) inhibits degenerative tissue neogenesis in association with prosthetic pulmonary valves.
Materials and Methods: In 12 pigs (35 kg) prosthetic pulmonary valves were implanted using a transcatheter technique. Tricuspid valves were prepared from a 0.1 mm thick titan-coated polymer and sewn into a self-explanding nitinol stent (diameter = 20 mm). These valve stents were placed in pulmonary position, where they remained for 3 months. In six animals, treatment with 2 mg everolimus per day was started 3 days before implantation and continued throughout the course of the experiment. The other six animals acted as controls. Adjuvant anticoagulation treatment consisted of ASS p.o. und Plavix i.m. After 3 months the hemodynamics of the valves was investigated using MRT. Postmortem the valve stents were explanted and subjected to macroscopic and microscopic examination.
Results: There were no undesired side effects under everolimus treatment; the everolimus level measured was 4 ± 2 ng/ml. In the everolimus group MRT showed laminar flow through the pulmonary valve and a regurgitation fraction of $9 \pm 5\%$; in the control group flow was turbulent with aliasing (evidence of stenosis-induced flow acceleration) and the regurgitation fraction was $35 \pm 11\%$. In macroscopic terms all control animals showed massive valve degeneration caused by tissue ingrowth, whereas in the everolimus group degeneration was absent or only slight. Microscopic evaluation in both groups showed neointima, smooth muscle cells and proteoglycan-collagen matrix. However, the thickness of these layers in the everolimus group was significantly greater ($p < 0.001$).
Conclusion: Oral administration of everolimus greatly inhibits tissue neogenesis and therefore valve degeneration in pulmonary heart valve stents in pigs.

O-47
Closure of atrial septal defects without fluoroscopy – a five year experience

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Introduction: Five years ago, Ewert et al. described transcatheter closure of atrial septal defects without fluoroscopy (Circulation. 2000;101:847–849). We present our experience with this technique.
Methods: Prospective observational study 04/00–12/05. Inclusion criteria: attempted occlusion of an interatrial communication. Exclusion criteria: planned fluoroscopy due to associated lesions, additional interventions or body weight below 10 kg. End points: need for fluoroscopy at any time during the procedure, feasibility of defect occlusion. 147 patients were enrolled: age median 7.9 (range 1–41) years, weight 24.4 (10–123) kg, length 123 (72–189) cm, BSA 0.9 (0.4–2.5) m^2. Right heart catheterization (pressure recording and oxymetry) was performed without visual guidance. Qp:Qs was 2.0 (1.4–5.0). Crossing of the defect, balloon sizing and Amplatzer device implantation were performed with transesophageal echocardiographic guidance in sedated (midazolam plus propofol) and spontaneously breathing patients. Defect size was native 14 (5–30) mm and balloon occluded 18 (7–32) mm, occluder size 18 (8–34) mm. Procedure time was 60 (30–242) min. Statistical analysis was performed to identify differences between patient groups (without fluoroscopy versus with fluoroscopy).
Results: Diagnostic catheterization and balloon sizing were feasible without fluoroscopy in all 147 patients enrolled. Device occlusion was completed without fluoroscopy in 115 (78%). Fluoroscopy was required in 32 (22%) patients due to difficult device configuration or dislocation and allowed for successful implantation in 20 more patients (14%). The overall occlusion rate was 135/147 (92%).

There were no differences in demographic data between the groups. Qp:Qs was 2.0 in the fluoroscopy groups versus 1.85 in the group without fluoroscopy (not significant). According to the protocol procedure time was longer (84 versus 53 min) in patients with fluoroscopy. The nominal occluder size was larger in the fluoroscopy group (22 versus 17 mm), which was also true when indexed for body surface area (23.3 versus 17.0 mm/m^2). Within the fluoroscopy group there was no difference in occluder size between success and failure.
Conclusion: Transcatheter closure of atrial septal defects is feasible without fluoroscopy in the majority of patients.Therefore it remains the standard approach in isolated atrial septal defects in our institution, thus reducing radiation exposure of patients and personnel.

O-48
Interventional cardiac catheterization in pediatric orthotopic heart transplant patients: transplant surgery is not the end of the road

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Introduction: Pediatric orthotopic cardiac transplantation (OHT) has been performed in 276 patients at The Children's Hospital in Denver, Colorado since 1988. Patients with complex anatomy and those who have undergone multiple palliative surgeries prior to OHT frequently have vascular access and great vessel abnormalities requiring intervention. There is a paucity of data on interventional procedures required in children and young adults following OHT.
Methods: A retrospective review of our pediatric cardiology catheterization and OHT databases was undertaken in order to quantify and characterize interventional procedures performed in children and young adults following OHT.
Results: From 1988–2005, 1475 catheterizations were performed following OHT. Of these, 58 interventions, excluding myocardial biopsies, were performed in 46 patients age 1.6 [0.1–26] years with a weight of 10.2 [3.5–96.0] kg. Sixteen patients had coarctation of the aorta and underwent 18 balloon angioplasties. No patient required stent placement. Iliofemoral vein obstruction was treated with isolated balloon angioplasty in 3 patients. Iliofemoral vein stent placement was undertaken in 10 patients on 12 occasions to restore femoral to inferior vena caval continuity. One to six stents per patient were required. Following final stent placement, femoral vein patency was confirmed at >1 year post procedure in 6 (60%) patients with the remaining 4 patients not yet having a follow up procedure. In 1 patient, recanalization was not feasible. Superior vena cava (SVC) obstruction was addressed with angioplasty and/or stent placement in 17 of 18 patients. The remaining patient had complete SVC obstruction making intervention not feasible. Other procedures included angioplasty and/or stenting of left pulmonary artery (LPA) stenosis in 4, atrial septal defect closure in 1, LPA to left SVC fistula occlusion in 1 and coronary artery stenting in 1. No major adverse events occurred.
Conclusions: Obstructive vascular lesions are common following pediatric OHT occurring in 17% of patients. Venous obstruction may be a particularly significant obstacle to ongoing care because these patients require life long surveillance. Femoral venous and great vessel obstructive lesions should therefore be aggressively sought as they can be successfully addressed using interventional techniques.

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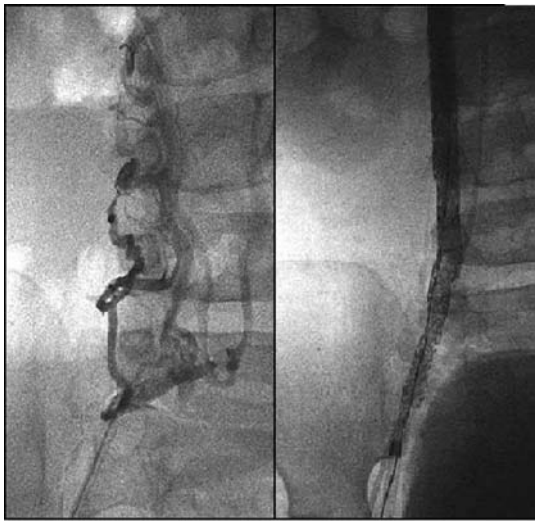

Pre and post angiograms demonstrating femoral to IVC recanalization

Session 9: Surgery

O-49

Plasma brain natriuretic peptide levels as a marker of right ventricular dilatation and recovery after pulmonary valve insertion in corrected tetralogy of fallot: correlations with cardiac magnetic resonance imagery

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Introduction: Cardiac volumetry by magnetic resonance imaging (MRI) can guide the timing for reoperation in asymptomatic patients with severe pulmonary insufficiency (PI) after corrected Tetralogy of Fallot (TOF). Plasma proBrain Natriuretic Peptide (BNP) is a marker of ventricular strain and dilatation, and levels may complement MRI volumetry to assess cardiac dilatation and remodelling in patients with severe PI, before and after pulmonary valve replacement (PVR).
Methods: Between 5.2004 and 10.2005, 23 consecutive asymptomatic patients with corrected TOF, severe PI and right ventricular end-diastolic volume index (RVEDVI) $>100\,ml/m^2$ underwent elective pulmonary valve replacement. Plasma proBNP levels and cardiac MRI were obtained one day before and 6 months after PVR, and analysed for correlations.
Results: There was no surgical mortality or morbidity. Pre-operative RVEDVI correlated with PI ($r = 0.52$), and inversely so with left ventricular ejection fraction (EF), reflecting inter-ventricular interaction ($r = -0.40$). Pre and 6 months post-operatively, logBNP was inversely correlated with RVEF. Mean pre-operative proBNP levels, RVEDVI, and PI diminished significantly 6 months after PVR (231 versus 114 ng/L; $p < 0.0001$, 184 versus $109\,ml/m^2$; $p < 0.0001$, and 44 versus 2%; $p < 0.0001$, respectively). However, correlation was weak between plasma BNP and RVEDVI before ($r = 0.20$) and 6 months after ($r = 0.28$) surgery.
Conclusions: ProBNP is elevated in patients with corrected TOF, severe PI and RV dilatation. Plasma levels drop and nearly normalize 6 months after insertion of a competent pulmonary valve, mirroring smaller RVEDVI, as documented by cardiac MRI. ProBNP promises as a marker of RV dilatation and recovery before and after surgery on the RV outflow, and may help in the operative timing of asymptomatic patients with chronic PI and slowly dilating right ventricles.

O-50

Bovine jugular valved conduit performs better in a dilated than in an obstructed pulmonary arterial tree

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Introduction or Basis or Objectives: Reoperations for degeneration of a valved conduit remain a major concern. Knowing that all the available biological conduits need to be replaced once, the strategy which allows longest survival needs to be pursued. We critically analyzed our cohort of consecutive 142 bovine jugular vein conduit (BJV) survivors to determine what led to best survival.
Methods: All survivors after BJV insertion for RVOT (2001–2005) were reviewed. There were 134 pts with 142 implantations: 87 TOF, 28 Ross Procedures, 12 Truncus arteriosus, 7 d-TGA VSD-PS and 8 miscellaneous. Three groups were identified based on the status of the pulmonary arterial tree: an obstructive (with RV pressure overload) (54 pts), a dilatative (with RV volume overload) (60 pts) and a normal (without pressure or volume overload as in Ross procedure) (28 pts). Follow-up (median 2 years) was 100% complete.
Results: Eleven conduits needed explantation and 4 continued to be palliated with balloon dilatations, due to an anastomotic stenosis. In none, the haemodynamic problem was primarily a regurgitation of the BJV. Reintervention was necessary in 10 pts in the obstructive, 3 in the normal and 2 in the dilatative group. Kaplan Meier freedom from reintervention at 42 months was 77% (+13, −13), 90% (+10.3, −11.1) and 94% (+6.3, −9.2) in the respective groups (See Figure 1). The difference between obstructive and dilatative groups was significant ($p = 0.02$).
Conclusions: BJV fare better in dilatative lesions than in obstructive or in normally loaded ones. These data suggest that during the primary repair of TOF/Pulmonary atresia and TGA VSD-PS, the implantation of BJV should be avoided as far as possible, in deference to reconstructive procedures. Residual regurgitation and the resultant pendulum flow after such reconstructions, promotes dilatation of the pulmonary arborization, thus effectively improving the longitivity of the conduit implanted secondarily.

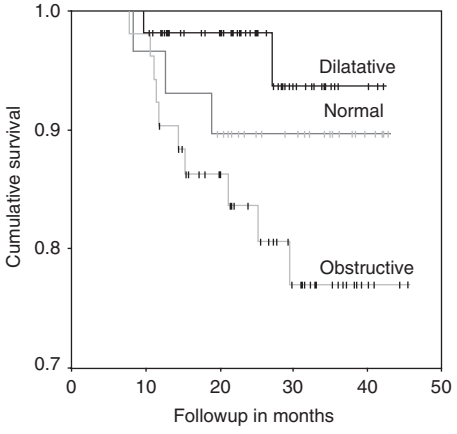

Figure 1. Kaplan Meier survival curves for obstructive, normal and dilatative pulmonary arterial systems.

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O-51
Outcomes of small right ventricle to pulmonary artery conduits

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Introduction: The right ventricle to pulmonary artery (RVPA) conduit has been used for palliation or cure since the 1960s. Studies have been done on conduit types and the larger sized conduits but few have looked at the smaller sized conduits.
Methods: All 163 RVPA conduits with an internal diameter ranging from 6 to 13 mm that were implanted from 1988 through June 2004 were retrospectively analyzed. These included the Homografts (Aortic, Pulmonary), Hancock (dacron conduit with porcine valve), Bovine (Contegra, Venpro), Porcine (Shelhigh, Tissuemed) and Gortex conduits. All cases were normalized to the patient's body surface area: z-score. The Kaplan-Meier method was used to estimate survival of conduit from change or death as well as from cardiological intervention (angioplasty and/or stenting). The Log-Rank test was used to assess any differences in survival.
Results: One hundred and fifty four patients were analysed with nine lost to follow to up. Seventy eight were male. Diagnostic categories included Pulmonary Atresia and its variants, Truncus arteriosus, Aortic Atresia and interrupted arch, Transposition of the Great Arteries, Absent Pulmonary Valve Syndrome and others. There were 42 deaths 22 were early (less than 30 days). Overall survival of all conduits was 94.4 ± 2.0% at 6 months, 87.1 ± 3.1% at 1 year, 40.5 ± 6.3% at five years and 21.71 ± 8.8% at ten years. Freedom from cardiac catheterisation interventions at 6 months was 87.0 ± 3%, 71.6 ± 4.0% at 1 year, 44.3 ± 5.6% at five years and 20.49 ± 9.81% at ten years. Factors which were analysed included body surface area (z score), conduit size, conduit type, diagnosis and gender.

Conduit type (p value 0.0396) was the only statistically significant factor which influenced outcome.

The z-score for the body surface area were banded into three categories larger z scores did survive longer but were not significant.
Conclusion: Contrary to the assumption that the size of conduit appears to lengthen the survival time of the RVPA conduit this study shows otherwise. What is significant is the type of conduit used. In this study Homografts are superior to all others both for freedom of surgical and catheter intervention.

O-52
Echocardiographic small residual ventricular septal defects after surgical repair – back on bypass or wait for spontaneous closure?

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Introduction: Small residual defects may be detected by intra or post-operative echocardiography after surgical closure of a ventricular septal defect (VSD). The hemodynamic relevance and rate of late closure of these defects are unknown.
Methods: Between 1994 and 2005, 239 consecutive patients underwent surgical closure of an isolated perimembranous VSD (n = 120), a malalignement Tetralogy of Fallot VSD (n = 66), or an inlet VSD in atrio-ventricular septal defect (AVSD) (n = 53). Post-operative transthoracic echocardiography (TTE) was performed on the intensive care unit (ICU), at hospital discharge, and during regular follow-up. Residual defects were graded as absent, between 1–2 mm, or greater than 2 mm.
Results: 82% of all residual defects <2 mm closed. Of 15 residual defects >2 mm on the ICU, only 6 closed after a median follow-up of 3.4 years. In these patients, they were hemodynamically insignificant, required no medication, and no endocarditis was noted. At last follow-up, residual VSDs were significantly more frequent after Fallot repair and isolated VSD closure, compared to that after AVSD correction (p = 0.045). Minor discrepancy was observed between ICU and discharge echocardiographic findings.

	ICU	Hospital discharge	Ambulatory (median 7.4 months; range 0.4–97.4)	Last follow-up (median 40.4 months; range 5.8–144.5)
AVSD (n = 53)				
No VSD	35	32	52	52
<2 mm	16	19	1	1
>2 mm	2	2	0	0
	34%	40%	2%	2%
Fallot (n = 66)				
No VSD	33	40	55	56
<2 mm	26	20	6	5
>2 mm	7	6	5	5
	50%	39%	17%	15%
Isolated VSD (n = 120)				
No VSD	83	77	103	109
<2 mm	31	37	12	7
>2 mm	6	6	5	4
	41%	51%	16%	10%

Conclusions: Post-surgical residual VSDs <2 mm closed spontaneously in the vast majority of cases within a year. Defects >2 mm are unlikely to close spontaneously. Residual defects after AVSD repair almost always close, whereas two thirds will remain open after Fallot or isolated VSD repair. Although defects >2 mm may remain hemodynamically and clinically irrelevant, revision of a perimembranous VSD on cardiopulmonary bypass at initial repair, guided by transesophageal echocardiography, may spare lifelong endocarditis prophylaxis.

O-53
Surgical repair of congenital mitral valve anomalies in pediatric age group: early and mid term results.

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Objective: To evaluate the effectiveness of mitral valve (MV) repair in patients with congenital mitral valve dysplasia, with or without associated cardiac malformations.
Methods: Between January 1987 and December 2005, 60 patients underwent MV surgical repair (M/F = 28/32, median 3.9 years, range 1.8 month–19.8 years). Patient with anatomic diagnosis of AV septal defect and univentricular heart were excluded from this analysis. We divided this population in: Group A, where MV anomaly was the main cardiac defect (26 patients), Group B where MV anomaly was associated to other intracardiac lesions (34 patients). MV prevalent dysfunction was: regurgitation in 35 (58.3%, 18/26 in Group A, 17/34 in Group B), stenosis in 25 (41.7%, 8/26 in Group A,

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17/34 in Group B). Seventeen patients (28%) had undergone previous operation for other cardiac or vascular anomalies. In Group B, subaortic stenosis was the most frequent associated lesion (60%), followed by ventricular septal defect (40%).

Surgical repair was tailored to patient's valve anatomy. Outcome for mortality, late reoperations and clinical follow up were analysed.
Results: MV reconstruction was possible in all the patients . There were 2 in hospital deaths (3%), all in Group B (2/34), related to poor preoperative cardiac dysfunction. All survivors were discharged home in good clinical conditions. Follow up completeness is 85%. At a mean follow up of 8.1 years (median 7.7 years, range 1 months–18.6 years) there were 2 late deaths, for prosthetic valve thrombosis, (1, Group A), and for sudden death (1, Group B). At 2-D echocardiographic evaluation, residual MV regurgitation was less than moderate in 78%, while only residual MV stenosis was detected in 10% . In the entire sample, in Groups A and B, freedom from MV replacement was 76, 88 and 73%, overall survival was 92, 95 and 90% respectively, at 18.6 years.
Conclusions: Surgical repair for congenital MV dysplasia is an effective and safe procedure, which has a favourable functional outcome in the medium term. It avoids the negative drawbacks of anticoagulation and most likely differs reoperation for MV replacement in older age, even in the severe forms.

O-54

Independent factors associated with mortality and reoperation in 377 children with total anomalous pulmonary venous drainage

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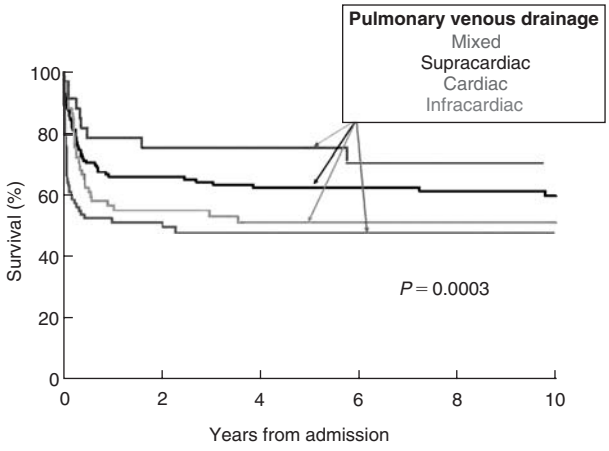
Introduction: We sought to describe morphologic characteristics, particularly pulmonary venous anatomy, and to determine the prevalence of mortality and reoperation in children with total anomalous pulmonary venous drainage (TAPVD).
Methods: From 1946–2005, 377 children with TAPVD without associated complex intracardiac lesions underwent 357 operations at our institution. Medical records were reviewed. Pulmonary venous anatomic drainage was characterized by review of angiography (54%), echocardiography (34%), operative (8%), or autopsy (4%) reports. Multivariable Cox proportional hazard models were used to determine the prevalence and associated risk-factors for the time-related events of death and reoperation.
Results: Median age at presentation was 1 day (range: birth–12 years), and mean weight was 3.9 ± 1.9 kilograms. Pulmonary venous anatomic type was classified as supracardiac in 44%, infracardiac in 26%, cardiac in 21%, and mixed in 9%. Pulmonary venous obstruction was present in 48% at presentation, and was most frequent in those with infracardiac type ($P < 0.001$). Overall survival from admission was 60% at 10 years and was dependent on anatomic type ($P < 0.001$, Figure). No intervention was performed in 50 patients, all of whom died. Complete repair of TAPVD was undertaken in 87% at a median age of 1.6 months (range: 1.8–14 years). Overall survival from repair was 64% at 15 years. Results improved over time ($P < 0.001$). Incremental risk-factors for death after admission included the presence of pulmonary venous obstruction at presentation (Hazard ratio (HR): 2.4; $P = 0.001$), infracardiac (HR: 2.8; $P = 0.005$) or cardiac anatomic TAPVD type (HR: 2.1; $P = 0.048$). Reoperations were performed in 30 children after complete repair, most commonly (53%) for pulmonary venous obstruction. Overall freedom from reoperation was 82% at 15 years following repair. Factors associated with an increased risk of reoperation included earlier birth cohort (HR: 1.1/year; $P = 0.03$), and the presence of pulmonary venous obstruction at presentation (HR 26.4; $P < 0.001$).
Conclusions: Outcomes in children with TAPVD have improved over time. Survival is lowest in those with infracardiac or cardiac TAPVD type. Pulmonary venous obstruction remains the most important risk-factor for both death and reoperation.



Session 10: Imaging

O-55

Cardio-pulmonary interaction in premature newborn with nasal continuous positive airways pressure (n-CPAP) respiratory assistance evaluated with echocardiography

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Introduction: Nasal continuous positive airways pressure (n-CPAP) is an effective treatment in premature infants with respiratory distress. The cardio-pulmonary interactions secondary to n-CPAP are well studied in adults, but less well described in premature infants. We postulated that there could be important interactions with regard to the patent ductus arteriosus (PDA).
Methods: Prospective study, approved by the local ethic committee. Premature infants less than 32 weeks gestation, <7 days-old, needing n-CPAP for respiratory distress, but without the need of additional oxygen were included in the study. Every patient had a first echocardiography with n-CPAP and then n-CPAP was retrieved. 3 hours later the echocardiography was repeated by the same investigator and then the patient replaced on n-CPAP.
Results: 14 premature newborn were included, mean gestational age of 28 ± 2 weeks, mean weight 1.1 ± 0.3 Kg and height 39 ± 3 cm. Echocardiographic measurements are depicted in Table 1.

Significant finding were observed between measurement on n-CPAP or without n-CPAP: on end diastolic left ventricular diameter (12.8 ± 1.6 mm vs. 13.5 ± 2 mm), on end systolic left ventricular diameter (8.4 ± 1.3 mm vs. 9.1 ± 1.5 mm), left atrium diameter (8.9 ± 2.2 mm vs. 10.4 ± 2.5 mm), maximal velocity on tricuspid valve (46 ± 10 cm/s vs. 51 ± 9 cm/s), calculated Qp (3.7 ± 0.8 L/min/m^2 vs. 4.3 ± 0.8 L/min/m^2). Only three patients have demonstrated a PDA during the study.
Conclusion: Positive end expiratory pressure (Peep) has hemodynamic effects which are: reduction of systemic and pulmonary venous return as shown by the changes on tricuspid valve inflow, on the calculated Qp and finally on the diameter of the left atrium and

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left ventricle. We found in premature infants the same hemodynamic effects than those described in adults but with lower Peep values. This could be due to the particular elasticity and weakness of the thoracic wall of premature infants. Interestingly the flow through a PDA seems also to be diminished with Peep, but the number of patients is insufficient to conclude. Further investigation will be needed to better understand these interactions.

Table 1. Echocardiographic measurement (mean (SD)).

	With n-CPAP	Without n-CPAP	*p* value
RV ED diameter (mm)	6.3 (1.7)	6.04 (1.1)	NS
LV ED diameter (mm)	12.8 (1.6)	13.5 (2.0)	<0.05
LV ES diameter (mm)	8.4 (1.3)	9.1 (1.5)	<0.05
SF (%)	34 (5)	33 (6)	NS
Ao valve diameter (mm)	7.4 (1.3)	7.4 (1.2)	NS
LA diameter (mm)	8.9 (2.2)	10.4 (2.5)	<0.05
Vmax Ao (cm/s)	70 (16)	71 (18)	NS
Vmax PV (cm/s)	69 (15)	72 (16)	NS
Vmax TV (cm/s)	46 (10)	51 (9)	<0.05
Vmax MV (cm/s)	53 (17)	54 (18)	NS
Qp (L/min/m^2)	3.7 (0.8)	4.3 (0.8)	<0.05
Qs (L/min/m^2)	4.0 (0.8)	4.0 (0.7)	NS
Qp/Qs	0.92 (0.14)	1.09 (0.23)	<0.05

RV: right ventricle, LV: left ventricle, ED: end diastolic, ES: end systolic, SF: shortening fraction, Ao: aortic valve, LA: left atrium, Vmax: maximum Doppler Velocity, Qp: pulmonary output, Qs: systemic output, NS: non significant.

O-56
Flow pattern in the inferior caval vein reflects fontan hemodynamics: quantification by magnetic resonance imaging

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Introduction: Suboptimal Fontan hemodynamics is a multifactorial problem and is dependent on breathing, pulmonary artery resistance und ventricular function. We investigated whether insufficient or slow forward flow in the inferior caval vein (ICV) reflects the hemodynamics with regard to chronic ascites.
Methods: Flow measurements using phase contrast MRI were performed in 41 of 105 patients who underwent different modifications of Fontan operation between 1992 and 2005. Median age at MRI was 11.5 (4–45) years. Median follow up was 5.7 years up to maximum of 13.1 years. The maximal, minimal and mean flow was determined and correlated with clinical symptoms, particularly with chronic ascites. Twenty-eight of these patients underwent heart catheterization at median 3.5 (1.0–9.5) years after surgery. Correlation of the mean pulmonary artery pressure (PAP) and end diastolic ventricular pressure (EDP) with flow velocity in IVC was also analysed.
Results: MRI measurements showed maximal forward flow of 51 ml/s, minimal flow of 14.8 ml/s and mean flow von 33 ml/s (all median values). Patients with chronic or recurrent ascites (n = 7) had the slowest maximal flow pattern in the IVC with a median of 33 ml/s vs. 65 ml/s in those without ascites (p = 0.002). Similar statistical relevance was observed in comparing the mean flow patterns for the correlation with ascites (34 ml/s vs. 17.6 ml/s, p = 0.004). Patients with a significant backward flow who had an overall adequate mean forward flow (median 27 ml/s) had clinically no ascites. Patients, who had a slow flow in the IVC had a tendency towards elevated PAP (>15 mmHg) or/and EDP (>8 mmHg).
Conclusions: Fontan hemodynamics can be analysed noninvasively by MRI. Patients with chronic ascites clearly show a reduced flow volume in the inferior caval vein in comparison to those with optimal Fontan hemodynamics. Absolute quantification of the flow pattern with MRI technique is helpful to optimize follow-up, therapy and decision making for re-interventions after Fontan operation.

O-57
Quantification mitral regurgitation by real time three-dimensional colour doppler echocardiography

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Aim: A pilot study to evaluate the use of real time three-dimensional colour doppler echocardiography (RT3CDE) to quantify the degree of mitral valve regurgitation.
Background: Accurate assessment of the severity of mitral regurgitation [MR] has been challenging using qualitative or quantitative two-dimensional transthoracic (2D TTE) colour Doppler techniques particularly in children. RT3CDE has been validated against cardiac catheter studies in adults with good correlation. The variation in body surface area and mitral valve annulus in children requires independent dataset to assess MR using RT3CDE.
Method: Prospective assessment of mitral regurgitation by RT3DE using a commercial real-time 3-dimensional imaging system Philips SONOS 7500 (Philips Co, Netherlands) with a 2–4 MHz matrix array transducer. The images were acquired and analysed off line by Q lab echo workstation version 4.1. The valve annulus area [VAA], Vena contracta and regurgitant orifice area [ROA] on colour doppler were measured.
Results: Ten consecutive children referred for RT3DE between November 2004 and January 2005 were included in the study. Age at study ranged from 2 months to 10 years. Six were symptomatic, out of which 3 had surgery. The mean ROA was 0.62 cm^2 [range 0.3–0.9]. The mean VAA was 3 sqcm [Range 1.4–4.4]. The mean regurgitation orifice area index [ROAI = (ROA/VAA)*100] was 21.2% [range 6–46%]. There was good correlation between severity of clinical symptoms, ROA and ROAI. The symptom to regurgitation orifice area index correlation was 0.67.
Conclusion: ROAI assessed by RT3CDE is the nearest accurate non-invasive quantification method independent of loading conditions available to date. The method we describe is feasible in children and detailed study is underway to establish reference values in various age groups.

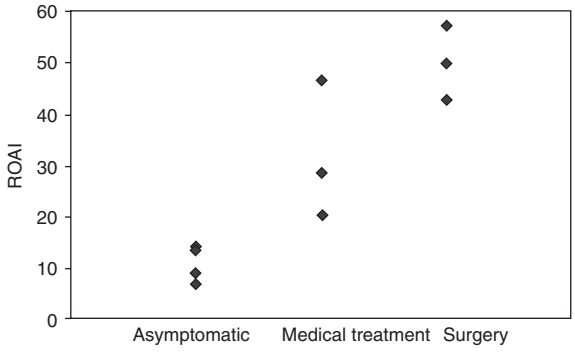

Figure 1. Correlation between ROAI and clinical Symptom ROAI–Regurgitant orifice area Index.

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O-58

Post systolic shortening is related to wall thickness and may indicate the presence of myocardial ischemia in children with hypertrophic cardiomyopathy

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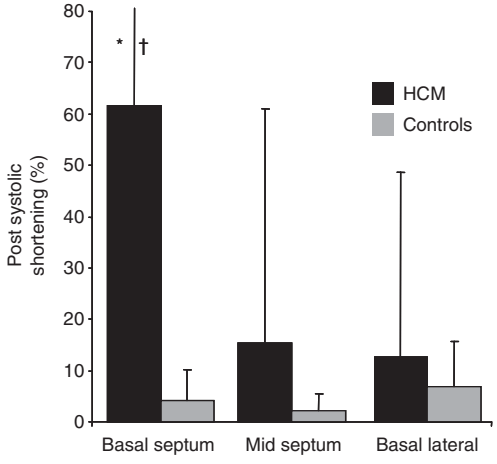
Background: In patients with hypertrophic cardiomyopathy (HCM), it has been shown that wall thickness correlates inversely with deformation. One might speculate that the development of hypertrophy leads to myocardial ischemia. The characteristic "strain pattern" (SP) on the ECG in patients with HCM has been associated with the presence of ischemia. Ultrasound based regional post-systolic shortening (PSS) is an established clinical marker of ischemia in dysfunctional myocardial segments.
Aims: To determine (1) whether PSS is present in children with HCM; (2) whether the presence of PSS is related to wall thickness and the SP on the ECG.
Methods: We examined 41 children with asymmetric HCM, (mean age 12.3 ± 5.5 yrs) and 29 age-matched controls. On ECGs, the presence of SP was qualitatively assessed: 0: no SP; 1: mild SP; 2: marked SP. Standard echocardiograms and tissue Doppler imaging were performed. End diastolic wall thickness (Z score) was measured at the basal interventricular septum and posterior wall. Peak systolic and maximal strain were measured at three myocardial segments: (1) Basal septum at thickest diameter; (2) Mid-apical septum without significant hypertrophy; (3) Basal lateral LV wall. PSS was defined as: (Maximal strain – Peak systolic strain)/Peak systolic strain.
Results: Nine HCM patients had an LVOT obstruction >20 mmHg. An ECG SP considered as "0" was seen in 11; "1" in 13; and "2" in 17 pts. Septal and posterior wall thicknesses were: 4.6 ± 2.4 Z scores and 2.3 ± 2.8 respectively. Heterogeneity in PSS in different myocardial segments is shown in Figure. In HCM pts, PSS was significantly higher at the basal septum compared to mid-apical septum and basal lateral segments; in controls a small amount of homogeneous PSS was noted. A significant correlation between PSS with wall thickness and ECG pattern were found r: 0.44, $p < 0.05$ and r: 0.44, $p < 0.05$.
Conclusions: The degree of PSS in pts with HCM is highest in the most hypertrophied myocardial segments. PSS correlates both with wall thickness and the SP on the surface ECG. These findings may be suggestive of myocardial ischemia as hypertrophy develops.



* $p < 0.01$ vs. basal septum controls,
† $p < 0.05$ vs. midseptum HCM and basal lateral HCM.

O-59

Different patterns of aortic wall elasticity in patients with Marfan syndrome – a noninvasive follow-up study

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Background: Unforeseen aortic complications determine life-expectancy of most patients with Marfan syndrome (MFS). To find out, if there is heterogeneity of aortic involvement among patients, and if there is, to characterise the aortic patterns and the response to long-term beta-blocker therapy, we investigated aortic elastic properties before and under beta-blocker treatment.
Methods: In 46 patients with MFS (age 17.4 ± 11.1 years) and 46 age- and sex-matched healthy control subjects aortic diameters at different sites as well as ascending aortic (AscAo) and descending aortic (DescAo) elastic parameters were determined non-invasively from M-mode echocardiographic aortic images and oscillometric blood pressure measurements using a semi-automated contour finding software. In patients investigations were performed before and, 39 ± 16 months later, during beta-blocker treatment with 0.79 ± 0.46 mg/kg/day of atenolol. The power of several aortic parameters was determined using logistic regression analysis and hierarchical cluster analysis.
Results: Diameters of the aortic root and the AscAo as well as AscAo and DescAo distensibility distinguished between MFS patients and controls with a sensitivity of 85.1%, and a specificity of 87.2%. Cluster analysis revealed 4 patterns of aortic phenotypic expression: a, reduced AscAo elasticity (46% of patients); b, diminished AscAo and DescAo elasticity (17%); c, minimal alterations of AscAo and DescAo elasticity (20%); and d, reduced DescAo elasticity (17%). After 39 ± 16 months of beta-blocker treatment with atenolol aortic elastic properties improved in 21 of 30 patients (70%) and deteriorated in 9 (30%) irrespective of beta-blocker dosage. Improvement was observed in 100% of patients (n = 7, age 5.3 ± 4.2 years) with enddiastolic aortic root diameters between 20 and 30 mm, and in 61% of patients (14 of 23, age 20.5 ± 10.0 years) with root diameters between 30 and 52 mm.
Conclusions: Noninvasive monitoring of aortic elastic properties in patients with MFS distinguished between patients and controls, revealed four patterns of aortic involvement, which may indicate the regions at risk for aortic dissection or rupture, and showed highest "success" of beta-blocker treatment in young patients with aortic root diameters <30 mm and AscAo distensibility >45 kPa-1·10–3.

O-60

Multi-detector CT of congenital vascular anomalies and associated complications in newborns and infants

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Introduction: To assess the value of multi-detector CT (MDCT) for evaluation of vascular anomalies (VA) and associated complications of the tracheobronchial tree in newborns and infants.

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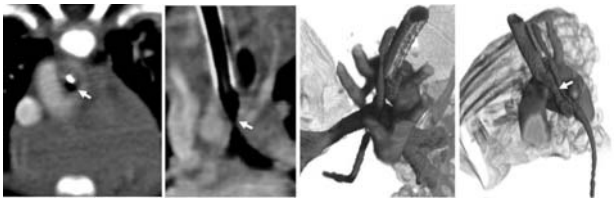
Materials and Methods: 95 children (mean age: 7 ± 7 months) with VA were examined using MDCT (4-, 8-, 16 or 64-row; collimation 0.5–1.25 mm; scan-time 3–30 s), which was performed under controlled ventilation or free breathing. The image quality was rated using a 5-point score. Image findings were correlated to echocardiography, conventional digital catheter angiography (DA), bronchoscopy, and intraoperative findings.
Results: High quality MDCT data were almost free of cardiac and respiratory motion. Images were scored for vascular contrast in 85% and for delineation of the tracheobronchial tree in 95% of all cases as excellent or good, showing a significant improvement with increasing number of detectors. VA morphology and topography in relation to adjacent structures, e.g. tracheal and esophageal compression caused by an aortic ring (see figure) or pulmonary sling, could be assessed exactly and allowed the final diagnosis. Even aberrant vessels such as aorto-pulmonary collaterals (MAPCA) with a diameter of less than 1 mm, could be identified and excellently visualized. Eighty-two percent (77/95) of all patients had benefited from MDCT: in 41 patient DA was neither necessary to perform surgical or interventional planning nor to exclude a VA, in an additional 36 patients radiation doses and sedation time due to interventional procedures could be reduced markedly.
Conclusions: MDCT can now be regarded as the modality of choice as a minimally invasive, robust, and accurate technique for the diagnosis of complex VA even in the group of newborns and infants or critically ill patients. Its accuracy for detecting VA appears at least equivalent to DA while it is more accurate in delineating potential life-threatening complications.



O-61
Cardiac function in children exposed to chemotherapy during intra-uterine life

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Background: The incidence of cancer diagnosed during pregnancy is increasing as childbearing is delayed to older maternal age. Currently chemotherapy (chemo), including anthracyclines, is used to treat malignancies during pregnancy. The effect of this on fetal development and pediatric cardiac outcome has been poorly studied.
Aim: To evaluate cardiac outcome and function in children whose mothers received chemo during pregnancy.
Methods: Eight children (age range: 3–68 months) were included in the study. The mothers were treated for different malignancies. The first chemo was given at 15–36 weeks gestational age (mean 21 weeks). Six of them received doxorubicin (mean dose: 340 mg/m^2). All children underwent a complete echocardiographic exam including tissue-Doppler imaging (TDI). Conventional as well as TDI-derived parameters for cardiac function were assessed. This included measurement of peak systolic myocardial strain rate and strain in different myocardial segments. Data were compared to normal age and gender-matched healthy controls.
Results: No morphologic cardiac abnormalities could be detected in any of the children. All functional parameters assessed, including TDI data were within normal range (see table).
Conclusions: Our study indicates that chemo given during pregnancy is safe and does not result in disturbance of cardiac development. In the short-term cardiac function in the offspring seems not to be affected by intra-uterine exposure to chemo. Long-term monitoring and larger studies are needed to further support this finding.

Table.

	Patients (n:8)	Controls (n:8)	P value
Heart rate	104.5 ± 34.5	100.4 ± 28.3	NS
LVEDD (mm)	29.6 ± 5.5	31.2 ± 6.3	NS
LV mass index (gr/m^2)	68.7 ± 12.9	82.97 ± 15.3	NS
Fractional shortening (%)	36.3 ± 4.5	33.9 ± 3.6	NS
E/A ratio	2.3 ± 0.6	2.3 ± 0.4	NS
IVRT (msec)	47.7 ± 8.7	52.4 ± 9.3	NS
LV ring motion (mm)	11.0 ± 2.4	12.1 ± 1.1	NS
Radial strain rate (1/s)	5.3 ± 0.9	4.5 ± 1.0	NS
Radial strain (%)	65.3 ± 16.7	67.1 ± 9.2	NS
Longitudinal strain rate	2.3 ± 0.5	2.5 ± 0.7	NS
Longitudinal strain	26.7 ± 7.6	29.9 ± 7.9	NS
IVA LV (cm/sec^2)	76.6 ± 16.1	73.0 ± 9.8	NS

Values are presented as mean ± SD; NS not significant.

O-62
Assessment of pulmonary valve and right ventricular outflow tract with real-time three-dimensional echocardiography

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Aim: To demonstrate the feasibility of real time three-dimensional echocardiography (RT3DE) in assessment of the morphology of pulmonary valve (PV) and right ventricular outflow tract (RVOT).
Methods: Fifty patients with adult congenital heart disease (mean age 32 ± 9.5 year, female percentage 60%) were examined with conventional transthoracic two-dimensional echocardiography (2DE) and RT3DE (Sonons 7500 philips, Netherland). For visualization of RVOT and PV, by 2De, parasternal short axis (PSAX) view at aortic valve level and parasternal long axis (PLAX) with superior tilting view were used. The RT3DE data were analyzes offline using TomTec software package (Munich, Germany). Visualization of the morphological features of RVOT and PV was evaluated by 4-point scale (1 = not visualized, 2 = inadequate, 3 = sufficient and 4 = excellent). Measurements obtained by 2DE and RT3DE included pulmonary valve annulus diameter (PVAD), RVOT diameter (RVOTD).
Results: RT3DE helps in sufficient visualization of PV (number of leaflets, commissures and line of closure) in 68% and excellent in 24%. PVAD and PVAA were achieved excellently in 88%. RVOT was visualized excellently in 40% but missed in 48%. 2DE measurements at PLAX were significantly higher than that measured by PSAX (19.7 ± 7.7 mm vs. 16.4 ± 6.8 mm; $p < 0.0001$) for RVOTD and (16.4 ± 7.3 mm vs. 13.5 ± 6.1 mm; $p < 0.0001$) for PVAD. By comparing measurements of 2DE and RT3DE, 2DE underestimated RVOTD (19.7 ± 7.7 mm vs. 22.2 ± 10.0 mm; $P < 0.001$) and PVAD (16.4 ± 7.3 mm vs. 19.4 ± 9.1 mm; $p < 0.0001$). The agreement for score of visualization by RT3DE was fair for the assessment of pulmonary valve (Kappa value: 0.59), good for RVOT (Kappa value: 0.71) and very good for PVA, PPA and DPA (Kappa value: 0.91). There was excellent concordance between all measurements when analysed by two independent observers ($r = 0.94, p < 0.0001$).

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Conclusion: RT3DE could help in assessment of RVOT and PV adding more anatomical details supplemental to routine assessment by 2DE and its feasibility encourage its clinical application for proper selection of therapeutic strategy.

O-63

Tissue doppler imaging, cardiac MRI and pro-BNP are suitable methods to quantify remodelling of the right ventricle after pulmonary valve replacement for severe pulmonary regurgitation

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Background: Analysis of right ventricular function remains difficult, but tissue doppler imaging and pro-BNP may be suitable parameters for the analysis of right ventricular function.
Objectives: Prospective clinical study to compare tissue doppler imaging and pro-BNP with cardiac MRI volumetric measurements.
Methods: Evaluation of the right ventricular function with cardiac MRI, pro-BNP and tissue doppler imaging before, 1 and 6 months after operative pulmonary valve replacement. Patients with severe right ventricular volume overload (RVEDVI, right ventricle enddiastolic volume index $> 150\,ml/m^2$) due to pulmonary valve regurgitation were included.
Results: 16 children, aged 9.9 ± 2.6 years, after total correction of tetralogy of Fallot were included.

	Preoperative	4 weeks postoperative	6 months postoperative
MRI (RVEDVI) [ml/m^2]	$196 \pm 54^{\star}$	–	$111 \pm 24^{\star}$
Pro-BNP [ng/l]	$260 \pm 157^{\star}$	187 ± 121	$155 \pm 110^{\star}$
RV longitudinal peak systolic strain [%]	$-39.5 \pm 3.8^{\star\star}$	$-25.4 \pm 8.2^{\star\star}$	-32.7 ± 6.2
RV longitudinal peak systolic strain rate [1/sec]	$-3.46 \pm 0.73^{\star\star}$	$2.15 \pm 0.53^{\star\star}$	-2.97 ± 0.82

Mean ± standard deviation, $^{\star}p < 0.001$ preoperative vs. 6 mo. postoperative; $^{\star\star}p < 0.05$ preoperative vs. 1 mo. postoperative.

Conclusions: Pro-BNP is elevated in patients with chronic pulmonary valve regurgitation and right ventricular volume overload and decreases in postoperative follow up. Pro-BNP is a suitable parameter for the right ventricular remodelling and correlates with the right ventricular volume relief documented in cardiac MRI. The reduction of longitudinal strain and strain rate of the right ventricle during postoperative follow up can be early detected.

Session 11: General Paediatric Cardiology

O-64

Rheumatic heart disease in children ≤12 years of age: current profile from a developing country

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Introduction: Rheumatic heart disease (RHD) remains a major public health problem in developing countries. According to some reports from these nations the disease is seen at a young age and has a rapidly progressive course. We report our experience in children with RHD ≤12 years who presented to our centre over four years.
Methods: Hospital records of all consecutive children who presented to paediatric cardiology outpatient department of this government funded tertiary care centre from year 2001 to 2004 were reviewed. Records were scrutinized for clinical information, laboratory parameters and echocardiographic data.
Results: A total of 349 children had RHD, this constituted 2.5% of all children seen during this period. Their age ranged from 2–12 years (mean 9.8 ± 2.2 years) and 220 were males. 45.4% (159/349) presented with acute rheumatic fever and carditis. Chronic valvular disease was present in 54.6%, 37% of these had previous history of rheumatic fever. In those with active carditis, arthritis was present in 93%, chorea in 4.4% and subcutaneous nodules in 1.9%. An elevated ESR was seen in 68% and an ASLO titer of >333 units was present in 86%. Over all, significant mitral regurgitation (MR) was present in 74.7%. Aortic and mitral valve lesions were seen in 14.3%, isolated aortic lesion was seen in only 3%. In children with active carditis, echocardiography showed nodules on mitral valve in 25% and mitral chordal rupture leading to severe MR in 3.7%. On a follow up period of 18 ± 15.2 months (available for 142 patients) MR regressed by at least one grade in 67.4% of children who had presented with carditis. No regression of valvular lesion was seen in the other group. Only 48% of children were taking penicillin prophylaxis regularly in both groups combined.
Conclusion: Our data shows that RHD is still common in young children in a hospital setting. A large proportion of these children present with acute rheumatic fever and carditis. Three fourth of children have advanced valvular lesions, possibly due to recurrent attacks of rheumatic fever as less than half of these children are compliant with penicillin.

O-65

Pulse oximetry as a population screening test in detection of critical congenital heart defects in presymptomatic newborns – polish multicentre study

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Objectives:

1. To evaluate the application of pulse oximetry as a screening test in detection of critical congenital heart disease (CCHD) before onset of symptoms in the large, non-selected population of newborns in Poland.
2. To evaluate the psychosocial attitude of mothers to the screening programme.

Material: 27200 asymptomatic newborns born between 15.03.2004 and 31.03.2005 (8% of all live births in Poland), were screened with pulse oximetry in 26 Polish hospitals of I/II/III grade levels of reference: including all 23 in Podkarpackie province (south-east Poland) and three other hospitals in different regions.
Methods:

1. Leg pulse oximetry performed on average at 25th hour of life; saturation (sat.) cut-off value 95%.
2. Psychosocial questionnaire for mothers.

Results:

1. Screening test – True positive: (CCHD in asymptomatic newborns – sat. < 95%) – 7 newborns: CoA (3), HLHS (2),

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TrAtr + CoA (1), TAPVD (1); False positive: (no CCHD; sat. < 95%) −13 newborns; False negative: (CCHD; sat. > 95%) −1 newborn – CoA; True negative: (no CCHD; sat. > 95%) − 27179 newborns. Sensitivity – 87%; Specificity – 99.9%; Positive predictive value – 35%; Negative predictive value – 99.9%.
2. Questionnaire – 99.8% of the questionnaire responders approved the test and suggested it as a regimen for whole newborns' population.

Conclusions:

1. Pulse oximetry test was effective in early detection of presymptomatic newborns with CCHD, particularly with duct-dependent systemic blood flow and anomalous pulmonary venous drainage.
2. Pulse oximetry test was accepted by families and medical staff.
3. High sensitivity and specificity in the study performed in a large, non selected cohort of newborns screened in hospitals of all reference levels, encourage us to propose pulse oximetry as a widely used screening test in detection of CCHD in population of presymptomatic newborns.

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O-66
Respiratory infections hospitalizations in young children with hemodynamically significant congenital heart disease: the 'CIVIC' epidemiologic study, Spain 2004–2005

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Objectives: Primary objective was to evaluate the incidence of acute respiratory infections (RI) hospitalizations in children under 24 months with hemodynamically significant congenital heart disease (HSCHD). Secondary objective was to describe associated risk factors, preventive measures, aetiology and clinical course.
Methods: Epidemiologic, multicentre (16 principal + 54 collaborative hospitals in Spain), observational (descriptive), follow-up and prospective study. Period: October 2004 to April 2005. 791 patients included with follow-up (mean 5 months) in 760; mean age 7 months. Hospitalized (H) and non-hospitalized (NH) were compared using multivariate logistic regression searching for associated risk factors.
Results: 79 patients (10.4%, CI 95%: 8.2% to 12.6%) required hospitalization, with 105 admissions. The incidence rate was 2.1 new hospitalizations for 100 patients and month, with a peak in December (point prevalence 4.1%). Significant associated risk factors were:

Risk Factor for Hospitalization		(CI95%)	
for RI	Risk	Lower limit	Upper Limit
Delection 22q11	8.1	2.5	26.3
Weight under percentile10	5.2	1.6	17.4
Previous respiratory disease	4.5	2.3	8.6
Incomplete RSV immunoprophylaxis	2.2	1.2	4.0
Trisomy 21	2.1	1.1	4.2
Cardiopulmonary bypass(CPB)	2.0	1.1	3.4
Siblings under 11 years old	1.7	1.0	2.9

There was a significant difference (H: 75.9% vs. NH: 86.2%, p = 0.015) in complete respiratory syncytial virus (RSV) immunoprophylaxis. Other vaccinations were similar. Bronchiolitis (51.4%), upper respiratory tract infection (25.7%) and pneumonia (20%) were the main diagnosis. Microbiologic studies performed were: RSV test (93%), culture of respiratory secretions (49.1%), blood cultures (41.5%) and antigen detection (32.1%). In 37 cases (35.2%) an infectious agent was detected: 25 RSV, 5 Streptococcus pneumoniae, 4 Haemophilus influenzae. Patients with incomplete RSV prophylaxis increased 3.05 (CI 95%: 2.14 to 4.35) the relative risk of RSV hospitalization. The median length of hospitalization was 7 days. In 18 patients (17.1%) the clinical course of RI was complicated by ICU admission (n = 17), mechanic ventilation (n = 7), sequelae (n = 4) or death (n = 3).
Conclusions: Hospitalizations due to RI in young children with HSCHD are mainly associated to extra cardiac conditions (genetic, malnutrition, respiratory) and to CPB. RSV was the most frequently detected infectious agent. An incomplete RSV immunoprophylaxis was related to an increased risk of hospitalization.

O-67
Ebstein's anomaly: factors predicting death in childhood a multi-center, long term Dutch study

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Introduction: Several risk factors have been associated with survival of adult patients with Ebstein's anomaly. The objective of this study was to establish prognostic criteria for the outcome (death versus life) of these patients during childhood and adolescence.
Methods: A retrospective study of all Dutch patients born between 1980–2005 followed by a paediatric cardiologist. Excluded were patients with corrected transposition of the great arteries. All possible risk factors for the outcome were obtained. Univariable and multivariate analyses were performed. Kaplan-Meier survival curve was plotted. The probability of death was calculated, and the model was validated using Efron's Optimism correction by bootstrapping.
Results: Of the 93 patients with Ebstein's anomaly, 18 (20%) died and 75 (80%) were alive at the last follow-up. Seventeen patients were already dead by 48 months, one patient by 49 months. Eleven patients were followed shorter than 48 months, leaving 82 (93–11) patients for further analysis of death versus life at the age of 48 months.

The age at presentation (<1 year), dyspnoea, respiratory insufficiency (not Prostin related), hepatomegaly, and the need for inotropics, diuretics, and Prostin at presentation were significantly associated with the outcome. Needing mechanical ventilation, pulmonary atresia, severe pulmonary stenosis, pulmonary valve defect (defined as pulmonary atresia or stenosis), patent ductus arteriosus, ventricular septal defects (VSD) and a peaked P-wave on the ECG were also significantly related to the outcome. Kaplan-Meier survival curve showed no death after 4 years. Using multivariate logistic regression analysis, a significant risk of death at the age of 48 months was associated with the requirement of mechanical ventilation (corrected OR 27; 95%CI 4.4–164), pulmonary valve defect (corrected OR 13.5; 95%CI 2.2–82.3) and VSD (corrected OR 25.2; 95%CI 3.2–196). The corrected area

under the curve (AUC) of the predictive probability of death within 48 months found with the combination of these 3 variables was 0.91 (95%CI: 0.88–0.91).
Conclusions: The overall survival of patients with Ebstein's anomaly during childhood and adolescence has dramatically improved as compared to earlier reports. The combination of requiring mechanical ventilation, pulmonary valve defect and VSD is a strong predictor of death.

O-68

Death rate in hypertrophic cardiomyopathy is higher in the 8–16 year age range than in the 17–30 year age range

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Introduction: Most studies on sudden cardiac death have emanated from adult cardiological units. These studies have reported a dramatic male preponderance in sudden deaths caused by hypertrophic cardiomyopathy (HCM), and there is a general perception that the risk is highest after puberty. However, a recent cohort study on HCM diagnosed in childhood showed that age-specific mortality rates were higher in the 9–13 year age range than in the 16–20 year age range, and that there was no male preponderance in the lower age group.
Methods: We have studied age-specific population-based mortality rates for HCM, as well as for the other common cardiac causes of sudden cardiac death in youth, using the National Swedish Cause of Death Registry for the six years 1997–2002, and cross-referencing with the national data base for forensic post mortem examinations, comparing the 8–16 year age range with the 17–30 year age range.
Results: In the 8–16 year age range HCM had the highest mortality rate at 0.115 deaths per 100 000 age specific population, followed by valvular aortic stenosis, 0.047, dilated cardiomyopathy (DCM) 0.034, myocarditis 0.017, and coronary malformations 0.016 deaths. There were no deaths attributed to arrhythmogenic right ventricular cardiomyopathy (ARVD) long QT-syndrome (LQTS) or coarctation of the aorta in this age range, although 0.066 deaths were coded R96.0 and probably contains LQTS. In the 17–30 year age range the mortality ascribed to HCM was only 0.042 (95% CI: 0.002 to 0.084), i.e. clearly below the mortality rate seen in 8–16 year olds. For HCM the proportion of male deaths were 33% in the 8–16 year age range, and 50% in the 17 to 30 age range. The death rates per 100 000 for DCM (0.150), ARVD (0.054), LQTS (0.021), coronary malformations (0.042) and myocarditis (0.022) all tended to be higher in the 17 to 30 years age range.
Conclusions: The risk of sudden death in HCM is at its highest in the 8–16 year age range, clearly higher than in young adults, and certainly at the lower age range there is no male preponderance for sudden death.

O-69

Impaired flow-mediated vasodilation, carotid artery intima-media thickening and elevated endothelial plasma markers in children after successful repair of aortic coarctation

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Introduction: Increased cardiovascular morbidity is manifested a long time after the repair of aortic coarctation (CoA). By way of impaired flow-mediated vasodilation (FMD) and increased intima media thickness (IMT), surrogate parameters of atherosclerosis, cardiovascular risk factors (RF) can be correlated with early vascular wall changes in children. Quantitative analysis of endothelium-derived soluble markers in plasma (von Willebrand factor (vWf), E-selectin and thrombomodulin) would be another diagnostic and/or prognostic approach. The present study aimed to assess vascular status (FMD, IMT) and to analyse plasma surrogate endothelial markers in children after coarctation repair as compared to controls.
Methods: We examined 28 children after successful repair of CoA versus 30 control subjects. All children underwent identical screening, comprehensive risk factor assessment, and measurement of E-selectin, vWf, thrombomodulin, FMD and IMT.
Results: CoA-children presented significantly ($p < 0.001$) impaired FMD ($4.87 \pm 2.6\%$ versus $10.2 \pm 3.1\%$) and higher IMT values ($p < 0.001$) than the controls, (0.48 ± 0.08 mm versus 0.38 ± 0.01 mm). Concentrations of soluble E-selectin (37.9 ± 14.9 versus 30.8 ± 9.9 ng/ml, $p = 0.08$) and thrombomodulin (34.1 ± 13.1 versus 29.5 ± 7.12 ng/ml, $p = 0.071$) were elevated in CoA-children, while vWf showed no differences between CoA-children and controls. The blood pressure during rest and exercise and the left ventricular mass were significantly elevated, but no additional RF could be identified in CoA-children. Only a remaining pressure gradient related significantly to early vascular wall changes.
Conclusions: The present study documented increased IMT, impaired endothelial function and elevated plasma markers of endothelial activation and injury in children after successful CoA repair. Arterial hypertension and a resting pressure gradient are the major contributing factors to the early atherosclerotic development and should be primary targets for therapy. Sonographic assessment of vascular status and the estimation of soluble endothelial plasma markers, combined with a comprehensive risk factor screening may form a rationale to monitor vascular changes during follow-up studies and therapeutic measures.

O-70

Use of enoxaparin in the treatment of catheter-related arterial thrombosis in infants with congenital heart disease

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Introduction: Catheter-related arterial thrombi are increasingly recognized complications of advanced intensive care and cardiac catheterization techniques used to treat congenital heart diseases (CHD) in infants. Little information is available on the optimal treatment of these thrombotic complications. This study aimed to investigate dose-requirements, efficacy and safety of enoxaparin for the treatment of catheter-related arterial thrombosis in infants with CHD.
Methods: Infants aged 0 to 12 months with CHD treated with enoxaparin for catheter-related arterial thrombosis from January 2002 to December 2005 constituted the cohort for this study. Main outcome measures included dose requirements of enoxaparin, resolution of arterial thrombosis by Doppler ultrasound, and bleeding complications associated with treatment.
Results: Thirty-two infants including 21 newborns and 11 infants aged 2 to 12 months were included in the study. Arterial thrombosis was located in the iliac/femoral arteries in 31 (97%) infants, and related to indwelling catheters and cardiac catheterization in 16 (53%) and 15 (47%) cases, respectively. One infant developed

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aortic thrombosis following umbilical artery catheterization. Arterial thrombosis occurred at a mean of 2.3 days after arterial catheterization. Enoxaparin therapy was started at a mean of 1.9 day after diagnosis. Newborns required increased doses of enoxaparin to achieve therapeutic anti-FXa levels (mean, 1.62 mg/kg/dose) as compared to infants aged 2–12 months (mean, 1.12 mg/kg/dose) ($p = 0.0002$). Complete resolution occurred in 29 (90.5%) infants at a mean of 23 days after initiation of enoxaparin therapy. Partial or no resolution was observed in 1 (3%) and 2 (6%) infants, respectively at a mean follow-up time of 4.3 months. Age, haematocrit, type of arterial catheterization, and duration of cardiac catheterization were not related to resolution of thrombosis. Bleeding complications occurred in 1 (3%) infant.
Conclusions: Results of this study show that enoxaparin is an efficient and safe form of anticoagulation for infants with catheter-related arterial thrombosis, possibly representing a valid alternative to the currently recommended intravenous UFH or thrombolytic therapy. Further studies are required to assess long term benefit of the use of enoxaparin in these infants.

O-71
The metabolic syndrome develops before puberty in obese children

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Introduction: Childhood obesity is a major public health problem and is associated with the metabolic syndrome, defined by body fatness, dyslipidemia, hyperinsulinemia, hypertension, and low aerobic fitness. There is some evidence that foundation of cardiovascular diseases may develop early in life in obese patients. The main purpose of this study was to assess components of the metabolic syndrome in prepubertal obese children.
Methods: This cross-sectional study included 47 prepubertal obese children and 47 matched non-obese controls aged 6 to 11 years (mean age 9.1 ± 1.5 years). We measured fasting blood lipids, glucose and insulin levels; body fatness by dual-energy x-ray absorptiometry; 24-hour systolic and diastolic systemic blood pressure using a Diasys Integra® monitor; maximal aerobic capacity (peak VO_2) by a treadmill test, and anthropometrics.
Results: Groups had similar age, height and pubertal stage (Tanner 1). Obese children had lower cholesterol-HDL (1.11 ± 0.26 vs 1.42 ± 0.32, $p = 0.001$), cholesterol total/HDL ratio (4.12 ± 1.31 vs 3.21 ± 0.72, $p = 0.001$), cholesterol-LDL/HDL ratio (2.83 ± 1.17 vs 2.00 ± 0.64, $p = 0.001$), and VO_2 peak (35.9 ± 6.6 vs 44.2 ± 7.8, $p = 0.001$) compared to controls, whereas insulin levels (4.6 ± 3.2 vs 12.0 ± 6.3, $p = 0.001$) and percentage of body fat (41.4 ± 9.2 vs 21.1 ± 7.4, $p = 0.001$) were significantly increased. In addition, mean 24-hour ambulatory systolic (124.7 ± 14.0 vs 101.8 ± 18.3, $p = 0.001$) and diastolic blood pressure (72.9 ± 7.1 vs 63.8 ± 5.4, $p = 0.001$) were higher in obese children than controls.
Conclusion: Our study demonstrates that components of the metabolic syndrome develop before puberty in obese children. We conclude that childhood obesity should be considered as a chronic disease and benefit of an early treatment. A healthy lifestyle including regular physical activity and a balanced diet should be encouraged. There is also an urgent need to develop prevention strategies in young children.

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O-72
Systemic hypertension is an early complication of childhood obesity and is associated with physical activity and aerobic fitness

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Introduction: Systemic hypertension is a major complication of obesity and may develop during childhood. Physical activity has beneficial effects on arterial function however, little is known in obese children. The main purpose of this study was to measure ambulatory 24-hour BP in prepubertal obese children and to evaluate its relationships with physical activity and cardiorespiratory capacity.
Methods: This cross-sectional study included 41 prebubertal obese children and 41 non-obese controls (mean age 9.0 ± 1.5 yr). We measured 24-hour systolic and diastolic BP and heart rate using a Dyasis Integra monitor with adapted cuffs. Standard deviation scores (SDS) were calculated using reference values of the German Working Group on Pediatric Hypertension ($n = 949$). Other measures included anthropometrics, maximal aerobic capacity (peak VO_2) by treadmill test; past 12 months physical activity by questionnaire; 7-day physical activity count by accelerometer Actigraph MTI; and body composition by dual-energy x-ray absorptiometry.
Results: Groups were matched for gender, age, height and pubertal stage. Compared to controls, obese children had significantly higher body weight, body mass index (BMI), and body fatness. Seven day physical activity count (309.3 ± 42.4 vs 394.4 ± 99.5 cpm, $p = 0.04$), past 12 months physical activity (0.8 ± 1.3 vs 3.9 ± 3.2 hours/week, $p = 0.001$) and peak VO_2 (35.7 ± 6.6 versus 46.1 ± 7.5 ml/kg/min, $p = 0.001$) were significantly lower in obese children than controls. Obese children had significantly higher systolic and diastolic BP than controls (table 1) and 16% of them had hypertension during the night (BP > 2 SDS). All BP measures were positively correlated ($p < 0.05$) with BMI and body fatness, and negatively with physical activity and peak VO_2.
Conclusion: Our study shows that obese children develop high systemic blood pressure early in life and it is associated with body fatness and low physical activity level. They should be encouraged to participate in adapted sports, to improve body composition and prevent premature vascular complications.

Table 1. Ambulatory systolic and diastolic blood pressure.

Groups	Day systolic BP (mmHg)	Day diastolic BP (mmHg)	Night systolic BP (mmHg)	Night diastolic BP (mmHg)
Obese children n = 41	127.9 ± 13.5**	77.7 ± 8.5**	119.7 ± 16.6**	64.9 ± 8.8**
Non-obese controls n = 41	108.4 ± 12.3	67.7 ± 5.2	99.9 ± 8.9	59.4 ± 7.2

**$p = 0.001$.

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Session 12: Intervention

O-73
The Solysafe septal Occluder for ASD and PFO: the first human experience

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Objective: We have previously reported on the animal testing results of a new device for closing Atrial Septal Defects (ASD) with transcatheter technique. This device (Solysafe®) has self-centering characteristics and is constructed of 8 metal wires (Phynox), one plastic wire-holder at each end and 2 polyester membranes in the middle. The device is mounted on a delivery system which gives full control of both ends and introduced through a short 10 F sheath over a 0,018″ guide wire. Once formed and locked in the septal defect, the device assumes a very flat profile. It can be repositioned and removed from the heart, even when the delivery system has been detached, as long as the guide wire is kept in position.
Methods: This device is now undergoing a first human study to evaluate the efficacy and safety of the procedure. The study is divided in two parts; one group with ASD and the second group with Patent Foramen Ovale (PFO) with paradoxical embolism. The study period is 6 months long with follow-up at 1, 3 and 6 months. Hospitals in Germany, Switzerland and Sweden are included in this multi-centre study.
Results: So far, 43 patients have had Solysafe® implantations performed, 29 with PFO and 14 with ASD. The youngest patient was ~6 years old. Eighteen patients have reached the 6 months follow-up. Eight serious adverse events have been reported (all PFO), 3 categorized as related to the device:

- A thrombus formed in the introduction sheath was seen sitting on the device when entering the heart. The device, with the thrombus could be removed and a new device implanted without sequel.
- Atrial fibrillation post implantation, no sequel.
- One device embolized to the aorta via the left ventricle. It opened upon release from the delivery system (not locked?), but could be retrieved percutaneously. A new device was implanted correctly, no sequel.

Conclusion: This preliminary report shows that the Solysafe® Septal Occluder is a promising new design with a very low profile and delivery over a wire. This provides added safety in the positioning of the device within the defect.

O-74
Transcatheter occlusion of ventricular septal defects using a novel Nitinol coil (PFM NitOcclud-VSD-coil)

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The transcatheter closure of ventricular septal defect (VSD) is considered as one of the most demanding interventional procedures. Using a Nitinol coil which was specially designed for the occlusion of PDA we have closed VSD in 45 patients with acceptable results. The most important advantage of VSD closure using the Nitinol coil is the freedom of rhythm disturbances, especially of AV-block.

We report our experience with the new PFM coil which is designed for the occlusion of VSD. Similar to the PDA Nitinol coil the VSD coil has a cone-in-cone formation. Different from the PDA-Nitinol coil the VSD-coil is stiffer and bears polyester fibers on its left ventricular cone. Our experience with the VSD-coil based on transcatheter closure of muscular VSDs (n = 9) and perim. VSDs (n = 35). The minimal VSD diameter ranged from 3 to 8 mm (mean = 4.5 mm). The mean shunt ratio Qp/Qs was 1.6 (1.1–2.7). The age of the patients ranged from 3 to 37 years (m = 14.7 y); the body weights ranged from 10 to 71 kg (m = 44 kg). In all cases the device was implanted using the transvenous approach with a transportation sheath of 6F or 7F. The implantation was abandoned in 1 patient with perimembranous VSD due to unintentional device detachment. The device could be removed using catheter technique without impairment of valves and other cardiac structures. There were no procedural problems or complications at all in the remaining 43 patients. The occlusion rate immediately after coil implantation by angiography was 70%.

Within 24 hours of implantation there was no high velocity residual shunt by echocardiography in 37 of 43 patients (86%). Within 4 weeks of implantation 41 of 43 patients (96%) were free of high velocity residual shunt. On follow-up evaluation, there was no evidence of arrhythmias, other conduction problems, device embolization, valve dysfunction, endocarditis or hemolysis.

The PFM NitOcclud-VSD-coil seems to be a promising device for transcatheter closure of VSDs with a minimal diameter up to 8 mm. Due to the soft implantation technique and flexible device configuration, no conduction problems occurred.

O-75
Transcatheter closure of exotic vascular malformations with Amplatzer devices

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Introduction: Fistulae and arteriovenous malformations usually associated with congenital heart disease have been occluded using different devices with variable degree of efficacy and safety. Recently Amplatzer devices have been used to occlude these lesions.
Objective: To evaluate the safety and efficacy of percutaneous transcatheter occlusion of vascular malformations with Amplatzer devices and the new Vascular Plug device in children.
Methods and Patients: Single institution, retrospective evaluation of clinical records and results of 36 vessels occlusion in 22 patients, performed from January 1999 to December 2005. Catheterization procedures were performed using standard technique, under general anaesthesia. Indication for occlusion was based on clinical criteria. The anomalous vessels were occluded with PDA and ASD occluding devices and vascular plug. To access technical safety and efficacy we recorded the success rate, immediate and late occlusion rate according to angiography and complication rate (embolization, vascular disruption and procedure-related).
Results: All vessels were successfully occluded: 16 pulmonary arteriovenous fistulae (one giant fistula) were occluded using 15 PDA and one ASD devices; four right-to-left venous shunts after Fontan surgery were occluded with four PDA devices; one veno-venous hepatic fistula after Fontan surgery was occluded with a PDA device; two peripheral arterio-venous fistulae were occluded with three PDA devices; three systemic-to-pulmonary shunt were occluded with two PDA and one ASD devices; eight bronchial fistulae were occluded with two PDA and six vascular plug devices and a MAPCA was occluded with a vascular plug. Complete vessel occlusion

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occurred within 10 minutes in 96% of vessels and in 100% within one year after procedure. There were no device embolization, vascular disruption, or procedure-related complications, nor late complications.
Conclusions: All Amplatzer occluding devices demonstrated to be safe and effective for embolization of different vascular malformations in children. For the same purposes, the vascular plug demonstrated to be easier to implant and less time consuming than other devices, with the same efficacy rate.

O-76
Fenestration of extracardiac TCPC tunnels with stents

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Introduction: In univentricular hearts, fenestrations in extracardiac TCPC tunnels allow to decrease right atrial pressure in high risk patients to maintain sufficient cardiac output in the early postoperative period as well as in case of late increase of pulmonary artery resistance. We report about transcatheter creation of fenestrations after TCPC with covered and uncovered stents.
Patients and Methods: From 10/01–12/05, 16 pts.(2–15 y) with Hypoplastic left heart (n = 6), Pulmonary atresia/IVS (n = 1), UVH (n = 5), Imbalanced AVSD (n = 3), TAT (n = 1) received palliative surgery with an extra cardiac conduit (EC). In 10 pts. a surgical fenestration was installed. Due to low cardiac output with pleural and abdominal effusions, catheterization was performed. During catheterization early spontaneous closure of the fenestration was documented in 5 pts. By guide wire technique (n = 5), transseptal needle (n = 9) and radiofrequency perforation (n = 2) the EC was perforated, subsequently ballondilated and afterwards covered stent-grafts 16 × 4 mm (Jo-Med) in 9 pts. and bare stents in 5 pts. were implanted. In one patient an Amplatzer VSD occluder, formerly implanted for closure of a fenestration was perforated and a Genesis stent was implanted through the wire mesh work.
Results: There were no procedural deaths. Pressure in the EC decreased from m = 23.5 mmHg to m = 18 mmHg. Improved haemodynamics and acceptable desaturation to 85–90% were observed. Stent occlusion was observed in 5 pts. 1–7 days afterwards, followed by successful additional implantation of bare stents or stent redilation.
Conclusion: Stent implantation in EC serve as an effective tool for treatment of low cardiac output in the failing fontan patient. Covered stents have the potential advantage of lower risk of bleeding. Stent obstruction can be solved by additional stents.

O-77
Percutaneous management of a Fontan fenestration: the search for the ideal restriction – occlusion device

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Objective: Several devices are currently available for percutaneous closure of a Fontan fenestration. However, no device meets all desired criteria such as ease of loading and deployment, non-bulky low profile with minimal material, non thrombogenicity early and late, perfect closure rate or possibility of partial obstruction.
Methods: A 15 mm PFO star was used as basis because of its nice profile once deployed; it consists of 2 umbrellas mounted on 2 × 15 mm legs. During the study period, some modifications were made: removal of the left disc to further reduce thrombogenicity in the left atrium, increase the size of the LA legs from 2 × 15 mm to 3 × 21 mm, pivot between the left and right umbrella. A partial occluder was made by removing 2 opposite quadrants from the proximal disk.
Patients and Methods: Deployment of a device was attempted in 41 patients 1.7 ± 1.8 years after Fontan operation at 4,63 ± 2,66 years; size fenestration 4,78 ± 0,51 mm (range 4,0–6,0 mm) in a 17,7 ± 2,1 (range 16–22 mm) conduit. Sheath size 9–10 French.
Results: In 3 patients, the device could not be deployed early in the experience: 2 because of repetitive prolapse of small 15 mm distal legs into a small 16 mm conduit; 1 because of anterior orientated fenestration and proximity of the atrioventricular valve. In 27 patients the fenestration was completely closed with increase of saturation from 85 ± 3% to 95 ± 2%. In 11 high risk patients with suboptimal Fontan circulation a partial occluder was used: saturation increased from 80 ± 5% to 90 ± 2%. No gradient across conduit after release of device was observed. In 1 patient a small clot was observed during the procedure; no thrombotic events were recorded during 1–4 years follow-up.
Conclusions: The modified PFO device can safely be deployed in Fontan patients to modulate (decrease or abolish) the right-to-left flow through a fenestration. It has a perfect profile with minimal amount of foreign material, and is therefore non-obstructive and minimally thrombogenic.

O-78
Dilatable pulmonary artery banding – a new hybrid approach to complex ventricular septal defects

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Background: Muscular VSDs may require pulmonary artery banding (PAB), later followed by PA debanding and VSD closure, if this did not occur spontaneously. Reoperation is always under cardiopulmonary bypass and can be complex.
Objective: To assess the results of a dilatable PAB allowing balloon dilatation and possible combination with percutaneous VSD closure, thus avoiding reoperation.
Patients and Methods: Dilatable PAB was performed by use of a 5/0 prolene™ single horizontal mattress stitch, following the Truler's rule, for palliation of complex conditions, including multiple or single large muscular VSDs. The records of 6 consecutive patients who had balloon dilatation of a PAB were analysed; 5 patients had had spontaneous VSD closure after 1 to 9 years of follow up and required PAB balloon dilatation only. One patient had a persistent large muscular VSD after 4.5 years of follow up; this patient had PAB balloon dilatation combined with percutaneous VSD occlusion.
Results: All patients had normal PA pressures distal do the band prior to dilatation. All had successful PAB dilatation; RV mean systolic pressure dropped from 89 (72 to 100) to 34 (32 to 39) mmHg. The patient with a large muscular VSD required two muscular VSD occlusion devices for complete VSD closure; these were implanted at different sessions, PAB dilatation was performed in the first session. No complications occurred and follow up is unremarkable to date.
Conclusions: 1. A dilatable PAB can be achieved by use of a single horizontal mattress stitch with 5/0 prolene™; this leaves the band stable but prepared for future balloon dilatation, offering a safe way of dealing with large/multiple VSDs; 2. In patients with large VSDs, PAB dilatation combined with percutaneous VSD closure is a feasible and safe hybrid approach; 3. This form of treatment may completely avoid or significantly simplify further surgery.

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O-79
Off-pump replacement of the pulmonary valve in large right ventricular outflow tracts: a transcatheter approach using a new device

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Introduction: Percutaneous pulmonary valve replacement (PVR) is under investigation in humans. It is, presently, limited to patients with a right ventricular outflow tract (RVOT) that does not exceed 22-mm in diameter. We report our experience of PVR in animals with large RVOTs using a percutaneous approach.
Methods: Twelve ewes were included in the protocol. We intended to implant a new stent as a first step to percutaneously reduce the diameter of the PA. Immediately following its insertion, we intended to implant a 22-mm valve mounted in a balloon expandable stent inside the restriction. Animals were sacrificed after valve implantation (group 1) and after a mean follow-up of 1.4 months (group 2).
Results: One animal died from an arrhythmia during delivery. The reducer was delivered successfully in all animals except in one where it was incompletely expanded. It allowed the pulmonary diameter to be reduced to 12-mm. In one animal, during PVR, we lost the wire position and the device embolized when trying to reposition the wire through it. Elsewhere, valved stents were successfully delivered inside the PA banding and valves were functioning perfectly at early and late evaluation.
Conclusions: Implantation of a pulmonary valve is possible in ewes through a transcatheter approach when the RVOT exceeds 22-mm in diameter with the use of a new device. The availability of this reducer will enlarge the present indication of PVR in humans.

O-80
Percutaneous pulmonary valve implantation: a cost-effective alternative to conventional surgery

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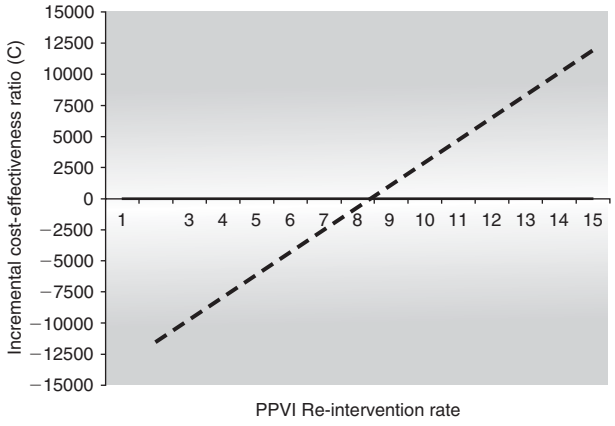
Background: Percutaneous pulmonary valve implantation (PPVI) is a novel, less-invasive alternative to surgery (PVR) for treatment of right ventricular outflow tract dysfunction, a common problem in patients with repaired congenital heart disease. With the introduction of this technology into clinical practice, cost-effectiveness will have important consequences for the planning of health care resources.
Methods: A Markov model was designed to test cost-effectiveness over 25 years in 84 patients who underwent PPVI and a contemporary population of 94 who underwent PVR. Total costs, presented in Euros (€), comprised the cost of the initial procedure and of documented and predicted complications or re-operations. Life expectancy was chosen as the measure of effectiveness. As follow-up was limited, extrapolation over 25-years was based on relevant literature and clinical assumption. One-way sensitivity analysis illustrated the implications of varying projections regarding mortality and re-intervention beyond 5 years. Additionally, we investigated the impact of the PPVI equipment price on overall costs.
Results: The mean cost of one PPVI procedure was €12,491.77, whilst PVR was €18,604.81. This was due to the longer procedural duration; intensive care and hospital stay for PVR. Total first year complication costs were €95,017.42 for PPVI and €205,614.44 for PVR. The PPVI group had a higher re-operation rate whilst the PVR group had higher mortality. The cost per life year gained at 25 years was €1,602.11 in PPVI and €3,426.79 for PVR. The cost of the PPVI equipment would have to increase by 3.87 times before 25-year costs met those of surgery, whilst mortality would have to reach 25% by 25 years before incremental effectiveness was lost. Importantly, it was found that the rate of repeat PPVI could reach 17% per year before PPVI became more expensive (Figure).



Conclusion: PPVI is less expensive and more effective in economic terms than PVR at all time points during a 25-year follow-up period. Furthermore, there is significant scope for repeating PPVI, in case of device failure, before incremental cost-effectiveness is lost. With the imminent introduction of PPVI into clinical practice, these findings are critical for the future planning of health care resources in this patient population.

O-81
Covered stents in patients with complex aortic coarctation

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Objectives: To evaluate the role of covered Cheatham-Platinum stents in patients with congenital heart disease.
Background: There are limited data in the literature about the use of covered stent in patients with congenital heart disease.
Methods: Between January 2004 and December 2005 we implanted covered Cheatham-Platinum stents into 16 patients with aortic coarctation or recoarctation (8 males, median age 20 years, range 8–45 years). Eight subjects had an associated aneurysm, 1 had patent ductus arteriosus, 3 had irregular wall, 4 had a subatretic native aortic coarctation.
Results: Group 1: The stents used ranged from 34 to 45 mm. The mean fluoroscopy and procedure times were 12 ± 8 and 70 ± 15 minutes, respectively. After implantation the gradient across the stenosis decreased significantly (pre-stent median value 37 mmHg (range 20–50 mmHg) vs post-stent: median value 0 mmHg (range 0–10 mmHg) ($p < 0.0001$). Vessel diameter increased from a median value of 6 mm (range 0–11) to a median value of 14 mm (range 10–23) ($p < 0.0001$). Stents were placed in the correct position in all subjects. No complications occurred and on angiographic control the stenoses had been relieved and the aneurysms completely excluded.
Follow-up: During a median follow-up of 8 months (2–21 months) the results were stable without complications.
Conclusion: Covered Cheatham-Platinum stents are very useful tools for treating various complex aortic coarctation.

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OP-1 P-20
Endothelial proliferation is mediated by multiple NOX proteins
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Increased levels of reactive oxygen species (ROS) have been suggested to contribute to the pathogenesis of various cardiovascular diseases, including pulmonary vascular remodeling which is characterized by enhanced proliferative activity of the media and intima. In neutrophils, NADPH oxidases generate large amounts of ROS in the innate immune response. Whereas in neutrophils the subunits p22phox and NOX2 form the catalytic core of the NADPH oxidase, several homologues of NOX2 (NOX1-NOX5) have been identified in vascular cells. In endothelial cells, a NOX2-containing NADPH oxidase has been described to be functionally active. However, the relative contribution of other NOX homologues to endothelial ROS production and proliferation has been controversial. We therefore compared the role of NOX2 with NOX1, NOX4 and NOX5 and in endothelial cells.

NOX2, NOX4 and NOX5 were abundantly expressed at the mRNA and protein levels whereas NOX1 expression was barely detectable. All proteins were present in a perinuclear compartment and colocalized with markers of the endoplasmic reticulum. In addition, NOX2, but not the other NOX proteins, also colocalized with actin at the plasma membrane. Bimolecular fluorescent complementation analysis supported by co-immunoprecipitation showed an interaction between the NOX proteins and p22phox. However, whereas NOX2, NOX4 and NOX5 were functionally active modulating ROS production and endothelial proliferation to a similar extent as was shown using expression vectors and specific siRNA, NOX1 did not significantly contribute to these responses. Taken together, these data show that endothelial cells express NOX1, NOX2, NOX4 and NOX5 in the endoplasmic reticulum where they interact with p22phox. Whereas NOX2, NOX4 and NOX5 were involved in basal endothelial ROS generation and proliferation, NOX1 did not contribute under these conditions. These findings indicate that a complex relation between several NOX homologues controls endothelial function and redox signalling.

OP-2 P-30
Carotid artery elasticity is reduced in patients with Tetralogy of Fallot
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Background: Tetralogy of Fallot (ToF) is a complex congenital heart disease characterized by ventricular septal defect. Animal embrionic data indicate that septation and large artery elastogenesis are related events, but human and clinical data are not available. We tested the hypothesis that elastogenesis was abnormal and large artery elastic function was impaired in patients with ToF.
Methods and results: We studied 23 ToF-patients aged 8–56 yrs, 11 ± 3 yrs after surgical correction and 23 healthy control subjects matched for age and sex. Carotid artery diastolic diameter and pulsatile distension was determined by echo wall-tracking; carotid blood pressure was measured by tonometry, and was calibrated by sphygmomanometric brachial pressure values. Carotid artery pulsatile distension was less in patients then in controls (565 ± 230 vs. 700 ± 205 μm), indicating reduced distensibility and compliance (4.2 ± 1.5 vs. 6.7 ± 2.7 · 10^{-3}/mmHg and 13.4 ± 8.6 vs. 20.0 ± 8.0 μm/mmHg, respectively).
Conclusion: Carotid artery is markedly stiffer in ToF-patients, suggesting that impaired elastogenesis may constitute part of the congenital abnormality. Since carotid artery stiffness has been established as an independent cardiovascular risk factor, this condition may have consequences in the clinical management of ToF-patients.

OP-3 P-67
Fetal pulmonary venous impedance is correlated to thickness of interventricular septum
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Introduction: It has been demonstrated that the pulmonary venous flow pulsatility index (PVPI) is higher in fetuses of diabetic mothers (FDM) than in fetuses of normal mothers (FNM). This may reflect an elevation in PV impedance due to a decreased ventricular compliance secondary to myocardial hypertrophy (MH). The correlation between fetal myocardial thickness and the behavior of PV flow in fetuses of either diabetic or nondiabetic mothers has not yet been assessed.
Objective: To test the hypothesis that a correlation exists between interventricular septum thickness (IVST) and PVPI in fetuses of diabetic and nondiabetic mothers.
Methods: We analysed 194 fetuses (79 FDM and 115 FNM) included of diastolic fetal function studies carried out in a tertiary Fetal Cardiology center. The PVPI was obtained by pulsed wave Doppler by the ratio (maximum velocity – minimal velocity/mean velocity) and IVST was analysed by two-dimensional echocardiography. For statistical analysis, Pearson's correlation was used with a significance level of 0.05.
Results: Mean maternal age was 31.6 ± 6.6 years (19–41). Mean gestational age was 28.1 ± 4.6 weeks (20–39). Mean IVST was 2.9 ± 0.08 mm (1.8–6.2) and mean of PVPI was 1.06 ± 0.42 (0.4–2.6). A linear significant correlation was demonstrated between these two parameters ($r = 0.62$, $p < 0.001$).
Conclusion: Fetal left atrial impedance to pulmonary venous flow, represented by its pulsatility index, is directly proportional to interventricular septal thickness, either in fetuses of diabetic or nondiabetic mothers. It may reflect a progressive modification of left ventricular diastolic function, with higher left atrial pressure as myocardial thickness increases.

OP-4 P-74
Assessment of the evolution of normal fetal diastolic function during mid- and late-gestation by spectral Doppler tissue echocardiography
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Background: High-frame rate spectral tissue Doppler imaging (TDI) may be used to quantitatively assess regional myocardial function during active relaxation in early diastole and passive chamber filling in late diastole. Using this approach, the purpose of this prospective study was to establish reference values for 1) early (Ea) and late (Aa) diastolic longitudinal wall motion velocities and 2) early diastolic

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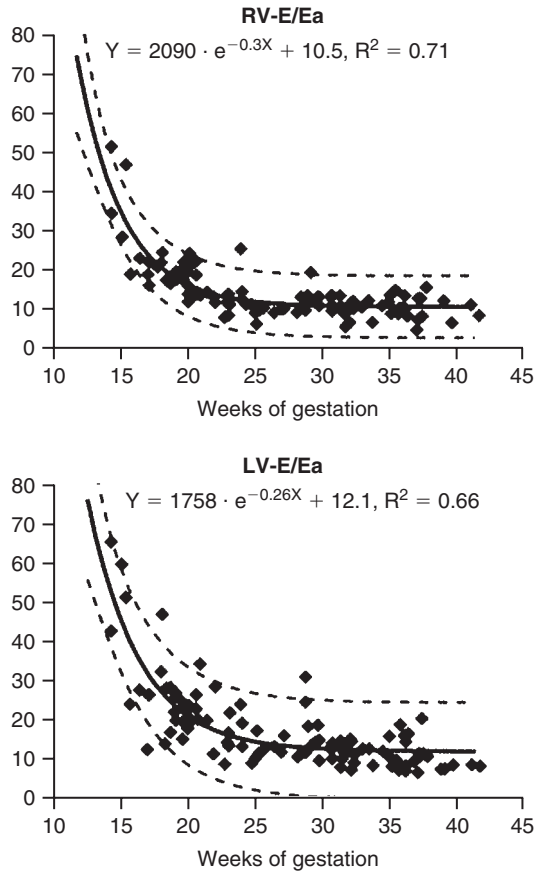
flow/wall motion velocity (E/Ea) ratios in a population of healthy singleton fetuses.
Methods: Peak TDI velocities were analyzed in 114 fetuses (range: 14–42 weeks) at the base of right ventricular (RV) free wall, ventricular septum (VS) and left ventricular (LV) free wall in 4-chamber view sections. The findings were compared to early (E) and late (A) diastolic peak Doppler flow velocities.
Results: Technically accurate recordings were obtained in 107 echo studies (94%) for LV inflow, 105/114 (92%) for RV inflow, 109/114 (96%) for RV free wall, 106/114 (93%) for VS, and 109/114 (96%) for LV free wall. A linear increase in Ea, Aa, and Ea/Aa ratio was documented at all sites with advancing gestation. The peak E flow velocities of both AV valves and the tricuspid peak A-flow velocity increased with gestational age, while the mitral peak A-velocity was unaffected. The highest early and late diastolic flow and longitudinal wall motion velocities were recorded from the tricuspid valve and lateral RV wall respectively, emphasizing the dominant role of the fetal right heart. In contrast, the ventricular septum reached the lowest velocity of lengthening. The ratio of maximal E/Ea velocities, which reflect the relationship between filling pressure and τ, decreased exponentially with advancing gestation due to a more rapid increase in Ea than E, to reach a stable E/Ea relationship only in the early third trimester (figure).



Conclusions: Spectral TDI is a feasible and reproducible imaging tool to quantify ventricular longitudinal myofiber lengthening. There was a strong positive correlation between Ea and Aa velocities and gestational age indicating improved early and late diastolic myocardial lengthening with advancing gestation. Reference charts for TDI velocities were established that will allow identification of fetal diastolic function anomalies.

OP-5 P-66
Experimental assessment of a new lead for in utero pacing for fetal congenital complete heart block
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Objective: The prognosis of congenital complete heart block is very poor when manifested with fetal hydrops as a result of profound heart failure. If low-output failure and/or fetal heart rate cannot be reversed by medical treatment with positive chronotropic drugs or steroids, fetal ventricular pacing seems to be the next logical form of treatment. However, premature labor following hysterotomy remains a major obstacle to open procedures. Therefore, we developed a new lead for fetal pacing, with the aim of minimizing fetal-maternal surgical trauma, thus decreasing the chances for premature labor.
Methods: Five fetal goats had the new lead deployed into the myocardium via a 20-G spinal needle. The prototype is a T-shaped bipolar lead that keeps it securely anchored to the myocardium. Complete heart block was achieved by cryosurgical ablation of fetal AV node. Electrophysiological parameters, hemodynamic and metabolic behavior of the fetus under different heart rates (40 to 140 bpm) were evaluated.
Results: The acute stimulation thresholds were consistently low. The voltage strength-duration curve remained relatively constant at pulse widths >0.6 msec. The stimulation resistance was 1050.4 ± 76.6 ohms, and the sensed fetal R wave was 8.6 ± 5.6 mV. Fetal heart rate bellow 60 bpm was associated to low cardiac output and low blood pressure ($p < 0.05$). Decreased fetal heart rate also determined oxygen saturation drop parallel to low cardiac output, with severe hypoxia below 60 bpm.
Conclusions: The percutaneous approach for fetal pacing with the new lead seems to be compelling and compatible with safe chronic stimulation. It allows a stable fixation to the myocardium that may prevent lead dislodgement after fetal recovery. This study suggests that a gradual increase in the fetal rate beginning on 60 bpm after implantation would be more adaptive and adequately augment the cardiac output. Prevention of premature labor after treatment may contribute for a better result in the treatment of congenital complete heart block associated with hydrops.

OP-6 P-69
Outcome of antenatally diagnosed congenital heart disease: 1-year survival of 221 pregnancies in a single institution over a 5-year period
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Introduction: Congenital heart disease carries a significant morbidity and mortality; however follow-up data on those diagnosed antenatally remains limited. We thus examined the outcome over the first year of life in 221 pregnancies where fetal echocardiography was performed, over a 5-year period.
Results: 87% of cases were referred because of an abnormal 4 chamber view, 5% due to an extracardiac anomaly and 4% with an arrhythmia. 10% of pregnancies had an associated structural abnormality such as congenital diaphragmatic hernia (31%), skeletal abnormalities (9%), exomphalos (9%), and cleft lip (9%). 66% of pregnancies resulted in live newborn, 21% in termination, 5% in intrauterine death, 6% are undelivered, with unknown outcome in 2%. 20% of

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the fetuses had a chromosomal abnormality [Trisomy 21 (47%), Trisomy 18 (16%) and 22q11 deletion (12%)].

Postnatal echocardiography was performed in all live born cases, with post mortem data available in only 12 other cases. The antenatal cardiac diagnosis was accurate in 69%, predominantly correct in 22% and incorrect in 9%. Of the live newborns, 19% had AVSD, 14% HLHS, 7% DILV, 7% VSD, 6% pulmonary atresia and 3% an arrhythmia. 8% had interventional catheterisation in the 1st year of life; of these 42% underwent septostomy, 25% RF perforation of RVOT, 17% pulmonary valvuloplasty, 8% aortic valvuloplasty and 8% balloon of coarctation. Of those undergoing catheter intervention, 42% had repeat catheter procedures, and 67% went on to have a surgical procedure in the 1st year of life. 34% underwent surgery as the primary intervention, 48% of which required a further surgical procedure in infancy. 19% of the cohort died without any intervention, due to elective palliative care and surgical incompatibility as a result of prematurity and other existing co-morbidities. 24% of newborns undergoing an intervention died following the procedure. The overall mortality of newborns with antenatal diagnosis of CHD was 30%.

Conclusions: The 1-year survival following antenatal diagnosis of CHD remains poor and detailed understanding of this is important for the appropriate and effective counselling of parents.

OP-7 P-84

The influence of antenatal diagnosis on postnatal recognition of life-threatening cardiac malformations

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Background: Most infants with cardiovascular malformations are asymptomatic at birth. Earlier diagnosis is likely to lead to a better outcome. This study examines trends in the timing of diagnosis of potentially life-threatening cardiovascular malformations.

Methods: Ascertainment of all cardiovascular malformations diagnosed in infancy in the resident population of one health region in 1985–2004. We considered life-threatening cardiovascular malformations to include all infants with hypoplastic left heart (HLHS), pulmonary atresia with intact ventricular septum (PA/IVS), transposition of the great arteries (TGA), or interruption of the aorta (IAA); and infants dying or undergoing operation in the first 28 days with coarctation of the aorta (CoA), aortic stenosis (AS), pulmonary stenosis (PS), tetralogy of Fallot (ToF), pulmonary atresia with ventricular septal defect (PA/VSD), or total anomalous pulmonary venous connection (TAPVC).

Results: Of 688167 live born infants in the 20 years of the study, 4444 had cardiovascular malformations diagnosed in the first year of life (6.5 per 1000). Cardiovascular malformations were potentially life-threatening in 685 (15%). Over the time of the study, 8% were recognised antenatally, 58% postnatally before discharge from hospital, 31% in life after discharge and 3% after death. Malformations most likely to remain undiagnosed at discharge were CoA (63%), IAA (50%), and TAPVC (47%). Over the 20 years of the study, the proportion of infants diagnosed antenatally increased from around 1% to 20% and no case was first diagnosed after death in the last 10 years. Antenatal diagnosis was made in 21% of PA/IVS and 20% of HLHS but in $<$10% of CoA and TGA and in 0% of TAPVC.

Conclusions: One baby in three with a potentially life-threatening cardiovascular malformation leaves hospital undiagnosed. In recent years better antenatal diagnosis has reduced this proportion to one in four. However, early recognition of such babies is unlikely to be improved by clinical examination and is more likely to come from further improvements in antenatal diagnosis and more widespread adoption of routine pulse oximetry.

OP-8 P-103

The particularities of clinical and electrocardiographic features in families with different genetics variants of long QT syndrome

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The LQTS is a cardiac disorder characterized by a prolonged QT interval and life-threatening tachyarrhythmias. The aim of this study was to determine the particularities of clinical and ECG-features in families with different genetics variants of LQTS, association between the localization (the type) of mutations and phenotype in these patients

Methods: 70 families (famil.) in which probands had prolonged QTc were clinically evaluated using family and medical histories, ECGs, Holter recordings, treadmill. We have tested 70 famil. with LQTS for mutation in 5 genes.

Results: Mutations (mut.) were found in 60% famil.: KCNQ1 in 73.8% famil., KCNH2 in 14.3% famil., SCN5A in 7.1% famil. and KCNE1 in 2.4% famil. In 1 famil. (2.4%) mut. in 2 genes were founded. Patients (pts) with KCNQ1 mutations have an early age at the first syncope, but not patients with KCNH2 mut. (6.3 ± 4.7 and 9.6 ± 5.4 years, $P < 0.05$). In LQT2 pts syncope often occurred in association with emotional (54.5%) and especially acoustic stimuli (81.2%), in LQT1 pts with exercises (83.3%) and especially swimming (33.3%). 50% LQT1 pts died during exercise and 50% – while swimming. 60% LQT2 pts died at rest or asleep without arousal and 30% – after acoustic stimuli. 83.3% LQT3 pts died during asleep and awake. Some HRV parameters were differences between pts with KCNQ1 and KCNH2 mut. Phenotype was more severe in pts with 2 mut. LQTS families with mut. in C-terminal region of KCNQ1 and KCNH2 had a milder phenotype.

Conclusions: The most significant clinical-ECG features which allow suggesting molecular-genetic variant of LQTS are: LQT1 – exercise-related syncope (Se 77%, Sp 78%), decrease of HRV (Se 60%, Sp 66%), typical "T" on ECG (Se 59%, Sp 92%), swimming-related syncope (Se 27%, Sp 100%); LQT2 – typical "T" on ECG (Se 95%, Sp 100%), noise-related syncope (Se 47%, Sp 100%). LQTS families with mutations in C-terminal area of KCNQ1 had a milder phenotype.The phenotype LQTS probably varies depending on specific mutation, amount of mutations (patients with 2 mutations in one and different genes have phenotype more severe) and gene.

OP-9 P-102

The frequency and nature of cardiac anomalies associated with pectus excavatum

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Pectus excavatum (PE) is a common congenital chest wall abnormality in children. Patients with PE may have an associated cardiac defect or impaired function secondary to the pectus deformity. However, the frequency and nature of associated cardiac problems has not been fully elucidated. This study aims to provide data to clarify this issue.

Methods: A retrospective evaluation was made of all patients at the Massachusetts General Hospital less than 21 years of age with PE referred for echocardiography from 1990 to 2005. We examined the medical record for presenting symptoms, associated syndromes (e.g. Marfan, Poland, Ehlers Danlos), and family history of PE, other musculoskeletal abnormalities, mitral valve prolapse, or structural heart disease. Echocardiographic records were examined for presence of structural anomalies, qualitatively determined valvular regurgitation,

aortic root dimension, mitral valve prolapse, and evidence of cardiac compression by the overlying chest wall deformity.
Result: A cohort of 129 patients was identified with a mean age of 12.9 years (range 1 month to 21 years), and a male predominance (77%). Associated syndromes were noted in 10.1%, of which 52% were Marfan or Marfanoid. In 16.5% of cases there was a family history of PE, mitral valve prolapse, Marfan's syndrome or other skeletal deformities. Aortic root enlargement (Z value > 2) was present in 5.4% with aortic insufficiency identified in 7% (trace and mild). Compression or distortion of the right ventricle was seen in 11.6% of patients. Although many patients had trace tricuspid insufficiency, mild tricuspid insufficiency was found in 8.5% and mild pulmonary insufficiency in 5.4% of all cases. Mitral valve prolapse was identified in 11.6%. Mitral insufficiency was present in 37.3% of all patients (34.9% trace, 1.6% mild, 0.8% mild to moderate). One patient had an atrial septal defect (0.8%), and 4.7% had a patent foramen ovale. One patient had a ventricular septal defect (0.8%).
Conclusion: Patients with PE have an increased likelihood of associated cardiac abnormalities. The majority of these appear to be related to geometric changes in chest configuration. In this cohort there is not a strong suggestion of genetic linkage between structural cardiac malformations and musculoskeletal abnormalities.

OP-10 P-83

Neurocognitive effects of open-heart surgery at school age

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Introduction: Children operated for complex congenital heart disease are at risk for adverse neurocognitive sequelae, which mostly become apparent at school age. We designed a longitudinal prospective study to measure these neurocognitive effects.
Methods: School-age patients indicated for open-heart surgery (6–16 years, no comorbidity) were requested to participate. At the time of this abstract, 26 patients (11 boys and 15 girls) completed two assessments: T1: before surgery and T2: one year after surgery. The patients' mean age was 11.9 years (range 6.8–16.1). Neurocognitive functioning was assessed by the WISC-III, the Developmental Test of Visual-Motor Integration, and the Bourdon-Vos test for sustained attention.
Results: At T1 the patients' cognitive functioning was significantly lower than the population mean of 100 (full-scale IQ = 95.1). At T2 a significant improvement was found (full-scale IQ = 99.1, $p < .05$) resulting from a significant increase in performance IQ ($T1 = 96.0, T2 = 101.3, p < .01$).

Regarding visual-motor integration, the patients' mean scores were significantly lower than the population mean of 100 at both assessments ($T1 = 93.1, T2 = 94.6$). No significant postoperative change was found.

The Bourdon-Vos test showed that 18 patients worked slowly compared to normative data at T1. At follow-up, a significant improvement was found in 16 patients. Four patients showed a weak working accuracy at T1. At T2, however, the accuracy of all patients was average or above average.

The complexity level of surgery (Aristotle classification), the duration of cardiopulmonary bypass, and other medical variables were not related to the neurocognitive outcome.
Conclusions: Visual-spatial and visual-motor integration skills seem to be most sensitive to effects of congenital heart disease and open-heart surgery (in accordance with other research). In contrast to other findings, however, our patients performed better after surgery. The cognitive improvement may be mediated by the patients' increased processing speed (and better sustained attention) after surgery.

Our study is still in progress. Fifty patients indicated for open-heart surgery will be compared with patients indicated for interventional heart catheterisation and with healthy controls. In addition to the tests referred to, specific and sensitive perceptual-motor tasks will be evaluated.

OP-11 P-181

Durability of Contegras compared to homografts at 5 years: a subgroup-forming metaanalysis

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Background: Contegra durability is considered well by most of the relevant publications; some authors like them less. Optimal use of the prospectively controlled European Contegra Multicenter Study information for comparisons with homografts is mandatory. We present a subgroup-forming metaanalysis comparing the results of 165 Contegra implantations to homograft durability reports.
Methods: 25 recent homograft durability reports with clearly described endpoints and 5-years-results were selected. These articles indicate 5-year-results of 125 subgroups. The subgroups were characterized by age, diagnoses, conduit size and previous operations, and the endpoints varied broadly from explantation for valve related reason to any reoperation, intervention or bad conduit performance. We modelled each of the 125 article subgroups in the Contegra population by selection, stratification and endpoint formation. We compared the event free rates after 5 years between the Contegra subgroup and the article subgroup. The difference of the freedom-from-percentages indicated the advantage (positive percentage) or disadvantage (negative percentage) for Contegras after 5 years.
Results: Including all comparisons equally, the median freedom from events at 5 years is 2.4% better for Contegras (interquartile range 13.9%, mean 4.7). Counting only 1 average value per article of all its subgroups, the median of the 25 articles is −0.2%, the average 3.9%. Roughly sorted by endpoints (degeneration, explantation, reoperation or death, any reoperation), the average/median advantage for contegras is 2.8/−0.4; −0.7/−0.6; 7.5/7.4; 3.1/−1.1%, respectively. Compared to aortic homografts, Contegras show an advantage of 25,5/27.4%, for pulmonary homografts there is a Contegra disadvantage of −1.5/−1%.
Conclusion: The Contegra results obtained within the European Multicenter Study are well comparable to homograft results as cited in literature.

OP-12 P-165

Reversible pulmonary artery banding V: intermittent X continuous ventricle overload of goats

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Objective: We sought to assess two different programs of systolic overload (continuous and intermittent) on the pulmonary ventricle

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(RV) of young goats with the aim of inducing rapid ventricular hypertrophy.
Methods: Twenty one 60-days-old goats were divided in three groups: control (n = 7, wt = 7.5 ± 1.9 kg), continuous stimulation (n = 7, wt = 9.3 ± 1.4 kg), and intermittent stimulation (n = 7, wt = 8.1 ± 0.8 kg). Pressure load was achieved by an adjustable pulmonary artery banding (PAB) device. The device was implanted on the main pulmonary artery and inflated percutaneously so that a 0.7 RV/LV pressure ratio was achieved. Echocardiographic and hemodynamic evaluations were performed every day. Systolic overload was maintained for 96 hours in the continuous group, while the intermittent group had four 12-hour periods of systolic overload, alternated with a resting period of 12 hours. The animals were then killed for water content evaluation.
Results: Both groups achieved a significant increase of RV mass (55.6% in the continuous and 88.9% in the intermittent group, $p < 0.05$). However, significant increase of the septum mass was observed only in the intermittent group, as compared to the control group ($p < 0.05$). Similarly, a significant increase in the RV wall thickness was observed in both groups ($p < 0.05$). Nevertheless, RV dysfunction was more frequently observed in the continuous group, with a trend for a greater RV diastolic volume and perimeter when compared to intermittent group. There was a trend for a lower RV ejection fraction in the continuous group throughout the protocol. There was no significant difference in RV myocardial water content between the study groups and control group.
Conclusions: Adjustable PAB has permitted RV rapid hypertrophy during a short period of time in both groups. Nevertheless, intermittent systolic overload has permitted a hypertrophic process more comprising than continuous overload. This study suggests that preparation of the pulmonary ventricle with intermittent systolic overload, similar to a fitness program, might provide better results for 2-stage arterial switch operation not only in patients with transposition of the great arteries (TGA) beyond the neonatal period but also in those who present with systemic ventricular failure in corrected TGA or after failed atrial baffle operations.

OP-13 P-172
Surgical treatment of HOCM in children with severe hypertrophy

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Objective: Treatment of children with HOCM is complicated by several factors including noncompliance medications and an increased risk of sudden death. The classic Morrow technique is not effective for HOCM children with midventricular and RVOT obstruction and extreme left ventricular hypertrophy.
Methods: The presented excision of the asymmetrical hypertrophied area of the interventricular septum causing obstruction of LVOT and RVOT simultaneously and midventricular obstruction is made from conal part of right ventricle transversely and anteriorly of the Lancisi muscle and moderator band but not through the whole thickness of IVS, that is, without penetration into the left ventricular cavity. 35 pediatric patients underwent this procedure. Ages ranged from 8 to 15 years (mean, 12.5). The midventricular obstruction was noted in 19 children, isolated RV obstruction in 1 patient. In 12 operated children the obstruction of LVOT and RVOT was noted simultaneously. The follow-up period was 38 ± 7 months.
Results: The mean echocardiographic intraventricular gradient in LV decreased from 78.9 ± 5.9 to 12.7 ± 5.2 mmHg ($p < 0.001$), the mean value of gradient in RVOT also reduced. In patient with isolated RVOT obstruction gradient decreased from 60 to 8.7 mmHg. Echocardiographically determined septal thickness was reduced 31.7 ± 6.5 versus 16.1 ± 4.6 mm ($p < 0.001$). Follow-up echocardiography showed reduction of left atrial size from 46.7 ± 7.1 to 38.5 ± 6.2 mm ($p < 0.01$). Magnetic resonance imaging showed an increase of the diastolic volume of RV and stroke volume. Sinus rhythm was noted in all children.
Conclusion: This method is a safe and effective technique for surgical treatment of pediatric patients with severe hypertrophic obstructive cardiomyopathy unresponsive to medical management.

OP-14 P-180
Outcomes after surgical repair of complete atrioventricular septal defect in children: factors associated with increased atrioventricular valve regurgitation over time

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Background: We sought to determine long-term outcomes, especially concerning atrioventricular valve (AVV) function and reoperation (REOP), after repair of complete atrioventricular septal defect (CAVSD) in children surviving into adulthood.
Methods: Records of all children (N = 51) born after 1948 who survived biventricular repair for CAVSD at our institution and who were followed for >18 years of age were reviewed. Kaplan-Meier method determined freedom from late death and REOP. Serial echocardiographic measurements (n = 271) were examined for trends over time. Factors associated with increased grade of atrioventricular valve regurgitation (AVVR) were sought using general estimating equations for ordinal outcomes.
Results: CAVSD type was Rastelli type A in 56% and Rastelli type C in 44%. A single papillary muscle was found in 6% of patients. A total of 66% of patients had Downs' Syndrome. Associated cardiac lesions included left (L) ventricular outflow tract obstruction in 7%, right ventricular outflow tract obstruction 6%, and coarctation of the aorta in 2%. Patients underwent repair at a median age of 1.1 years (y) (range: 1 month–9 y). One-patch technique was used in 66%; two-patch technique was used in 34%. LAVV cleft was completely closed in 59%, partially closed 17%, and left open in 24%. No deaths occurred during a mean follow-up interval of 21 ± 6 y, except for one accidental death at age 19 y. REOPs were undertaken in 21 patients, including 6 LAVV repairs, 7 LAVV replacements, and 4 subaortic resections. Overall freedom from REOP was 74% at 18 y following initial repair, with one occurring beyond age 18 y. LAVVR grade increased in all patients nonlinearly ($P < 0.003$). Factors associated with a higher grade of AVVR at all time points were LAVV leaflet prolapse ($P < 0.03$) and lower age at initial repair ($P < 0.03$). At 20 years from repair, 48 patients (92%) had no cardiac symptoms and were asymptomatic.
Conclusions: Adults who have survived repair in childhood of CAVSD have no cardiac-related mortality and excellent functional status, but REOP during childhood for residual LAVVR is an important source of morbidity. Complete closure of the LAVV cleft at initial repair may decrease the risk of worsening regurgitation and subsequent REOP.

OP-15 P-169
Mitral valve repair using a biodegradable ring in children

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Objectives: Although conventional rings currently available on the market respond to the needs of the adult population, no annuloplasty ring has thus far been designed for children, in whom preservation of the growth potential of the native annulus is an important issue in terms of long-term stability of valve repair procedures. The aim of this study is to report the midterm results of conservative mitral valve repair supported by a new biodegradable mitral ring (Kalangos-Bioring®) in children (ring size <26 mm).
Methods: Between 2000 and 2006, 25 consecutive patients aged 5.2 ± 1.3 years underwent mitral valve repair associated with degradable annuloplasty ring placement (Kalangos-Bioring®). Mitral valve disease leading to surgery included incompetence ± stenosis due to rheumatic fever (14 pts) or congenital heart disease (10 pts) or bacterial endocarditis (1 pt). Mean preoperative mitral regurgitation was graded on surface 2D-Echo 3.2 ± 0.7. Follow-up patient examination was performed at least with 6 serial surface 2D-Echo investigations at 1, 3, 6, 8, 12 and 24 months postoperatively.
Results: All patients survived the operation. During a mean follow-up time of 64 ± 17 months, the mean mitral regurgitation (0.4 ± 04) was significantly reduced compared to preoperative one ($p < 0.001$), whereas a mean gradient of 1.6 and 4.2 mmHg was recorded in the rheumatic and congenital group, respectively. Two patients required mitral valve replacement and 1 patient is awaiting surgery for significant recurrent mitral incompetence, accounting for an overall reoperation rate of 11%. No deaths occurred.
Conclusion: The support provided by a biodegradable ring may optimize the results of conservative mitral valve repair in pediatric patients and provide satisfactory midterm results.

OP-16 P-175
Factors predicting surgical treatment and postoperative survival of paediatric patients with Ebstein's anomaly

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Objective: To establish a) factors that predict the probability of having surgery for children with Ebstein's anomaly (EA) and b) prognostic criteria for mortality of the operated patients during childhood and adolescence.
Methods: All records of Ebstein's patients born between 1980 and 2005, from six paediatric heart centres in the Netherlands were studied. Demographic, diagnostic and clinical parameters of the operated group were assessed. Cox regression analysis was used to study a) factors predicting surgery, b) factors predicting postoperative death and c) probability of postoperative death within 36 months after first surgery. Relative Risk (RR) and 95% confidence limits (95%CL) were calculated.
Results: On a total of 93 patients with EA, 30 (32%) patients had been operated on. Median age at diagnosis and at the first operation was 0 month (range 0–112) and 38.5 months (0–185), respectively. The patients had their first surgery between 1989 and 2005. Fourteen (46.7%) patients were operated on more than once (twice 10, three times 4). All but four underwent a Glenn or Fontan procedure. Significant predictors for surgical treatment were: dyspnoea (RR 9.5; 95%CL 2.6–33.8), pulmonary valve atresia or stenosis (=pulmonary valve defect) (RR 4.4; 95%CL 1.7–11.4), patent ductus arteriosus (RR 3.8; 95%CL 1.4–10.0) and the use of digoxin (RR 2.7; 95%CL 1.0–7.2) and Prostin (RR 5.5; 95%CL 2.1–14.7).

The median postoperative follow-up was 45 (6–149) months. Thirteen (43.3%) patients had complications, e.g., arrhythmias and pericardial effusion. Four patients died after first (within 1 month) and 2 after their second operation, due to cardiogenic shock (n = 5) and untractable hypoxemia (n = 1). Death in the operated group was significantly associated with dyspnoea at diagnosis (RR 21.6; 95%CL 3.8–124.0). Without dyspnoea the probability of death within 36 months after the first surgery was 11% (0–21.3%) and with dyspnoea 92% (26.3–99.02%).

The overall survival of the un-operated group (n = 63) versus the operated group (n = 30) was 81% and 80%, respectively (p = 1.0).
Conclusion: In this large multicenter cohort, only one third of the patients with Ebstein's anomaly were operated on. Clinical and echocardiographic factors predicting surgical treatment were established. Dyspnoea at diagnosis was strongly associated with postoperative death.

OP-17 P-7
Comparison of cryoablation versus radiofrequency ablation for pediatric supraventricular tachycardia: a clinical decision analysis

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Introduction: Evidence-based medicine seeks to integrate the best research evidence with clinical expertise and patient values in the context of clinical decision-making. Cryoablation (CRYO) of pediatric SVT has different success and complication rates than radiofrequency (RF) ablation.
Methods: A clinical decision analysis model representing plausible outcomes was constructed for CRYO and RF. Probabilities of outcomes were derived from published reports. Relative values for outcomes were obtained through standard gamble interviews. Probabilities were multiplied by values and summed across the different outcomes to give final (probability and value weighted) scores (ranging to 1 for a perfect procedure). Final scores were further adjusted for procedural disutility (magnitude of preference for a specific procedure type) and perceived procedural mortality. Sensitivity analyses were performed to determine threshold values that would alter the preferred decision.
Results: Interviewees (n = 42) included 15 lay persons, 13 pediatric cardiology trainees and 14 pediatric cardiologists. Before the interview, 55% favored CRYO over RF for ablation of AV node reentry (AVNRT), and all favored CRYO for accessory pathway (AP) ablation. Respondents estimated the mortality associated with CRYO at a mean of 0.09% and for RF 0.16% (p = 0.001). From the literature, the probability of procedural success was estimated at 87.5% for CRYO vs. 97.2% for RF, arrhythmia recurrence at 9.2% for CRYO vs. 7.7% for RF, and complications at 0.7% for CRYO vs. 2.4% for RF. Overall final probability and value weighted scores were similar for CRYO (median 0.9996) vs. RF (0.9991; p = 0.45), but were significantly higher for CRYO after adjusting for estimated mortality (p = 0.02). Final scores weighted for disutility were similar for CRYO (0.999) vs. RF (0.999; p = 0.65) for AVNRT, but significantly better for CRYO (0.9996) vs. RF (0.990;

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$p < 0.001$) for AP, with similar findings after further adjustment for estimated mortality. Sensitivity analysis showed RF to be favored under the following scenarios for CRYO: procedural success <51%, recurrence >38%, complications >1.9%.
Conclusions: When outcomes and their values are analyzed in a systematic manner, CRYO is favored over RF for ablation of pediatric SVT. Interviewees chose the reduced complications of CRYO over the acute success and lower recurrence rates of RF.

OP-18 P-8
ICD-implantation in infants and small children: the extracardiac technique

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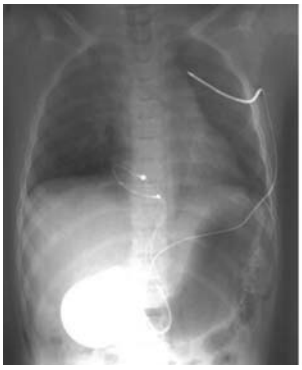
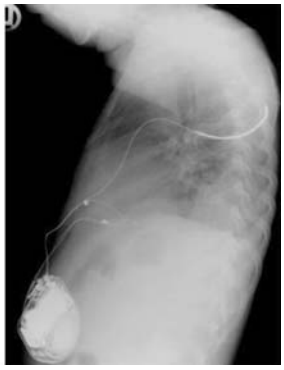
Introduction: There is no convincing concept for implantation of an internal cardioverter-defibrillator (ICD) in infants and small children. This study was performed to assess the efficacy and safety of a new extracardiac ICD implantation technique in pediatric patients.
Patients and Methods: Since July 2004 an extracardiac ICD-system was implanted in 7 patients (age: 0.3–8 years; body weigt: 4–25 kg). Under fluoroscopic guidance a defibrillator lead (Medtronic Transvene 6937 SN, 35 cm) was tunneled subcutaneously starting from the anterior axillar line in a subscapular fashion along the course of the 6th rib until almost reaching the vertebral column. After a partial inferior sternotomy, bipolar steroid-eluting sensing and pacing leads (Capsure Epi 4968, 25 cm) were sutured to the atrial wall (n = 2) and to the anterior wall of the right ventricle (n = 7). The ICD device (Marquis DR 7274) was implanted as "active can" in the upper abdomen (see Figure). Sensing, pacing, and defibrillation threshold (DFT) as well as impedance were verified intraoperatively and 3 months later, respectively.
Results: In 6/7 patients, intraoperative DFT between subcutaneous lead and device was <15 J. In the seventh patient ICD implantation was technically not feasable due to a DFT > 20 J. During follow-up (mean 13.1 months) appropriate and effective ICD discharges were noted in two patients. DFT remained stable after 3 months in 3/5 patients retested. In two patients a shift of the defibrillation lead position resulted in an increase of the DFT requiring revision.
Conclusions: In infants and small children, extracardiac ICD implantation was technically feasible. Experience and follow-up are still limited. The course of the DFT is unknown facing further growth of the patients.





OP-19 P-153
Illustration of complex congenital heart morphology in the living by individual RepliCast and RepliCardio models

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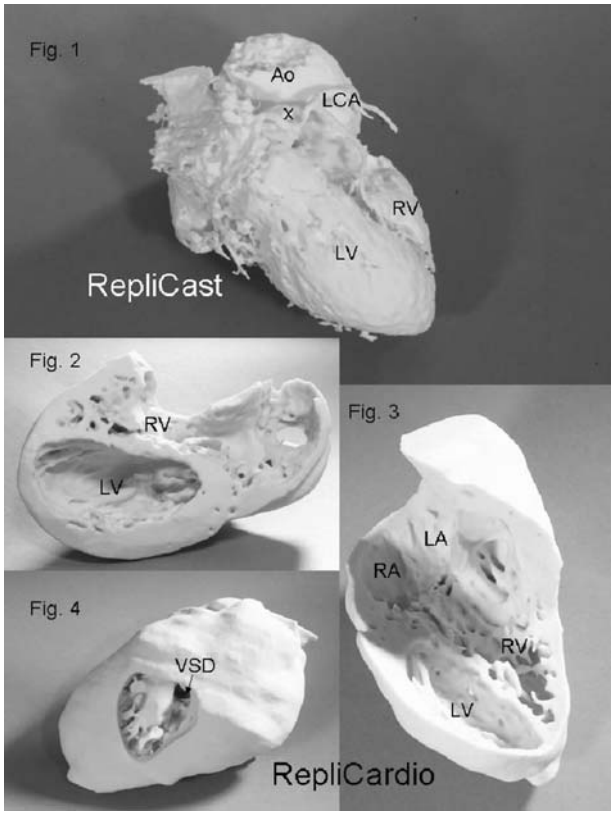
Introduction: Up to now, only pathologic specimen could illustrate the morphology of complex congenital heart disease in detail. In the living, we managed to produce the first models of casts (RepliCast) and muscle and vessel walls (RepliCardio).
Methods: On MRI data sets of three patients with complex congenital heart disease and one normal subject the epicard was segmented with automatic interpolation by software developed at our institution. Additionally, automatic endocardial and endoluminal border detection was performed after determination of the threshold value. The RepliCast was printed out 3-dimensionally on basis of the virtual cast. For the RepliCardio the virtual cast was subtracted from the epicard. As the opening of the surface was mandatory for production and view of the inner surface a software tool was developed allowing for accurate placing of fenestrations and curved cut lines. The results could be viewed as virtual 3-dimensional surface models using red-blue-glasses before printout.



Results: RepliCasts showed the inner volumes and even the course of the coronary arteries (Fig. 1: LCA) and the positions of abnormal structures such as an atretic subpulmonary outflow tract (x). The first patient demonstrated a morphologically left ventricle (LV) on the right side and an aorta (Ao) rising from the morphologically

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right ventricle (RV) on the left side. The virtual cutting instrument allowed for separating of the RepliCardios into two halves (Fig. 2: top halve; Fig. 3: basal halve) as well as exact positioning of fenestrations (Fig. 4) giving view for example on papillary muscles and the rim of the ventricular septal defect (VSD). Thereby, the view into the open heart could be simulated for surgeons and cardiologists. Additionally, the touching of the models eased the imagination of complex morphology by far.
Conclusions: RepliCasts present inner volumes and RepliCardios reflect the muscle and vessel walls of living hearts. The presented technique is expected to be invaluable for diagnosis, treatment planning, patient education, and as reference during repeat heart surgery.

OP-20 P-39
Free-breathing time-resolved parallel 3D MRA of congenital heart disease in infants and small children: a validation study
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Introduction: Until now, contrast-enhanced MR angiography (MRA) could only be considered an alternative modality to conventional catheter angiography (CCA) in older and cooperative children, since a breathhold acquisition was required. With parallel imaging the acquisition time of MRA can be significantly shortened without reducing the spatial resolution. The aim of this study was to assess the feasibility of free-breathing time-resolved parallel 3D MRA in the preoperative evaluation of congenital heart disease (CHD) in infants and small children.
Methods: Twenty-two children (median [range]: 1 year [1 week to 7 yr]) with suspected vascular anomalies were examined: TOF with PA (n = 10), PS (n = 4), vascular ring (n = 6), coarctation (n = 2). Time-resolved 3D MRA was performed at a 1.5 T magnetom (Symphony, Siemens) during free breathing. If necessary, the children received a conscious sedation: 3D FLASH pulse sequence with parallel imaging: TR/TE/α = 2.4 ms/0.9/60°, GRAPPA, factor 2, 24 reference lines, FOV: 300 mm, matrix: 192, spatial resolution: $\sim 1.4 \times 1.4 \times 2.0\,mm^3$, TA: ~2.8 s, 0.2 mmol Gd-DTPA/kg b.w. i.v. @ 1–2 ml/s. Image data were post-processed using 2- and 3-dimensional reconstructions (MIP, MPR, volume-rendering) and compared to CCA (n = 19).
Results: In all patients, the image quality was of diagnostic value. Despite the free-breathing acquisition no relevant motion artifacts were observed. The diagnostic information of MRA regarding the visualization of vascular anomalies was nearly equivalent to CCA. This included the visualization of smallest aberrant vessels such as multiple aortopulmonary collateral arteries (MAPCA). Only in 2 patients CCA showed one further MAPCA (diameter <1.5 mm). In addition to the visualization of vascular morphology, the high temporal resolution of the MRA technique facilitated the functional characterization of the vascular anomalies. This included the definition of pulmonary venous drainage and pulmonary perfusion in patients with TOF+PA (MAPCA n = 7; PDA n = 3).
Discussion: Our results indicate the feasibility of free-breathing time-resolved 3D MRA as a non-invasive and non-ionizing imaging tool also in infants and small children with CDH in their preoperative evaluation. While the accuracy for the visualization and characterization of vascular anomalies appears equivalent to CCA, it seems to be more valuable than CCA regarding the visualization of complex morphology.

OP-21 P-34
Real-time 3-D echocardiography: echocardiographic–anatomic correlation
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Introduction: Real-time 3-D echocardiography is now available commercially. However, the correlation between the 3-D image and the cardiac morphology remains unclear.
Aim: We performed experiments in an open-chest animal model to compare real-time 3-D echo images with the anatomical specimen to better understand the appearance of cardiac structures in the 3-D images.
Methods: Six pigs were anesthetized, intubated, ventilated and cared for according to the animal investigation protocol. The heart was exposed and 3-D images of cardiac structures were acquired using a standard Philips Medical System Live 3D Ultrasound system. At the end of each experiment the pig was euthanized and the heart was fixed in situ under a distending pressure similar to the venous pressure. The heart was dissected and photographed with a digital camera.
Results: 3-D images of venous structures and the atrial wall corresponded exactly to the specimen. Chordae tendinea of the AV valves appeared thicker than the specimen so that we initially mistook them for papillary muscles. The rete at the chordae-leaflet junction had the expected appearance of chordae because of drop-out of the thin leaflet material. The ventricular muscle corresponded well with the specimen, with trabeculations and muscular ridges apparent, as did the semilunar valves and arteries.
Conclusion: We conclude that the correspondence between real-time 3-D echo and anatomic specimens is excellent but care must be taken to correctly identify the components of the AV valves.

OP-22 P-44
Function of the interventricular septum in patients with systemic right ventricle – a tissue Doppler study
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Introduction: After Mustard or Senning repair for transposition of the great arteries (d-TGA), qualitative assessment of the systemic right ventricle (RV) remains difficult. The function of the interventricular septum (IVS) is closely connected to right ventricular function. Based on myocardial Doppler echocardiography, strain (S) and strain rate (SR) can describe regional myocardial function. We wanted to evaluate right ventricular function and determine the role of the interventricular septum (IVS) in Senning patients using tissue Doppler echocardiography.
Methods: We performed an echocardiographic examination in 11 patients (age 19.3 ± 3.4 years) several years after Senning procedure (18.9 ± 3.6 years post surgery). Doppler myocardial imaging data were recorded and regional myocardial function was analysed. Velocity curves, regional SR and S profiles of the IVS and both the right and left ventricular (LV) free wall were obtained for analysis of longitudinal function. For radial function, data of the IVS and LV free wall were analysed. In addition, right ventricular ejection fraction was determined using endsystolic and enddiastolic planimetry of the RV (EF = (Ad−As)×100/Ad).
Results: 1. Myocardial velocities of the IVS were found to be reverse in Senning patients compared to a normal population (towards the

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RV instead away from the RV). 2. Radial strain of the IVS was increased in Senning patients (44 ± 16% vs. 16 ± 7%) and radial strain of the left ventricular posterior wall (LVPW) was found to be decreased (23 ± 11% vs. 52 ± 18%). 3. Also, radial strain rate of the septum was higher than radial strain rate of the LV (SR IVS 3.40 ± 1.30 /s; SR LV 2.09 ± 1.29/s). The ratio of radial SR IVS/LV showed a negative correlation with RVEF ($r = -0.51$) indicating an increased radial septal function in right ventricles with an impaired function.

Conclusion: Septal radial function is increased after Senning procedure, which may be due to the remodeling process of the RV. Further studies on this finding are needed to evaluate the clinical importance.

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OP-23 P-42
Left ventricular growth is impaired in children after repair of tetralogy of Fallot – a longitudinal echocardiographic study

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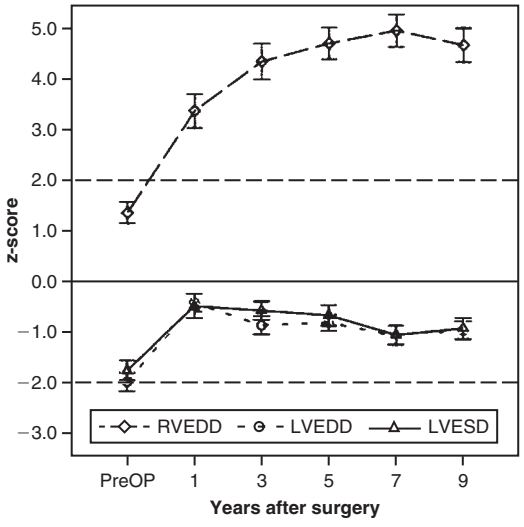
Introduction: Pulmonary regurgitation (PR) is a frequent long-term complication after repair of tetralogy of Fallot (TOF). The aim of the study was to determine long-term echocardiographic measurements of left and right ventricle (LV, RV) in children after repair of TOF in relation to somatic growth. We hypothesized that by "interventricular interaction" LV growth is impaired by chronic RV volume overload resulting from PR.



Methods: Retrospective longitudinal cohort study of 88 consecutive children who had repair of TOF between 1981–2001. Outcome variables were M-mode echocardiographic measurements of LV enddiastolic (LVEDD) and endsystolic diameter (LVESD) and RV enddiastolic diameter (RVEDD). Ventricular diameters were standardized by z-scores in relation to body surface area using published reference values. Covariables were gender, systemic to pulmonary shunt, type of surgery, VSD patch leak, degree of PR. Correlations between time courses of LV and RV diameters and influence of covariables were analyzed using mixed multiple regression models.

Results: LV diameters were significantly decreased preoperatively, improved following surgery, but decreased again over time. RVEDD significantly increased beyond normal range over time following repair surgery, corresponding to the degree of PR. Mean LV diameters were significantly lower than normal population means at all times. There were only trends for a correlation between LV and RV diameters. The longitudinal course of RVEDD was significantly positively correlated with the degree of PR ($p < 0.001$), while courses of LVEDD ($p = 0.03$) and LVESD ($p = 0.01$) were significantly negatively correlated with the degree of PR. Mean LVEDD were significantly increased in patients who had required a systemic to pulmonary shunt compared to patients without shunts ($p = 0.012$) and were similar to the normal population mean by 7 years after repair surgery. Mean LVESD courses showed a similar trend.

Conclusions: LV impairment is associated with severity of PR and may contribute to overall cardiac dysfunction. RV size and degree of PR are the main predictors of the need for pulmonary valve replacement. In line with the concept of LV-RV interaction to follow LV growth in TOF after repair surgery can be a new additional criteria helping to decide about the optimal timing for reoperation.

OP-24 P-48
Assessment of right ventricular function by Tissue Doppler Imaging in children operated on for tetralogy of Fallot

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Introduction or Basis or Objectives: To analyze systolic, diastolic and global right ventricular (RV) function in children operated on for tetralogy of Fallot using Tissue Doppler Imaging technique (TDI).

Methods: Study population consisted of 73 children divided into two groups: Group 1: 30 children operated on for tetralogy of Fallot (Post TF), aged 96.1 ± 61.8 months. Mean pulmonary regurgitation was +1.7 ± 0.6 of 4 grades and mean pulmonary pressure gradient 33.2 ± 12.4 mmHg. All patients were in NYHA class 1. Group 2 (control group): 43 healthy children aged 99.8 ± 65.0 months.

Systolic RV function was assessed using longitudinal tricuspid annular systolic velocity by TDI (Sm) and RV isovolumic contraction time (IVCT). Diastolic RV function was assessed using RV isovolumic relaxation time (IVRT). Global RV function was assessed using the myocardial performance index (Tei index) calculated as follows: time interval between the onset and end of tricuspid annular velocities (a) minus the duration of systolic wave (b) was divided by b, i.e. (a − b)/b.

Results:

Table. Study results

	Post TF	*Control Group*	*P-value*
Sm (cm/s)	10.4 ± 1.9	15.1 ± 2.3	P <0.001
IVCT (msec)	103.7 ± 39.8	42.8 ± 26.6	P <0.001
IVRT (msec)	80.0 ± 27.8	30.5 ± 15.1	P <0.001
Tei index	0.80 ± 0.28	0.34 ± 0.06	P <0.001

There was a highly significant difference of all investigated RV functional parameters between the post TF and control group. Tricuspid annular systolic velocity was significantly lower; there was even a moderate RV volume overload in the post TF group. As both IVCT and IVRT were significantly prolonged, the calculated RV Tei index was extremely higher in the post TF Group, indicating a very altered global RV function. On the basis of ROC (receiver operating curve) analysis, the cut-off value for the Tei index for the Pos TF patients was 0.41 as to the sensitivity of 100% and specificity of 89%.

Conclusions: A significant impairment of RV systolic, diastolic and global functions were found in children operated on for tetralogy of Fallot (NIHA I functional group). The RV TDI-Tei index was found to be a very sensitive parameter for early recognition of global RV dysfunction.

OP-25 P-51

A pilot study on the effects of Carvediol on right ventricular remodelling and exercise tolerance in patients with systemic right ventricle

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Background: Patients with atrial repair for transposition of the great arteries and patients with congenitally corrected transposition have a right ventricle (RV) in the systemic position and they may develop RV dysfunction and exercise intolerance with advancing age. No data is available on the effect of carvedilol in patients with a dysfunctional systemic RV.
Methods: We studied with cardiovascular magnetic resonance (CMR), cardiopulmonary exercise testing, and standard 12-leads electrocardiogram, 8 adults (median age 26 years, range 18–31) with chronic stable heart failure and systemic RV dysfunction (6 patients with atrial repair and 2 patients with congenitally corrected transposition). Assessment was done before and after 12 months of carvedilol administration. The initial dose was 3.125 mg twice daily, and the target dose was 25 mg twice a day.
Results: Carvedilol administration was safe and the target dose was achieved in 5/8 (62%) patients. Right ventricular end-diastolic (119 ± 31 vs. $112 \pm 28\,ml/m^2$, $p = 0.01$) and end-systolic volumes decreased (79 ± 17 vs. $65 \pm 14\,ml/m^2$, $p = 0.006$), and RV ejection fraction improved (34 ± 6 vs. $42 \pm 7\,\%$, $p = 0.004$). Left ventricular ejection fraction increased (44 ± 8 vs. $49 \pm 9\%$, $p = 0.01$), suggesting a positive biventricular remodelling. Peak oxygen uptake did not change with carvedilol (26.8 ± 5.3 vs. $27.3 \pm 5.7\,ml O_2/Kg/min$, $= 0.58$), whereas exercise duration increased (13.4 ± 2.6 vs. 17.3 ± 3.1 min, $p = 0.008$).
Conclusions: In this small cohort, carvedilol administration was safe and it was associated with positive RV remodelling as well as improved exercise duration.

OP-26 P-49

Cardiac function analysis with MRI: assessment of measurement accuracy in a prospective multicenter study

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Objectives: MR volumetry is considered the gold standard to assess cardiac function but interobserver variability remains a practical limitation. We sought to assess the impact of semi-automatic postprocessing software and consensus criteria for image reading to reduce interobserver and interexamination variability (IOV, IEV).
Methods: We designed a prospective multicenter study involving patients with Tetralogy of Fallot (TOF, n = 30) and controls (n = 10). All subjects were studied at 1.5 T scanners (Philips) using well defined imaging protocols acquiring: (1) LV and RV enddiastolic/systolic volumes (EDV, ESV) with multislice SSFP in a short-axis and transversal plane; (2) muscle mass of interventricular septum, LV and RV freewall. MRI data was analyzed by customized software using semi-automated border detection by threshold adjustments. Images were acquired at three centers. Data were blinded and analysed by three experienced independent observers before and after informed consent about standardized guidelines for image analysis.
Results: Interexamination variability was low (<4%) in all studies and there was no systematic measurement error for either operator ($p > 0.62$). However, without informed consent there was significant IOV with a maximum of 18% for RV-EDV measured in short-axis plane at TOF and a minimum of 6% for LV-ESV in controls. After informed consent variability decreased significantly ($p < 0.01$) to 6% and 4%, respectively. Bland-Altman test showed agreement between measurements acquired in short-axis and transversal planes, however, IOV was significantly higher ($8 \pm 7\%$, $p < 0.05$) in short-axis when compared to analysis done in transversal planes.
Conclusions: Even in the presence of high-quality SSFP source images for MR volumetry, semi-automatical postprocessing software and consensus guidelines and training for image analysis were essential to reduce IOV from 18% to less than 6% to enable safe quantification of cardiac function in patient follow-up and clinical research. Transverse-plane volumetry is less prone to IOV than short-axis volumetry.

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OP-27 P-151

Transcatheter treatment for superior vena cava obstruction

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Introduction: SVC obstruction occurs as a complication of cardiac surgery, chronic SVC instrumentation or extrinsic compression. We assessed the safety, efficacy and freedom from re-intervention in patients who have undergone transcatheter treatment for SVC obstruction.
Methods: We retrospectively reviewed medical records and catheterisations in patients with SVC obstruction and congenital heart disease with biventricular circulation, and in patients with no cardiac disease who had developed SVC obstruction for other reasons.
Results: 56 patients with variable underlying anatomy underwent 75 transcatheter interventions for SVC obstruction (Mustard/Senning n = 22, ECMO n = 10, chronic lines n = 7, baffle repair of PAPVR n = 6, cardiac transplant n = 5, Warden operation n = 3, miscellaneous n = 2, tumor n = 1). The patients' median age and weight was 5.4 years (range 1 month–42 years) and 15.9 kg (range 3–114 kg), respectively. Forty-two patients were symptomatic, of which 23 had symptoms directly related to SVC obstruction, ie: SVC syndrome or effusions. The remainder had other symptoms (ie: syncope, heart failure) that prompted imaging and intervention (MRI in 2, other catheterization procedure in 8, recannalisation for transvenous pacing in 9). Thirty patients had stent implantation, (preceded by SVC predilation in 40%), and 26 patients had balloon dilation only. The mean SVC gradient decreased from 10.7 ± 6 mmHg to 2.5 ± 2.3 mmHg ($p < 0.0001$); high SVC

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pressure decreased from 18 ± 5.7 mmHg to 11.4 ± 4.6 mmHg ($p < 0.0001$). SVC diameter increased from 3.4 ± 2.8 mm to 9.1 ± 3.7 mm ($p < 0.0001$). Four patients, each with balloon dilation alone, developed an SVC tear; one of these patients required surgical repair. Stent malposition occurred in 2 patients and each was repositioned successfully. Re-interventions were performed in 12 patients (21%), due to SVC syndrome (n = 4), during other catheterisation procedures (n = 6) and after incidental detection of recurrent significant gradient by echocardiography (n = 2). Of these patients, three of 4 who initially underwent balloon dilation alone had a stent placed, and five of 8 with an existing stent had developed intimal hyperplasia.
Conclusions: SVC obstruction can be successfully relieved by balloon dilation or stent implantation in patients with a variety of underlying conditions. Complications occur in approximately 10% of patients but can often be dealt with in the catheterization laboratory.

OP-28 P-124
A cost and efficacy comparison of three methods for device closure of fenestrated Fontan

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Introduction: Following a fenestrated Fontan procedure many patients require device closure to restore normal saturation. This study evaluated the comparative cost and outcomes of three methods for fenestration closure (Flipper detachable coil (Cook®),Amplatzer®, double Ivalon umbrella (Cardia®).
Methods: Review of the patient database from the two participating institutions between 1996–2005.
Results: 58 patients age 3.6–13.9 (median 5.5) yrs, weight 6.1–74 (median 12.5) kg underwent fenestration closure (coil N = 32, Amplatzer® N = 12, Cardia® N = 14). All patients had test occlusion prior to device closure to ensure suitable hemodynamics. Pre-device Fontan pressure = 10.6 (7–15) mmHg; post = 12.8 (7–16) mmHg. Aortic saturation pre = 81% (71–92); post = 95% (91–98); $p > 0.05$ between devices. The 32 coil patients required a total of 51 coils (13 single, 16 double, 2 three coils). Delivery system: Coils 4 F; Amplatzer® 6 F (4 or 5 mm device); Cardia® 9–10 F (15 mm device). Repositioning was required in 67% of patients prior to coil release with no embolic events. No implant complications occurred with any device. Three Cardia® devices, after extruding both umbrellas, were retrieved and satisfactorily redelivered. Residual flow immediately post-implant: Coils 12/32; Amplatz 0/12; Cardia® 7/14. Follow-up (colour Doppler): Coils 2/32 (4 months–10 years); Amplatzer® 0/12 (1–7 year); Cardia® 0/14 (3 month–4 year). Cost per implant Canadian $ (EUR): Coils (device + delivery system $285/coil + $320 = $605 (432 EUR) + additional coil; Amplatzer® $6,480 (4,627 EUR); Cardia® $4,200 (2,999 EUR).
Conclusion: All three methods provide effective and long-term closure for fenestrated Fontan with no observed late complications during the study period. Each device has its own specific advantages and disadvantages. Coil occlusion has the least cost, smallest delivery system using standard coil occlusion techniques, although it requires the creation of a specific "clipped tube fenestration". The Amplatzer® has a similar F-size delivery system, simple placement technique and provides the highest immediate closure rate; although it has the highest device profile and cost. Cardia® requires the largest F-size delivery system, with a more intricate loading mechanism, similar deployment technique and final occlusion rates, with the advantage of a lower metal content (than Amplatzer®), and has the lowest device profile within the Fontan circuit.

OP-29 P-147
Stent implantation in restrictive atrial communications in patients with hypoplastic left heart syndrome

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Background: The presence of an appropriate interatrial shunt in patients with hypoplastic left heart syndrome (HLH-S) is essential. Without sufficient interatrial communication hypoxemia caused by pulmonary hypertension will araise. Ballon atrial septostomy (BAS) may not be sufficient in all cases.
Procedure: By femoral venous access interatrial stents were implanted in 5 patients with HLH-S, who were treated before by bilateral pulmonary banding and duct stenting. Using a 6 F long sheath exact stent placement was guided by hand-injections of contrast medium. Patient A, B received a 17 mm Jo-Stent (Jo-med) hand crimped on a 10 × 20 mm Balloon catheter, patient C, D ,E received premounted 10 × 19 mm Genesis-Stents (Cordis/Johnson & Johnson). Stent expansion was performed to create a diabolo shaped form which fitted in the septum.
Results: In consideration of the clinical condition, hypoxemia, and particularly the echocardiographic data, interventions were successfully performed as an elective or rescue procedure. In 4 of 5 patients left atrial pressure (LAP) was measured 22 ± 4.9 mmHg before intervention.

	Age	Mean LAP (b. s.) mmHg	Transcutaneous SaO_2 (%) (b.s.)	Transcutaneous SaO_2(%) (a.s.)
A	2 Mo	21	84 + O_2	92
B	3 Mo	29	67	77
C	2 Mo	25	72	84
D	2 Wk	not done	34	90
E	6 Wk	18	65	95

b.s. = before stent implantation; a.s. = after stent implantation

After stent placement patients clinical condition improved. O_2-saturation increased from 64% ± 18% to 88% ± 7%, $p < 0.05$). Considering the potential risk of stent dislocation LAP was not measured invasively after stent implantation.

Duct stenting, bilateral pulmonary banding as well as interatrial stent placement worked as a brigde to combined Norwood stage I and stage II operation in 4 patients and to heart transplant in one patient. The mean follow up until the following operation was 2.5 months. During this time the patients were not treated with any anticoagulants. There were no thrombotic events or other early or late stent related complications. Surgical removal of the stents was uneventful.
Conclusion: Stent implantation in patients with HLH-S is safe, effective and allows in selected cases a creation of an appropriate communication which prevents pulmonary hypertension and clinical deterioration when BAS alone is not sufficient.

OP-30 P-144
The Amplatzer membranous septal occluder in isolated and complex VSD's

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Introduction: The Amplatzer membranous septal occluder (AMVSO) is designed for transcatheter closure of perimembranous ventricular

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defects (VSDs). The most actual complication reported, has been complete heart block. We present our results in isolated and complex VSDs.
Patients: From 9/03–01/06 transcatheter closure of membranous VSDs was attempted in 42 pts (24♀ and 18♂) with significant left to right shunt (n = 18) or aortic-/tricuspid regurgitation induced by the VSD (n = 24). The age was 6 dd–26 yy (m = 9.3 yy), weight 3.3–79.1 kg (m = 28 kg). Diagnoses were isolated VSD (n = 37), VSD/TGA (n = 4), residual VSD after surgical VSD closure (n = 1).
Methods: Implantation was performed transvenously in 38 pts or intraoperatively in 4 newborns with d-TGA during arterial switch operation. In this patient population we did only use the AMVSO, the size ranged from 4–12 mm with a measured size of the defect of 3.5–11 mm (m = 5.7 mm). The distance of the defect to the aortic valve ranged from 1.7–12 mm.
Results: Device implantation was successful in 39/42 pts and complete closure was achieved in 36 pts; trivial residual shunt was detected in 3 pts. Device instability, intraoperative dislocation and impossibility to place the long sheath occurred in unsuccessful procedures. Preexisting tricuspid/aortic regurgitation improved in most of the patients. In 3 pts mild aortic-/tricuspid regurgitation was seen after device placement. Right bundle branch block occurred in 2 pts, transient left bundle branch block in 1 patient. Complete heart blocks did not occur.
Conclusion: The AMVSO is effective in isolated and complex membranous VSDs implanted percutaneously or as a hybrid approach during complex heart surgery. Preexisting aortic-/tricuspid regurgitation improve mostly after device placement.

OP-31 P-135
Novel technique of bifurcation stenting in pulmonary artery stenosis
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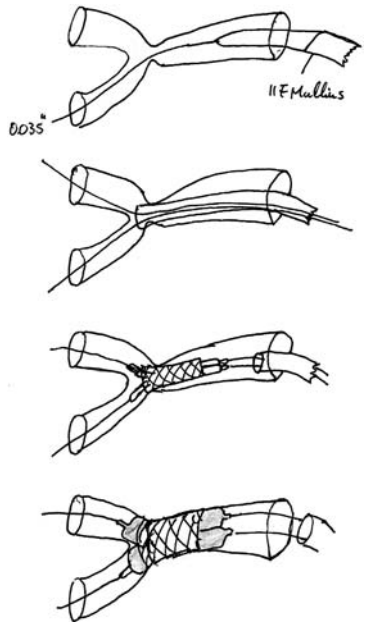
Background: Stenting of central or peripheral pulmonary artery bifurcation stenosis remains challenging. The choices are:

a) To use one stent to treat the proximal lesion or
b) To use 2 stents simultaneously to overlap into the proximal stenosis or
c) To treat the larger vessel and attempt side branch stenting.

The outcome is frequently limited with potential complications including jailing of side branches and vessel occlusions. We propose a novel technique of pulmonary artery bifurcation stenting and present our initial experience.
Technique: The lesion is assessed using appropriate biplane angiographic projections. An 11–16 Fr Mullins sheath is introduced over a guide-wire placed in the larger distal vessel beyond the proximal stenosis. A further guide-wire is placed through the Mullins sheath into the smaller branch vessel. Two balloon valvuloplasty catheters are selected according to the size of the branch vessels. The selected stent is mounted onto the 2 valvuloplasty balloons placed in parallel. When fully crimped this unit is introduced over the appropriate guide-wires into the Mullins sheath and advanced to the bifurcation. The Mullins sheath is retracted and the 2 valvuloplasty catheters are advanced in tandem into the side branches, taking care not to dislodge the mounted stent. Once a good position is achieved the 2 valvuloplasty catheters are inflated simultaneously. Repeat simultaneous inflations are carried out to achieve appropriate distal flaring of the stent.

This technique was utilised in 3 patients with very satisfactory results and no complications. In 1 case with severe recurrent bifurcation stenosis after a 23 mm Shellhigh conduit, it was not possible to introduce the stent mounted on a tandem 18 mm and 12 mm balloon through a 16 Fr Mullins sheath. Thus the stent was mounted on a tandem 18 Fr balloon and a 5 Fr catheter to protect the right pulmonary artery and avoid jailing.
Conclusion: Stent implantation into proximal bifurcation stenosis can be achieved utilizing this novel technique with good immediate results and reduced risk of jailing of side branches.



OP-32 P-131
Re-intervention after primary treatment of coarctation of aorta – balloon angioplasty versus surgery
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Aims: To assess the efficacy of each primary intervention (balloon angioplasty [BA] versus surgery [Sx]) in the treatment of coarctation of aorta in infants and children.
Methods: Retrospective review of patient records with the diagnostic code of coarctation of aorta, born between January 1990 and October 2005, and had surgical or catheter intervention were included in the study. Patients with hypoplasia of the aortic arch or interruption were excluded.
Results: We found 541 patients with diagnosis of coarctation of aorta which needed intervention over 15.8 years. 144 (26.6%) patients were initially treated by coarctation balloon angioplasty (15 neonates), whilst 397 (73.4%) patients had surgery (276 neonates). Of these, 108 patients required re-intervention for re-coarctation (42/144 in BA group [29.1%] and 66/397 in Sx group [16.6%], $p = 0.002$). Outside the neonatal age group, there was significantly older age at first intervention for the BA group, irrespective of need for re-intervention ($p < 0.0001$ for both re-intervention and non-re-intervention groups). The age at re-intervention was however significantly younger for the surgical group ($p = 0.0001$). However, interval between re-intervention was not statistically different for BA and SX group ($p = 0.44$).

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	Catheter balloon angioplasty (BA)			Surgery		
	No re-intervention	With re-intervention		No re-intervention	With re-intervention	
Age at initial treatment	>30 days	>30 days				
Number of patients	94	35		331	66	
Age 1st intervention						
• Median (days)	351	194		15	10	
• Range (days)	39-5348	37-1438		0-5302	1-754	
Age at re-intervention						
• Median (days)	NA	522		NA	119	
• Range (days)		47-3110			28-5465	
Interval between initial and 1st re-do						
• Median (days)		160			101	
• Range (days)		6-2605			17-5404	
Re-intervention by type		BA	Sx		BA	Sx
• 1st re-do	NA	12/35	23/35	NA	65/66	1/66
• 2nd re-do	NA	3/7	4/7	NA	7/8	1/8
• 3rd re-do	NA	0	0	NA	0	0

Conclusion: Coarctation of aorta can be successfully treated by balloon angioplasty and surgery. Irrespective of the age group, balloon angioplasty had significantly increased re-intervention rates [7/8 neonatal (47%), 35/129 beyond neonatal age group (27%)]. Patients treated with initial balloon angioplasty were significantly older, for both the reintervention and non-reintervention groups ($p < 0.0001$ for both). There is a significantly earlier re-intervention age for patients who were initially treated by surgery, but no significant difference in interval between re-intervention.

OP-33 P-137

Stentimplantation for treatment of recurrent and native coarctation in children <20 kg body weight

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Introduction: Balloon angioplasty for coarctation of the aorta may be unsuccessful due to elastic recoil or unfavorable anatomy, such as tubular narrowing or hypoplasia of the isthmus. Intermediate follow-up in patients with stents at the site of coarctation shows excellent gradient relief with minimal complications. We report of our experience with stentimplantation for treatment of recurrent and native aortic coarctation in children <20 kg body weight.
Patients: Between March 2003 and January 2006 we treated 38 patients with coarctation of the aorta with stents. 16 of these weighed <20 kg. 7/16 patients were s/p Norwood operation (average weight 6.3 kg (4.4–9.8)), 2/16 had native coarctation, 7/16 had recurrent coarctation post surgery (average weight 14.4 kg (5.5–19.8)). 8 patients underwent balloon dilatation before stent implantation.
Results: Under curative indication we used Palmaz Genesis XD Stents in 12 patients (max. stent diameter of 20 mm), under palliative indication we used 2 PG 124P (max. stent diameter 10 mm) and 2 JoStents 6–12 mm (max. stent diameter 14 mm). In 6/7 Norwood patients the implantation was performed antegradely from the femoral vein. In all patients the therapy was effective. Immediately after stent implantation the peak systolic gradient fell from 33 mmHg (16–70) to 3 mmHg (0–10). There were no complications. No aneurysms, stent dislocations or severe bleedings were observed. One patient had a weak peripheral pulse secondary to arterial access. Heparin therapy led to complete resolution. In 3/16 patients growth related redilatation of the stent was already performed.
Conclusion: Implantation of stents at the site of recurrent and native coarctation of the aorta shows excellent gradient relief. It is also a safe and effective therapy in smaller patients.

OP-34 P-132

Ductus stenting in duct-dependent systemic blood flow: self-expanding or balloon expandable stents?

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Introduction: The technique of duct stenting in newborns with duct-dependent systemic blood flow (DA-SBF) has dramatically changed over the last decade. Particularly, since percutaneous duct stenting combined with bilateral pulmonary artery banding (PAB) is established as a new strategy in newborns with hypoplastic left heart (HLH) lesions. Since one year percutaneous duct stenting is performed with self-expanding nitinol-stents (SENS), and balloon-expanding stents (BES).
Methods: In 2005, 18 newborns (body weight 1.7–3.9 kg) with DA-SBF were treated by duct-stenting in addition to PAB, but two. 6 newborns had a classical HLH-S (syndrome), 5 HLH-C (complex), and 7 other complex lesions with DA-SBF. 16 SENS (width 7–9 mm, length 12–20 mm) were implanted in 14 newborns (5F sheaths). Four newborns received 6 BES (Herkulink and Genesis). The detailed technique of duct-stenting was reported previously.
Results: Stent placement was feasible in all, with no death related to stenting. One pt. died despite successful stent placement. He didn't recover from cardiogenic shock. Four pts. needed a second stent because of stent slipping or uncovered duct tissue. An univentricular approach with aortic arch reconstruction (AOAR) together with bidirectional Glenn (BDG) was successfully performed in 4 pts. at an age of 3 to 6 months, in two such approaches are provided. Three pts. received heart transplantation, one is still waiting. Three pts. got biventricular repair, in 4 corrective repairs are foreseen.
Conclusion: Based on our experience of duct-stenting SENS should be preferentially used in unrestricted ducts, BES in stenotic ductus. Surgical removal seems to be easier in SENS.

OP-35 P-129

Hybrid catheter interventional and surgical therapy in patients with congenital heart disease: single-center experience of 3 years including 48 patients

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Introduction: For relief of congenital heart defects, sophisticated interventional and surgical therapeutic approaches are available. It is hypothesized, that the combination of both strategies in a so called "hybrid" approach, improves the outcome. We report the results of our combined surgical-interventional approach to a variety of congenital heart defects.
Methods: The data base was searched for patients with combined surgical-interventional approaches conducted during 03-2003 and 12-2005. Hybrid procedures were divided in preparative, intraoperative and early-postoperative (within the first postoperative days) approaches for residual defects. Late interventions for residues like fontan fenestrations, paravalvular leaks or residual septal defects were not categorized as "hybrid" and were therefore not included into this study.
Results: Using the above strategy, we identified 48 patients with hybrid approaches. Preparative procedures were performed in 29 patients and included 14 patients weighing <3 kg and 9 patients

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<2 kg. Preparative procedures included relief of RVOT obstruction in tetralogy of fallot with hypoplastic pulmonary arteries (n = 13), ductal stenting (n = 6), balloon valvuloplasty of critical aortic stenosis (n = 8), balloon angioplasty in critical coarctation (n = 1) and stent implantation into an obstructed right ventricular outflow tract in an 8 years old patient with tetralogy of fallot with mapcas in preparation for surgical correction. 11 patients were treated with intraoperative stent implantation into obstructed pulmonary arteries during the bilateral Glenn operation (n = 6), RV-PA conduit exchange (n = 4) or aortic valve replacement in a patient s/p alloprosthetic pulmonary valve replacement and no transcatheter access to the pulmonary arteries. In a patient (body weight 2.4 kg) with truncus arteriosus and long-segment interruption of the aorta and severe hypoplasia of the ascending aorta the ductus was stented in combination with bilateral banding. All intraoperative procedures were performed without angiography or fluoroscopy. Early postoperative catheter interventions were performed in 8 patients: closure of a residual large ventricular septal defect (n = 1), stent implantation into pulmonary arterial (sub-) occlusion (n = 3), angioplasty of aorto-pulmonary shunts (n = 2), Sano-conduits (n = 1) and angioplasty of RVOT obstruction early post arterial switch (n = 1).
Conclusion: The combination of surgical and catheter-interventional procedures leads to improved outcome in selected patients with congenital heart defects.

OP-36 P-126
Multi-detector computed tomography as an adjunct to the management of in-stent stenoses in small children with congenital heart disease
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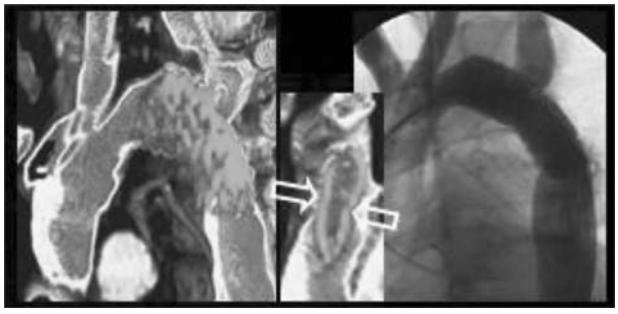
Introduction: Our purpose was to investigate the diagnostic reliability of multi-detector computed tomography (MDCT) in assessing in-stent stenoses compared to digital angiography (DA) in small children due the little knowledge about the feasibility of MDCT to assess stents placed to treat children with congenital heart disease (CHD).
Methods and Results: Fifty-four patients with transcatheter placed stents and suspected of having potential stent associated hemodynamic problems on the basis of echocardiographic findings could not be adequately imaged and underwent therefore MDCT exams. Twenty-five (out of 54) children with a median age of 3¼ [range: ½ to 12] years with 45 transcatheter placed stents (median diameter of 7.2 [3.4–16.3] mm; pulmonary arteries (n = 37), aorta (3, see Fig.), PDA (2), SVC (3)) underwent both MDCT and DA for diagnostic and studies due to suspected hemodynamic problems. Of 133 stent segments, 124 (93%) could be evaluated with both MDCT and DA by assessing stent and minimal luminal diameters. The interobserver variability of MDCT was low (mean difference: 0.4, SD 0.9 mm; correlation r = 0.97; $P < .0001$). The grade of stenosis on MDCT correlated high with DA (r = 0.90, $P < .0001$; mean error 2.8 ± 10.3%). The sensitivity of MDCT for ⩾20% stenoses (40/115 (35%) segments) was 98% respectively (no specificity available due to absent negative results), which was better than for all grade of stenoses (sensitivity 58%, specificity 97%). There were no significant diagnostic limitations for MDCT on the basis of the stent diameter employed. All stent associated complications (fracture (4), vascular narrowing (11)) were diagnosed by MDCT. Furthermore, MDCT lined out clearly one inward collapse of lumen of a aortic stent (see Fig.) which was difficult to appreciate at DA.
Conclusion: MDCT is a feasible method for assessing stent associated complications in the treatment of CHD. Cardiac surgeons and interventional cardiologists might rely on this imaging modality to plan specific intervention more precisely and to assess the results upon follow up.



P-1
Nonfluoroscopic mapping reduces radiation exposure in children and adolescents during RF ablation of supraventricular arrhythmias
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Background: Radiofrequency (RF) catheter ablation has become the treatment of choice for supraventricular arrhythmias also in pediatric patients. Minimizing the amount of irradiation is in the interest of young, growing patients as well as EP laboratory personnel. The pediatric RF ablation registry data show a decrease in mean fluoroscopy time from 51 (1991–1995) to 40 minutes (1996–2000), and to 35 minutes in 1999–2003. The utilization of new nonfluoroscopic mapping systems can further decrease fluoroscopy times. We evaluated the impact of the NavX® system on fluoroscopy times in pediatric RF procedures for common supraventricular tachycardias (SVT).
Methods: The procedure data were collected retrospectively from patient charts. NavX® mapping system was used in common SVT ablation procedures in 23 consecutive patients. Fluoroscopy time with and without NavX® was compared in 23 age and size matched patient pairs with equal tachycardia mechanisms and success rates. A two-tailed paired t-test was used in the analysis, $p < 0.05$ was considered significant.
Results: Using NavX®, the mean fluoroscopy time decreased from 26.5 minutes to 15.9 minutes ($p < 0.003$). The procedure time (2.96 h with fluoro only vs. 3.13 h, p = 0.50) or RF lesion count (1.5 with fluoro only vs. 2.0, p = 0.14) did not differ between the two groups. The tachycardia mechanism was AV-nodal reentry (AVNRT) in 9 and accessory pathway (WPW or concealed) in 14 patient pairs. The mean AVNRT fluoroscopy time was 6.3 minutes (3–11) with NavX® and 15.0 min (5–24) without (p = 0.032). Mean accessory pathway fluoroscopy time was 22.0 min (9–34) with NavX® and 33.9 min (11–81) without (p = 0.034). The overall mean fluoroscopy time in our laboratory in 2000–2003 was 33.8 min and showed a slightly decreasing tendency.
Conclusions: Utilization of the NavX® system reduces fluoroscopy time in pediatric RF ablations for common SVT. In developing children and adolescents, the use of a nonfluoroscopic mapping system can be recommended also in standard RF procedures, not only in complicated arrhythmias.

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P-2

Spontaneous ventricular tachycardia in a pediatric population

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Little is known on the epidemiology of spontaneously occurring episodes of ventricular tachycardia (VT) in a general pediatric population. This study aimed at defining the incidence, risk factors and outcome associated with VT in a pediatric population.

Methods: Tertiary referral centre serving a populatin of 1.4 million of which 252,000 are younger than 16 years. In that defined area there are no other pediatric cardiology services available. A 10 year period ending in June 2005 was studied. Based on a chart search, all in-house and outpatients <16 years of age and diagnosed as having VT were evaluated. Only patients with spontaneously occurring VT were studied, excluding patients in the immediate (<30 days) post cardiac-surgery period, those taking drugs with a known potential of proarrhythmia and VT occurring during other medical interventions.

Results: In the period studied, 26 children were observed with documented VT (accounting for 1 in 5880 patients seen in our institution diagnosed as having VT). There was a wide spectrum of underlying disease with idiopathic VT in a structurally normal heart being the most frequent clinical entity (n = 10). Cardiomyopathy was present in 5 patients, idioventricular rhythm was seen in 3 neonates, 3 children had previously operated heart defects, 2 had long QT syndrome and one each a cardiac tumor, WPW syndrome and coronary artery disease in hypercholesterolemia. 5 children died suddenly of VT (the one with coronary artery disease, one with long QT and 3 with cardiomyopathies). Symptoms associated with VT were observed in only 8 patients (30%). 13 of the patients (50%) were managed exclusively on an outpatient basis. Treatment consisted of antiarrhythmic drugs in 7 patients, ablation in 2 and causative therapy of the underlying disease in 6 patients. 13 children were managed without specific therapy. At last follow-up, 17 of 21 survivors were untreated and asymptomatic, 4 still had some form of treatment.

Conclusion: Spontaneous VT in children is rare and outcome is highly dependent on the underlying pathological substrate.

P-3

Age and sex-related changes in QTc in children with long QT syndrome (LQTS) 1 and 2

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Objective: Evaluation of age and sex-related changes in QTc in children with LQTS.

Methods: QTc was measured (lead II) in all available ECGs of 47 children with genotypic LQTS1 and 2 followed-up at the Academic Medical Centre, Amsterdam.

Group 1: 20 children with LQTS1. Group 2: 27 children with LQTS2.

Results: 260 ECGs were analysed representing 180 follow-up years.

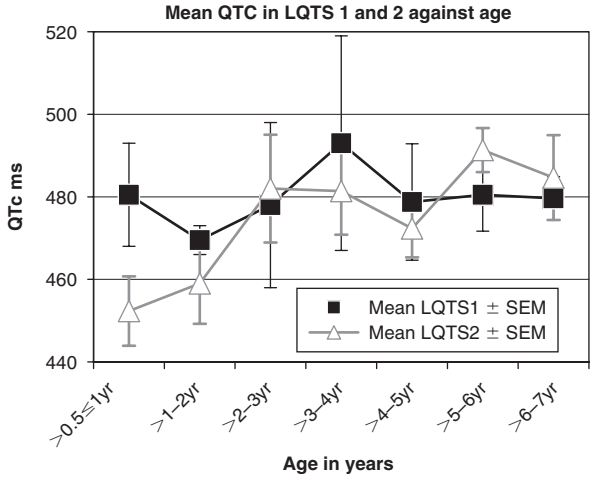
	Mean F/U/pt (yrs)	Male: Female	Median age at study (yrs)	Mean QTc (ms)	Mean QTc males (ms)	Mean QTc Females (ms)	
LQTS1	4.4 ± 3.7	8:12	10 (3–21)	489 ± 29	471 ± 22	499 ± 28	P < 0.0005
LQTS2	3.6 ± 2.8	17:10	11(0.3–17)	479 ± 30	485 ± 32	470 ± 26	P < 0.02
				P < 0.025	P < 0.02	P < 0.0005	

Mean QTc is significantly longer in LQTS1 than LQTS2.

Within the groups, mean QTc is significantly longer in LQTS1 females than in males. This relationship is reversed in LQTS2.

QTc is significantly longer in LQTS1 females than LQTS2 females and significantly longer in LQTS2 males than LQTS1 males.

All 3 LQTS1 patients with documented ventricular tachycardias were females.



QTc increases significantly in LQTS2 between 1–6 years ($0.025 < p < 0.05$).

QTc in LQTS1 remains relatively constant during the first 12 years.

Conclusions: There is a sex-related difference in QTc in the LQTS types studied.

An age-related increase in mean QTc in children with LQTS2 was seen. This may explain the different ages at first presentation of syncope in LQTS1 and 2, consistent with a strong relationship between the risk of torsades de pointes and degree of QTc prolongation. Drug therapy in LQTS2 patients can possibly be delayed until late in the first decade.

P-4

Does sex, age or gender influence the mechanism of supraventricular tachycardia in pediatric patients?

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Background: The affect of age and gender has been proven to have an influence on the mechanism of supraventricular tachycardia (SVT) in adults. The purpose of this study was to determine if age, sex, or race has an impact on the mechanism of SVT in pediatric patients.

Methods: A search of the Pediatric Electrophysiology Registry identified patients (pts) whom had undergone a radiofrequency catheter ablation from 89–04. Pts were grouped into three categories based upon age; <7, 7–<12, and >12–18. Statistical analysis was conducted using the Chi-Squared and Fischer's exact test with a $p < 0.05$ considered significant.

Results: There were 3,554 patients (<7 = 378, 7–<12 = 964, 12–18 = 2212) with 1948 males (M), 2,916 White, 266 Black,

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248 Hispanic, 63 Asian and 61 Others. For accessory pathway (AP) tachycardias, there were 2,417 pts (1404M) and for AV node reentry tachycardia (AVNRT) there were 1,137 pts (544M). Significant differences were found in sex distribution between AP (845M/571F) and AVNRT (353M/443F) in the >12–18 age group with females more likely to have AVNRT than AP ($p < 0.0001$). When all data was combined, females are still more likely to have AVNRT (593/1137) than AP (1013/2417) ($p < 0.0001$). Additional significant difference was identified when comparisons were made between Whites and Blacks for AP (White = 558, Black = 44) vs. AVNRT (White = 209, Black = 32) in age group 7–<12, with Whites being more likely to have AP ($p < 0.025$).

Conclusions: The mechanism of SVT in pediatric patients is affected by sex, age, as well as race. Whites were more likely to have an AP then Blacks.

SVT type by race and sex

	AP (M)	AP (F)	AVNRT (M)	AVNRT (F)
White	1138	823	442	513
Black	102	73	47	44
Hispanic	115	78	37	18
Asian	25	20	8	10
Other	24	19	10	8
Total	1404	1013	544	593

P-5
Use of novel approach of radio frequency assisted perforation for placement of pacemaker leads in patients with congenital heart disease

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Introduction: We describe a novel use of radio frequency assisted perforation to allow placement of pacemaker leads after intra-atrial repair operations for transposition of the great arteries.

Patient 1: A 14 year old boy after a Senning operation had progressive right ventricular dysfunction and intermittent complete heart block. The ventricular function continued to deteriorate despite dual chamber pacemaker and medical therapy. He also had coarctation, treated by stent angioplasty during which he was resuscitated from a ventricular fibrillation arrest. Hence, we planned for biventricular pacemaker and defibrillator strategy.

Using subclavian approach the ventricular lead was extracted and defibrillator lead placed into left ventricular apex. The septum between pulmonary and systemic venous atrium was perforated with the Nykanen RF perforation system. The perforation wire was exchanged for a Cordis SV5 0.018′ wire and the sheath was exchanged for a short transeptal sheath and dilator to enlarge the atrial perforation. Select Secure sheath and dilator were then passed to the pulmonary venous atrium and a pacing lead was positioned in the RV (Fig. 1a). He has good improvement in interventricular conduction delay (from 90 to 50 milliseconds) and exercise tolerance in follow up.

Patient 2: A 28 year old man after a Mustard operation presented with presyncope secondary to sinus arrest. Angiography showed moderate length pathway occlusion with a small baffle leak (Fig. 1b). A Nykanen radiofrequency catheter system was used to perforate the atretic segment via jugular approach. A guide wire circuit was made by snaring a wire in the systemic venous pathway from a femoral vein approach. A 38 mm long CP covered stent was deployed from the femoral venous access. This restored patency of the superior caval pathway and closed the baffle leak. A dual chamber permanent pacemaker system was the inserted from a subclavian approach (Fig. 1c). He has had good relief of his symptoms following pacemaker implantation.

Conclusions: These are the first described cases of radiofrequency assisted perforation procedures to allow pacemaker lead implantation after intra-atrial repair operations for transposition. This technique is an alternative to epicardial leads or the use of transeptal needle procedures.

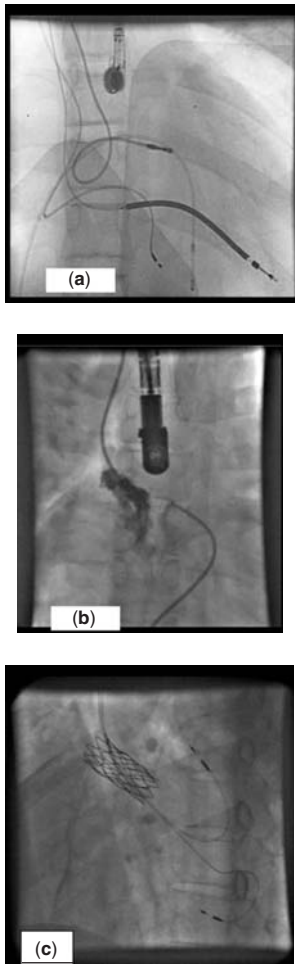

Figure 1.

P-6
Early and midterm results of ICD (Implantable Cardioverter Defibrillator) implantation in patients with repaired Tetralogy of Fallot and pulmonary stenosis

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Introduction: Repair of Tetralogy of Fallot is associated with an increased risk of ventricular tachyarrhythmia and sudden death in young adulthood. This study aims to determine the rate of appropriate and inappropriate discharges and the effectiveness of ICD therapy in this population of patients.

Materials and Methods: This is a retrospective review of patients with repaired Tetralogy of Fallot (n = 18) and pulmonary stenosis (n = 2) with implantable cardioverter defibrillators managed at

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our centre. Patients were identified from our outpatient database and data collected on the incidence of appropriate and inappropriate therapies and the success rate of ICD therapy.
Results: Of the 20 patients, 18 had previous repair of Tetralogy of Fallot and 2 had pulmonary valvotomy and infundibular resection for pulmonary stenosis between 1969 and 1989. 70% (n = 14) of these patients required reoperation with 10 patients having pulmonary valve replacements (PVR).

The median age at implantation was 22 years (16.4–43 years). Early post procedural complications included atrial lead displacement (n = 1) and pneumothorax requiring drainage (n = 1).

During a median follow up of 1.6 years (0.03–4.5 years) several episodes of inappropriate therapies were noted in 6 patients (30%) especially early after implantation (33 episodes of inappropriate anti-tachycardia pacing (ATP) and 19 episodes of inappropriate cardioversion). Appropriate ATP was instituted in 4 patients (25%) with successful termination of all 20 episodes (100 % success rate) of ventricular tachycardia. 1 patient required cardioversion with successful termination of VF. 1 patient (5%) with troublesome tachyarrhythmia died suddenly of unknown cause, 10 months after AICD implantation having had no detections or therapies on his device.

Late complications of ICD implantation included lead failure in 1 patient requiring replacement 3.3 yrs after implantation and generator replacement in a patient due to an advisory issued by the manufacturer regarding the risk of sudden battery depletion.

Conclusions: In our study we found a rate of 0.6 appropriate and 1.4 inappropriate therapies (0.9 episodes of inappropriate ATP and 0.5 episodes of inappropriate cardioversion) per patient-year of follow up following ICD implantation. Anti-tachycardia pacing was very successful in terminating tachyarrhythmia in our population with 100% success in terminating ventricular tachycardia.

(For P-7, please see OP-17)

(For P-8, please see OP-18)

P-9
Prevalence of WPW syndrome in a multicentre study of paediatric Ebstein patients in the Netherlands

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Introduction: Ebstein's anomaly (EA) is the most commonly occurring congenital abnormality associated with Wolff-Parkinson-White (WPW) syndrome. WPW has been reported in 14–20% of all Ebstein patients. Objective of this study was to assess the prevalence of WPW in the population of paediatric patients with EA in the Netherlands.
Methods: A multicenter retrospective study was performed, combining medical records of paediatric patients diagnosed with EA between 1980 and 2005 from the six paediatric heart centers in the Netherlands. Using this database, patients who also had a diagnosis of WPW syndrome were selected and reviewed.
Results: Nine (9.7%, 95%CI: 4.5–17.6%) of the 93 Ebstein's patients in the database were diagnosed with WPW syndrome. The diagnosis WPW was made at the same time as the diagnosis EA in all 9 patients. Six (67%) of the 9 patients were diagnosed at birth, the other three at 42, 74 and 141 months. Eight (89%) patients were alive at the end of the study. The one death was not due to an arrhythmia.

In six (67%) patients a supraventricular tachycardia was registered; 2 of them also experienced an episode of ventricular tachycardia. Due to the ventricular tachycardia, one patient had a cardiac arrest at birth but was successfully resuscitated. Later, this patient underwent two catheter ablations unsuccessfully, and remained on anti-arrhythmic medication (calcium antagonist) at the last follow-up. The second patient received a beta-blocker therapy.

A Holter registration had been made of 3 patients with WPW syndrome. The first had an AV-nodal re-entrant tachycardia with episodes of sinus rhythm, the second a sinus rhythm with pre-excitation and ventricular extra systole, and the third had multiple supraventricular and ventricular extra systole, but no pre-excitation could be found. The first two underwent catheter ablation, successfully. The latter remained symptomatic and received a beta-blocker at the last follow-up.

The patients not receiving any medication (n = 6), reported to be asymptomatic at their last visit.
Conclusions: The prevalence of WPW syndrome in our patients with Ebstein's anomaly during childhood and adolescence was lower than reported in the literature. Life threatening rhythm disturbances are not frequent early in life.

P-10
Cryomapping offers advantages for ablation near the atrioventricular junction in pediatric patients

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Objectives: Ablation in young children using radiofrequency (RF) energy can be challenging when the arrhythmia substrate is located in close proximity to the atrioventricular (AV) node due to a significant risk for inadvertent AV block. Cryomapping can theoretically decrease the occurrence of iatrogenic AV block. Aim of our study was to determine the effect of cryomapping during ablation of arrhythmia substrates located in the triangle of Koch in a pediatric patient group.
Methods: This was a single center, prospective, non-randomized study. During the study period (August, 2004–November, 2005) a total of 59 pediatric patients (age range: 7.5 to 17.7 years) were screened for supraventricular arrhythmias. After a diagnosis of AVNRT or AVRT with an anteroseptal accessory pathway (AP) was made, cryomapping was performed cooling the catheter tip temperature to a max. of −40°C. Mapping was performed either during ongoing tachycardia or in sinus rhythm. Ablation was performed only if cryomapping terminated the tachycardia or blocked the AP conduction without prolongation of the AV conduction.
Results: In 14/59 pts (24%) RF ablation was abandoned because of arrhythmia substrate in close proximity to AV node (11 anteroseptal AP, 3 AVNRT's). Cryoablation was successful in all 3 pts with AVNRT. Elimination of AP conduction without AV block was observed in 5/11 pts and cryoablation was performed successfully. In 6/11 pts high degree AV block developed simultaneously with elimination of AP conduction with during cryomapping in patients with anteroseptal AP's. Cryoablation was not attempted in these cases. This resulted in an acute success rate of 57% for cryoablation (8/14 pts). No permanent complications related to the use of cryoenergy were observed. Only one (12%)

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patient experienced recurrence of AP conduction during a 12 months follow up period.
Conclusions: 1. Cryoablation is a safe alternative to RF ablation in pediatric patients, when the arrhythmia substrate is located in close proximity of the AV node. 2. Cryomapping improves success rate on an intention to treat basis. 3. Cryomapping identifies high risk patients for inadvertent AV block, but results in a considerable decrease of the overall ablation success rate.

P-11
Evolution of paced QRS and QTc intervals in children with epicardial pacing leads
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Background: Some children with permanent pacing develop left ventricular dysfunction due to dysynchronized ventricular activation. There is no information whether a dysproportionate increase of paced QRS intervals over time may be a cause of ventricular dysfunction. Therefore, we evaluated the evolution of paced QRS and QTc intervals in children with permanent ventricular pacing.
Patients and Methods: ECG and pacemaker data of 45 consecutive children who received a permanent pacing system at a median age of 6.3 years (range 0.1–18.3) for bradycardia pacing were analyzed. Bipolar steroid eluting epicardial leads (Medtronic 10366 and 4968) were implanted and connected to various pulse generators. Median follow up time was 3.1 years (range 1.1–11.5). Standard deviation scores (SDS) for paced QRS and QTc intervals were calculated from Garson's standard-ECG norm-values. As a measure for QRS and QTc interval changes, regression slope coefficients were calculated for each patient's course. Data are presented as median (range) or [interquartile range]. For statistical analysis, groups with right (group 1, n = 18) and left (group 2, n = 27) ventricular pacing were compared using Mann-Whitney-test.
Results: At first and last follow-up, median SDS for paced QRS intervals in group 1 were 4.68 (2.45–7.90) and 4.62 (3.77–6.90), in group 2 they were 4.18 (2.00–7.00) and 4.50 (2.95–7.27). Slopes for QRS intervals in group 1 and 2 were 0.014 [−0.31–0.10] and −0.013 [−0.26–0.35], respectively. For QTc intervals the slopes were 0.015 [−0.10–0.12] and 0.043 [−0.13–0.16], respectively. No statistical significance was seen between the two groups. Slopes for QRS intervals in group 1 showed a tendency to be skewed towards negative values.
Conclusion: Substantially prolonged paced QRS intervals were observed in right and left ventricular epicardial pacing. However, epicardial pacing of the right or left ventricle did not cause dysproportionate QRS and QTc interval increases over time.

P-12
Percutaneous coronary injection of bone marrow cells in small experimental animals: small is not too small
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[1]Necker Enfants Malades, Paris; [2]Salvatore Maugeri Foundation, Gussago; [3]University of Ferrara, Italy

Background: Intracoronary infusion of bone marrow cells (BMc) is thought to induce cardiac regeneration in ischemic heart disease and dilated cardiomyopathy. Although small experimental animals are largely utilized in this field of research, the size of peripheral vessels and coronary arteries has prevented intracoronary BMc injection so far. We developed a new method to inject BMc into coronary arteries of small experimental animals.
Methods: Transient atrioventricular block (AVB) was obtained in 25 rats and 25 hamsters through intracarotid injection of adenosine 5'-triphosphate (ATP). Contrast echocardiography was obtained. BMc (0.2–0.5 ml) were collected through femoral puncture, stained with PKH26 and injected into the carotid artery. Ten animals were immediately sacrificed and 10 followed during one month.
Results: Induction of transient AVB was possible in all animals by injecting 20 to 30 mg of ATP. Animals recovered a basal cardiac activity spontaneously or by dopamine injection. Flash injection of contrast medium through the carotid artery induced opacification of aortic root, coronary arteries and ventricles but not of ventricular cavity. BMc injection was possible in all cases, over few seconds. No immediate or late ECG changes were observed. Hystological examination showed, immediately after injection, the presence of BMc into small coronary arteries and, after 1 month, the absence of infarction.
Conclusions: ATP-induced AVB block allows percutaneous intracoronary injection of BMc in small experimental animals with no immediate or late mortality and morbidity. This method offers new perspectives to investigation of BMc coronary infusion in heart diseases.

P-13
Hypoxia increases PPARα and PGC-1β expression in neonatal rat cardiomyocytes
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Introduction: Szintigraphic studies in children with congenital cyanotic heart disease have shown a reduction in cardiac fatty acid metabolism. The aim of the present study was to investigate if hypoxia influences not only the activity but also the mRNA expression of enzymes involved in cardiac fatty acid oxidation. In addition to the enzymes themselves, a number of transcription factors involved in the transcription control of these enzymes were looked at.
Methods: Neonatal rat cardiomyocytes were cultured under 5 or 10% SaO_2 in the ambient air. Effective hypoxia was confirmed by measuring medium pO_2 by blood gas analysis, increases in hypoxia inducible factor (HIF) dependent genes (VEGF, GLUT1) by PCR, as well as phosphorylation of AMP activated proteinkinase (AMPK) by Western blot analysis. mRNA expression of the transcription factors of the peroxisome proliferator activated receptor (PPAR) family (α, β, γ), their Co-factors (PGC-1α and β, RXRα and γ) and that of PPAR regulated enzymes of fatty cardiac acid oxidation was determined by real time PCR.
Results: Hypoxia resulted in a statistically significant increase in mRNA expression of PPARα (+190% and 130% (at 5 or 10% O_2 respectively)) and PGC-1β (each + 60%). PPARβ and γ, PGC-1α, as well as RXRα and γ mRNA expression did not change under hypoxia, neither did that of the PPAR target genes involved in fatty acid oxidation (LCAD, ACOX, MCD, CPT-1).
Conclusion: 24 h hypoxia results in an increase in expression of the transcription factor PPARα and its co-factor PGC-1β. This change has no effect upon the expression of PPAR target genes. Studies of other groups have shown that an increased PPARα expression and an activation of the AMPK result in an increase in fatty acid oxidation. However, in short term hypoxia additional mechanisms appear to be important. If the reduction in fatty acid oxidation as seen in long term hypoxia (e.g. in cyanotic heart disease) is mediated by changes in PPAR regulation remains to be investigated.

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P-14

Total antioxidant capacity in children undergoing cardiac surgery

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Background: In this prospective study we tested the hypothesis that cardiopulmonary bypass (CPB) in children influence plasma anti-oxidant capacity, and that preoperative cyanosis aggravates oxidative stress by reducing anti-oxidant capacity. Finally, we examined the clinical relevance of postoperatively changed total antioxidant capacity.
Patients and Method: 26 infants/children with cyanotic or non-cyanotic cardiac defect were investigated. All patients were operated with support of hypothermic CPB receiving mechanical ventilation with pure oxygen. Plasma total antioxidant capacity was assessed by Trolox Equivalents Anti-Oxidant Capacity (TEAC) prae-, intra- and postoperatively. TEAC was analyzed with respect to operation variables (duration of CPB, aortic clamping, cardio-circulatory arrest) and clinical outcome variables (hemodynamic variables including dosage of inotropic drugs, markers of renal- and liver function, oxygenation index, and duration of stay on the intensive care unit).
Results: TEAC significantly decreased after induction of anesthesia with hyperoxygenation ($p < 0.001$). During CPB, there was a continuous increase up to the end of CPB. Postoperatively there were no differences. Preoperative TEAC values were similar in cyanotic and non-cyanotic patients and were significantly higher in cyanotic patients than in the others 4/24 hours postoperatively, respectively.

TEAC values 4/24 hours postoperatively were correlated with duration of aortic clamping (respectively). Postoperative TEAC values were negatively correlated with mean arterial blood pressure and positively correlated with dosages of inotropic drugs, liver enzyme levels, duration of mechanical ventilation and duration of the stay on the intensive care unit.
Conclusion:

1. General anesthesia with pure oxygen ventilation and not extra-corporeal circulation with ischemia-reperfusion is related to a significant decrease of the total antioxidant capacity in children undergoing cardiac surgery.
2. This decrease is transient, lasting up to the first postoperative day
3. Preoperative hypoxemia does not influence peri-operative total antioxidant capacity as assessed by plasma levels of TEAC.
4. Patients needing longer operative support and having the worse clinical outcome show the higher TEAC values after the operation, suggesting release of antioxidants from damaged tissue as a response to higher stress rather than higher regeneration of the antioxidant capacity.

P-15

Use of bivalirudin as an anticoagulant for pediatric cardiac interventional procedures

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Introduction: The practice of pediatric catheter intervention has evolved to include implantation of many different vascular and cardiac devices, but no data exists as to the optimal periprocedural anticoagulation regimen for these procedures. Heparin, the traditional agent, has a variable response in children, especially neonates, based on its reliance on ATIII as cofactor and variable protein binding. Neonates have very low levels of ATIII but normal functioning thrombin. Bivalirudin, a thrombin specific inhibitor, has considerable data as the periprocedural anticoagulant for adult percutaneous coronary interventions (PCI) and offers several potential benefits over heparin for children undergoing catheter based procedures, e.g., linear dose response, no cofactor requirement, short half life, low volume of distribution.
Methods: Pediatric patients received bivalirudin IV (0.25–0.75 mg/kg bolus) followed by an infusion to maintain an ACT >200 sec during intervention. The weight based dose of 0.75 mg/kg bolus and 1.75 mg/kg/hr infusion was used for patients weighing >40 kg, lower doses for patients those <40 Kg.
Results: In 2004–5, thirty pediatric patients, ranging in age from 5 days to 25 years (Mean ± SD = 7 ± 7.64) with a BSA of 0.19 to 2.1 m^2 (Mean ± SD = 1 ± 0.55) received bivalirudin as the anticoagulant during various procedures, including insertion of ASD or PDA closure devices, stent insertion, and valvuloplasty. Average ACT 15 minutes post bivalirudin bolus was 259 sec. All patients completed the procedure without visible thrombus formation, though one patient required post procedure anticoagulation for an absent distal pulse, which resolved within thirty minutes. There were no other clinical events (bleeding or thrombus formation) and all patients were discharged without complication.
Conclusions: Bivalirudin has intrinsic properties that would make it a favourable agent for pediatric antithrombin therapy. Although further work is required to establish the optimal dose for children of all weights, this initial experience indicates the feasibility of using bivalirudin as a procedural anticoagulant for pediatric catheter based interventions.

P-16

Impaired endothelial function is an early marker of arterial dysfunction in young pre-pubescent obese children

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Overweight and obesity in very young children have become major public health issues. No information about vascular function is available in pre-pubescent obese.

The aim of the present study was to investigate non invasively the effects of early obesity on mechanical properties of an elastic artery, the common carotid artery (CCA) and the endothelial function of the brachial artery using a high-resolution ultrasonography.

35 pre-pubescent obese subjects, 8.9 ± 0.3 years old, weight 51 ± 13 Kg, height 139 ± 9 cm, body mass index (BMI) 26 ± 5 Kg/m^2 and lean subjects n = 13, 8.8 ± 1.5 years old, weight 31 ± 5 Kg, height 138 ± 9 cm, BMI 16 ± 2 Kg/m^2 were examined. The local CCA pulse pressure (LPP) was determined non-invasively with tonometry of applanation. Using LPP, intima media thickness (IMT) measurement, systolic and diastolic diameter of CCA, mechanical parameters were determined: cross sectional compliance (CSC), cross sectional distensibility (CSD) and incremental elastic modulus (Einc). The vascular endothelial function was evaluated through measuring brachial artery vasodilatation, after transient ischemia (flow-mediated dilatation, FMD%). Endothelial independent vasodilation was also measured after administration of sublingual glyceryl trinitrate (glyceryl trinitrate mediated dilatation, GTNMD).

No differences were evident between obese and lean subjects in LPP (37 ± 13 vs 34 ± 11 mmHg), IMT (0.49 ± 0.03 vs 0.47 ± 0.02 mm), CSC (0.19 ± 0.12 vs 0.22 ± 0.09 $mm^2 \cdot mm\,Hg^{-1}$), CSD (0.9 ± 0.6 vs 1.2 ± 0.6 $mm\,Hg^{-1} \cdot 10^{-2}$) and Einc (14 ± 6 vs 11 ± 6 $mm\,Hg \cdot 10^2$). Flow-mediated dilation (FMD) impaired in

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obese subjects relative to lean control subjects (5.7 ± 2.7% vs 8 ± 2%, p = 0.002); GTN dilation was not altered in the obese group (21 ± 7 vs 25 ± 7%).

In pre-pubescent young obese children, the mechanical properties and the remodelling of the CCA are not yet altered. However the endothelial dysfunction, a well-demonstrated cardiovascular risk factor, appears to be the initial altered index of arterial function.

P-17

The serum- and glucocorticoid-inducible kinase Sgk-1 is involved in pulmonary vascular remodeling: role in the regulation of tissue factor by thrombin

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A prothrombotic state is frequently associated with pulmonary hypertension and can promote vascular remodeling. The stress-responsive serum- and glucocorticoid-inducible kinase Sgk-1 has been associated with fibrosis and hypertension, but the function of this kinase in vascular remodeling and thrombosis has not been elucidated. We previously showed that thrombin enhances tissue factor (TF) expression as well as Sgk-1 expression in pulmonary artery smooth muscle cells (PASMC). In this study we investigated the molecular mechanisms linking Sgk-1 with thrombin action and tissue factor expression in PASMC.

Thrombin enhanced TF promoter activity, expression and procoagulant activity in PASMC. Thrombin also activated the transcription factor nuclear factor kappa-B (NFkappaB) whereas inhibition of NFkappaB diminished TF promoter activity and expression in response to thrombin. Furthermore, Sgk-1 increased NFkappaB activity as well as TF expression and procoagulant activity whereas activation of NFkappaB or TF upregulation by thrombin were diminished by inactive Sgk-1. Similarly, activation of NFkappaB or expression of TF by thrombin were not detectable in fibroblasts from mice deficient in sgk-1 (sgk1-/-). Furthermore, treatment with dexamethasone, known to enhance Sgk-1 and TF, failed to induce TF expression and activity in lung tissue from sgk1-/- mice, confirming the role of Sgk-1 in TF regulation in vivo. Finally, Sgk-1 and TF were specifically detected in lung tissue in the media of small pulmonary vessels with signs of vascular remodeling.

These data show that Sgk-1 increased TF expression and activity by activating the transcription factor NFkappaB. Since enhanced procoagulant activity can promote pulmonary vascular remodeling, and Sgk-1 and TF were present in the media of remodeled pulmonary vessels, this pathway may play a critical role in promoting vascular remodeling in pulmonary hypertension.

P-18

Nitric oxide modulates Rac activation by thrombin: role in the redox-sensitive regulation of angiogenesis

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Endothelial dysfunction characterized by enhanced levels of reactive oxygen species (ROS) and reduced nitric oxide (NO) bioavailability as well as increased thrombogenicity have been associated with pulmonary hypertension and appear to play a central role in promoting vascular remodeling processes including angiogenesis. However, the underlying signaling pathways are poorly understood.

The GTPase Rac has been shown to be required for ROS production by NADPH oxidases and to enhance NO levels. We therefore investigated whether Rac is involved in the regulation of angiogenesis by the coagulation factor thrombin in human microvascular endothelial cells.

Stimulation with thrombin stimulated ROS generation, activated the endothelial NO synthase (eNOS) and enhanced NO production and increased Rac activity and protein expression.

In the presence of antioxidants or inactive Rac, thrombin-induced ROS production was diminished whereas active Rac enhanced ROS levels. On the other hand, exposure to N-nitro-L-arginine (L-NNA), which inhibits eNOS activity, increased ROS levels and Rac activity whereas antioxidants prevented these effects. Furthermore, thrombin and active Rac as well as L-NNA increased endothelial proliferation and stimulated in vitro angiogenesis. In contrast, antioxidants and inactive Rac diminished proliferation and angiogenesis in response to thrombin.

Taken together these data show that Rac and ROS are required for activation of endothelial proliferation and angiogenesis by thrombin. Thus, activated Rac may play an important role in linking oxidative stress with a procoagulant state and angiogenesis in pulmonary hypertension. Concomitant activation of NO production by thrombin antagonizes activation of Rac and subsequently endothelial proliferation and angiogenesis, thus limiting the deleterious consequences of endothelial dysfunction on pulmonary vascular remodeling processes.

P-19

PAK upregulates plasminogen activator inhibitor-1 and promotes proliferation of pulmonary artery smooth muscle cells: the role of calcium and reactive oxygen species

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A prothrombotic state due to enhanced levels of thrombin and plasminogen activator inhibitor-1 (PAI-1), an inhibitor of fibrinolysis, is frequently found in pulmonary hypertension and has been associated with remodeling processes in the vascular wall. However, the molecular mechanisms connecting these factors with proliferation of pulmonary artery smooth muscle cells (PASMC), the main cell type involved in pulmonary vascular remodeling, are not resolved. We previously showed that the p21-activated kinase (PAK) can be activated by thrombin in PASMC. In this study we investigated the role of PAK in the regulation of PAI-1 and proliferation of PASMC in response to thrombin.

Thrombin rapidly activated PAK, and this response was inhibited by the calcium chelator BAPTA-AM. Subsequently, thrombin and activated PAK were able to enhance the levels of reactive oxygen species (ROS) in a calcium-dependent manner. Furthermore, thrombin and activated PAK stimulated PAI-1 promoter activity as well as mRNA and protein expression whereas pretreatment with BAPTA-AM and antioxidants prevented these responses. Concomitantly, inactive PAK inhibited upregulation of PAI-1 by thrombin. Furthermore, thrombin and activated PAK increased expression

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and activity of the hypoxia-inducible transcription factor HIF-1alpha, and this response was inhibited by antioxidants, BAPTA-AM, or inactive PAK whereas depletion of HIF-1alpha by siRNA prevented upregulation of PAI-1 by thrombin. Finally, thrombin and activated PAK stimulated proliferation of PASMC whereas antioxidants, BAPTA-AM or an inhibitory antibody against PAI-1 inhibited proliferation of PASMC in response to thrombin.

Taken together, these data show that thrombin stimulates PASMC proliferation via calcium-dependent activation of PAK, subsequent ROS formation and induction of PAI-1 via activation of the transcription factor HIF-1alpha. Thus, this mechanism may play an important role in promoting vascular remodeling in pulmonary hypertension.

(For P-20, please see OP-1)

P-21
Multi-lineage stem cells derived from the thymus gland of children during cardiac surgery as a source for regenerative medicine

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Introduction: Stem cell transplantation and tissue engineering are attractive future strategies for functional myocardial repair. Multi-lineage stem cells (MLSC) are well qualified for this purpose and have mainly been isolated from the bone-marrow so far. In the present study we aimed to isolate MLSC from the thymus gland of infants undergoing cardiac defect repair, where partial thymectomy represent a standard procedure for better cardiac access.
Methods: After sternotomy, the thymus gland of 10 neonates/children undergoing cardiac surgery was removed. Tissue was minced, digested and cultured. Multi-lineage potential was tested every three culture passages using differentiation cultures towards the osteogenic, chondrogenic and adipogenic lineage. Surface antigen expression was investigated in detail by FACS-analysis and results were compared with MLSC from the bone-marrow.
Results: While the majority of isolated cells were non-adherent lymphocytes, we observed two types of adherent cells in primary culture. The epithelial cell colonies were refractory to passage. The remaining cells showed almost unlimited proliferation. Throughout the proliferation phase of more than 40 doublings a multi-lineage potential was re-evaluated. Full differentiation potential is maintained during this proliferation phase as confirmed by differentiation cultures and FACS-analysis. The thymic MLSC showed similar characteristics as compared to bone-marrow derived MLSC.
Conclusions: The human thymus gland is an alternative source for isolation of MLSC in neonates and young children undergoing cardiac surgery since the thymus is routinely resected during pediatric cardiac surgery. Children who might undergo further cardiac surgeries might profit from the use of thymic MLSC for tissue engineering.

P-22
PTPN11 mutations in Turkish children with noonan syndrome

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Introduction: Noonan syndrome (NS) is a developmental disorder characterized by distinctive facial findings, short stature and cardiac defects. A gene responsible for the disorder (PTPN11) was recently identified, and mutations in this gene explain 30–50% of the cases clinically diagnosed possible with NS. The aims of this study are to determine the PTPN 11 mutations in Turkish children with NS and to assess genotype-phenotype correlations in a cohort of clinically well-characterized pediatric patients with NS.
Methods: Fourteen unrelated patients (4 months – 15 years old, 9 male – 5 female) with the clinical diagnosis of NS ascertained according to standardized inclusion criteria were enrolled. Genomic DNA from each participant was isolated from blood lymphocytes using standard procedures. Mutational analysis was performed by direct sequencing of the exons 2–4, 7–9, 12–13 of PTPN11 where mutations have previously been reported in this gene followed by PCR-RFLP screening for certain mutations.
Results: Heterozygous point mutations in the PTPN11 gene were thus far detected in 5 patients. The p. Asn308Asp (in exon 8) mutation was detected in 3 children. The p. Tyr63Cys and p. Ala72Ser were detected in single patients. All three mutations have been previously described. Clinical features of mutation-positive and negative patients were shown in Table 1.

Table 1. Clinical features of mutation-positive and negative patients.

Clinical feature	Mutation (+), n:5	Mutation (−), n:9
Pulmonary stenosis	5	6
Hypertrophic cardiomyopathy	–	2
ASD/VSD	3	6
Typical facial features	5	9
Short stature	5	5
Speech delay/learning disability	2	2
Thorax deformity	2	6
Cryptorchidism	4	–

Conclusions: This is the first study reporting PTPN11 mutation screening in Turkish patients. Although the number of included patients is not sufficient to make a firm conclusion and the mutation screening is not completely finished yet, some clinical features appear to be more or less frequent in the mutation positive group. Assessing the genotype–phenotype correlation in different ethnic groups will be helpful to clarify the clinical effects of individual mutations.

P-23
Lack of evidence of association between congenital heart disease and methylenetetrahydrofolate reductase gene C677T and A1298C polymorphism

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Introduction: Hyperhomocysteinemia is frequently associated with neural tube defects and congenital heart defects (CHD). A common missense mutation in the MTHFR gene is C to T substitution at position 677 changing valine to alanine, resulting in higher plasma levels of homocysteine. The second common mutation in MTHFR gene is the A1298C polymorphism, reduces the enzyme activity but the homozygosity for this mutation is not associated with elevated tHcy levels. The aim of this study is to investigate

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association with MTHFR gene C677T and A1298C polymorphism and also compound heterozygosity with CHD.
Methods: This study included 142 children with isolated congenital cardiac defects consecutively seen by our Pediatric Cardiology Department. Patients with any other congenital defect or systemic disease were excluded from the study. Seventy-one age- and sex- matched children without any known systemic disease or congenital defect served as a control group. For analysis of the MTHFR gene polymorphisms, DNA samples extracted by Oiogen kit from peripheral blood were amplified using polymerase chain reaction method. The C677T and A1298C polymorphisms were analyzed by Hinf I and Mbo II restriction enzyme digestion, respectively. Allelic distribution were analyzed with Chi-Square test.
Results: The overall genotype frequencies of the MTHFR C677T and A1298C polymorphism were not significantly different between the CHD patients and the healthy control ($p > 0.05$).
Conclusion: We did not find sufficient evidence for an association between MTHFR C677T genotype and congenital heart disease in our study group. Different results about this association may be due to some heterogeneity in the development of various subtypes of CHD or to population genetic structure.

P-24
The effect of nicotine on myocardium of newborn rats whose mothers received nicotine during gestation and lactation
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Objectives: The aim of this study was to evaluate the effects of nicotine on myocardium of newborn rats whose mothers received nicotine during gestation and lactation, by using biochemical and pathological parameters and to evaluate the role of antioxidant enzymes, free oxygen radicals, and lipid peroxidation products which have role in the etiopathogenesis of the nicotine induced injury.
Methods: The experiment was carried out on rats of Spraque-Dawley type. The rats were divided into 3 groups. Group I: Low dose nicotine (1 mg/kg/day, n = 10). Group II: high dose nicotine group (6 mg/kg/day, n = 10). Group III: Control group (n = 10). Nicotine was administered by subcutaneous way and control group received 0.2 cc subcutaneous saline. Four weeks later rats were mated. Nicotine was administered from the beginning to end of third week of lactation, (average 11 week, during pregnancy and lactation). Plasma malondialdehyde (MDA), nitric oxide (NO), glutation peroxidase (GSH-Px) and superoxide dismutase (SOD) levels were measured at the end of the study. After the sacrification, the levels of myocardial MDA, NO, GSH-Px and SOD were analyzed and myocardial tissue was examined histopathologically with haematoxylin – eosin stain.
Results: In group I and II the levels of plasma and tissue MDA and tissue NO were higher; whereas the levels of myocardial GSH-Px and SOD were lower than group III ($p < 0.05$). Heavy cardiomyopathy characterized by swelling in myocardial filaments, interstitial edema, disorganization and necrosis was determined in Group I and II.
Conclusions: Our results demonstrate the destructive effects of nicotine on myocardium: decrease in myocardial antioxidant enzymes activities and increase in free radicals and lipid peroxidation products which have important roles in the pathogenesis of nicotine – induced myocardial injury.

P-25
An experimental study on protective effects of L-triptophan on hypoxic heart
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Objectives: The aim of this study was to evaluate the protective effect of triptophan on experimentally produced hypoxic myocardial injury by biochemical and pathological parameters and the role of free oxygen radicals, lipid peroxidation products and antioxidant enzymes in hypoxic injury etiopathogenesis.
Methods: A total of 26 New Zealand rabbits were divided into three groups. The first group (n = 9), which only exposed to hypoxia, second group hypoxic and L-triptophan received group (n = 10), and third group was control group (n = 7). Before the hypoxic injury serum samples were taken for troponin-I, CKMB, LDH, GSH-Px, SOD, MDA and NO analysis. In group I and II hypoxic condition was obtained by a funnel which was drained by 5 lt/min 10% oxygen and nitrogen mixture for 10 minutes. After the hypoxic injury, rabbits in group II received L-triptophan (200 mg/kg/day) orally for 5 days. After the medication second samples for troponin I, CKMB, LDH, GSH-Px, SOD, MDA and NO were taken and rabbits were sacrificed and the myocardium samples taken and myocardial NO, MDA, SOD, GSH-Px enzyme activity levels were studied by histopathologically with hematoxylene and eosin staining.
Results: In group I severe cardiomyopathy was demonstrated histopathologically, and GSH-Px and SOD activity decrease, and significant troponin-I, CKMB, LDH elevations were detected in this group. In group II: tissue GSH-Px and plasma SOD activity increased significantly and tissue MDA levels decreased. Mild to moderate cardiomyopathy was demonstrated histopathologically.
Conclusions: Our findings support that, there is a clear effect of free oxygen radicals and lipid peroxidation products on hypoxic cardiomyopathy, and L-triptophan supplementation has a strong protective effect on hypoxic heart by antioxidant activity.

P-26
Comparison of various brain protective strategies in the setting of deep hypothermic circulatory arrest
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Objectives: The metabolism of the brain, although reduced, is not suppressed by hypothermia at 18°C, a common target temperature in surgical practice. Various brain protective strategies, as antegrade selective cerebral perfusion (ASCP) and other adjuncts, are in clinical use, although poorly defined. We assessed and compared the efficacy and safety of these protective strategies.
Methods: 4 groups (1 to 4) of 6 mini piglets each were randomized to undergo DHCA, DHCA after premedication with GPIIb/IIIa-receptor-inhibitor, DHCA with low-flow-ASCP (10 ml/kg/min) or DHCA with high-flow-ASCP (30 ml/kg/min) respectively. The CPB protocol consisted of 40 min cooling, 60 min DHCA and 40 min rewarming followed by 30 min normothermic bypass. ASCP perfusion pressures of 50–60 mmHg were employed. Intravital microscopy of the pial vessels was assessed every 10 min. Histopathology and immunocytology were obtained at the completion of the experiment.

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Results: Indices of cerebral perfusion (functional capillary density, red blood cell velocity and tissue oxygenation) were significantly improved in group 2 and 4 compared to group 1 and 3 ($P < 0.01$). Biochemical alterations, followed by ultrastructural and then structural changes could likewise be shown in group 1 and 3. A significantly increased capillary permeability as well as an VEGFR2-upregulation in group 1, 3 and 4 compared to group 2 were measured ($P < 0.05$).
Conclusions: GPIIb/IIIa-receptor-inhibitor and high-flow-ASCP improved capillary perfusion. However, adequate clinical ASCP-flow-monitoring is lacking. Further, ASCP is somewhat surgically demanding.

P-27
S100B in brain cell culture under conditions of deep hypothermia and rewarming
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Background: Increased concentrations of the astrocytic protein S100B in serum and cerebrospinal fluid in association with hypothermic perfusion of cardiopulmonary bypass have been associated with a higher incidence of postoperative neurological dysfunction after corrective surgery of congenital heart disease. In vitro, S100B has beneficial effects in picomolar or nanomolar concentrations, while micromolar S100B levels lead to detrimental effects. However, the role of S100B in hypothermia-associated neuroregeneration and inflammation has not yet been investigated in vitro. Therefore, we analyzed the impact of nanomolar S100B administration on hypothermia-treated primary astrocytes, BV-2 microglial cells, primary neurons and organotypic brain slice cultures subjected to dynamic changes of hypothermic temperature.
Methods: Brain cells and organotypic brain slice cultures were pretreated with nanomolar S100B concentrations and incubated according to a hypothermia protocol mimicking temperature changes during cardiac surgery in children: deep hypothermia (2 h at 17°C, phase 1), slow rewarming (2 h up to 37°C, phase 2), normothermia (20 h at 37°C, phase 3).

In all cells, release of the proinflammatory cytokine IL-6 was measured. In brain slices, axonal outgrowth modulation was analyzed microscopically.
Results: Deep hypothermia significantly induced secretion of the pro-inflammatory cytokine IL-6 by astrocytes, microglial cells and neurons. Application of S100B reduced hypothermia-induced IL-6 release by microglial cells and neurons. In contrast, S100B synergistically increased hypothermia-induced IL-6 secretion by astrocytes. In brain slice culture, S100B significantly suppressed hypothermia-induced axonal outgrowth.
Conclusion: Deep hypothermia induced an inflammatory response in all brain cells studied. S100B has cell-specific anti-inflammatory effects under conditions of hypothermia and rewarming. The effect of S100B on neuronal cell activation and axonal outgrowth under these non-physiological conditions needs further investigation.

P-28
Fibronectin-binding protein A enhances endothelial procoagulant activity and monocyte interaction in the pathogenesis of S. aureus infections such as endocarditis
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Staphylococcus aureus is among the most important bacterial pathogens responsible for endocarditis in children and adults. Essential for the infection process is the high propensity of S. aureus to colonize endovascular tissues, allowing these pathogens to spread via the bloodstream to other tissues. Our recent studies emphasize the significant contribution of fibronectin binding proteins of S. aureus to endothelial cell adhesion in the pathogenesis of endovascular S. aureus infections and demonstrate their role in inducing a variety of proinflammatory endothelial responses resulting in leukocyte accumulation, cell damage and fibrin deposition.

In the present study we investigated the role of fibronectin binding protein A (FnBPA) concerning endothelial pro-coagulant activity via the extrinsic pathway and further examined whether monocyte interaction is mediated by FnBPA leading to enhanced procoagulant activity.

S. aureus FnBPA was constitutively expressed in the non-invasive organism Lactococcus lactis by means of gene transfer. L. lactis piL 253, carring the empty expression plasmid, showed virtually no adherence to cultured human venous endothelial cells and was therefore taken as a control.

Incubation of endothelial cells with the FnBPA-expressing L. lactis strain led to endothelial tissue factor (TF) m-RNA production and endothelial TF-antigen expression. The extrinsic coagulation pathway was activated which was investigated by increased FXa activity. Monocyte adherence was increased in L. lactis FnBPA infected endothelial cells with 70–80% of monocytes bound. Monocyte interaction led to increased TF-expression as well as enhanced pro-coagulant activity. Further investigations to analyse the role of single regions of the FnBPA molecule were done with the consecutive L. lactis strains. Incubation with strains containing the parts of the FnBPA molecule which mediate fibronectin binding led to a high endothelial adhesion rate and further more to activation of the endothelial cells.

We conclude that S. aureus FnBPA plays an important role in the TF-mediated endothelial pro-coagulant activity and mediates monocyte-endothelial interaction. Further investigations stated the importance of the fibronectin binding regions of the FnBPA molecule as pathways to evoke inflammation, tissue damage and fibrin deposition at the infected endovascular sites.

P-29
The protective effect of melatonin on nicotine induced myocardial injury in newborn rats
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Objectives: One of the most dangerous substances in tobacco is nicotine. By passing the placenta nicotine increases vascular resistance, fetal heart rate and decreases umbilical blood flow. The aim of this study was to evaluate the protective effects of melatonin; an antioxidant which used for maternal nicotine induced myocardial injury in newborn rats, by using biochemical and pathological parameters.
Methods: The Spraque-Dawley rats (n = 49) were divided into 5 groups. Group I: high dose nicotine (n = 10). Group II: high dose nicotine and melatonin group (n = 9), Group III: low dose

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nicotine (n = 10). Group IV: low dose nicotine and melatonin (n = 10). Group V: Control group (n = 10). Nicotine was administered by subcutaneous way, as 6 mg/kg/day to high dose group, and 1 mg/kg/day to low dose group, from the beginning to end of third week of lactation, (average 11 week, during pregnancy and lactation). Melatonin was given 10 mg/kg/day during gestation and lactation period, and control group received distilled water. Rats were mated at fourth week. Plasma malondialdehyde (MDA), nitric oxide (NO), glutation peroxidase (GSH-Px) and superoxide dismutase (SOD) levels were measured at the end of the study. After the sacrification, the levels of myocardial MDA, NO, GSH-Px and SOD were analyzed and myocardial tissue was examined histopathologically.

Results: In group I and III the levels of plasma and tissue MDA and tissue NO were higher; whereas the levels of myocardial GSH-Px and SOD were lower than group III ($p < 0.05$). Heavy cardiomyopathy characterized by swelling in myocardial filaments, interstitial edema, disorganization and necrosis was determined in Group I and III.

There was a significant decrease in the levels of myocardial MDA and NO, but a significant increase in the levels of myocardial GSH-Px and SOD in the melatonin administered groups (group II and IV). Histopathological signs were improved in group IV when compared with group III.

Conclusion: It was shown that a decrease in myocardial antioxidant enzymes and an increase in the free radicals and lipid peroxidation products have important roles in the pathogenesis of nicotine induced myocardial injury. Also melatonin can particularly protect the nicotine induced cardiac injury as an antioxidant.

(For P-30, please see OP-2)

P-31
A registry of familial cases of congenital heart defects to identify new predisposing genetic factors

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Introduction: About 15% of congenital heart defects (CHD) are syndromic or related to chromosomal anomaly. For the 85% remaining cases, it is commonly accepted that CHD results from the combination of 1 to 3 genetic factors with or without an adverse environmental factor. Currently, only in rare occasions one genetic factor is evidenced. In order to increase the number of genetic factors responsible for CHD, we decided to set up a registry of familial CHD including clinical information and blood sample of affecteds and their first degree relatives. It is hypothesised that familial cases correspond to stronger genetic factors than sporadic cases.

Methods: Twenty three paediatric centres participate to this registry. Familial cases of CHD are given the information about the registry. If they contemplate a participation, the pediatric cardiologist forward telephone number and a detailed description of CHD to the registry centre which will contact family members to set up a complete pedigree tree with surname, first name and medical history. Blood samples of affecteds and first degree relatives are shipped to the registry centre and DNA is extracted. Systematically, the DNA of 2 affecteds/family is screened for mutation in 3 genes (ZIC3, NKX2.5 and GATA4) by dHPLC and/or sequencing.

Results: So far, 443 individuals of 63 families were enrolled. DNA of 250 individuals including 154 affecteds were obtained. Eleven additional families were excluded because either they did not provide blood samples (5) or only one affected was still alive (6). Four families have 4 living affecteds, 3 have 5 and 1 have 6 affecteds. The other families have 2 or 3 affecteds. Ten families are consistent with autosomal recessive inheritance, 3 are apparently X-linked, the remaining are autosomal dominant with incomplete penetrance or undetermined. Mutation screening is underway and already two mutation were evidenced (one in ZIC3 and one in NKX2.5) in 2 different families.

Conclusions: This resource will be extended to 150 families. Families with no mutation will be analysed by linkage analysis and mutation screening in candidate genes. This process is expected to evidence new genetic factors and greatly improve genetic counselling.

P-32
Comparison of electrical velocimetry and Fick's principle for measuring cardiac output in children with congenital heart disease

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Introduction: Impedance cardiography (ICG) has been used extensively to estimate cardiac output (CO) from changes of thoracic electrical bioimpedance (TEB). However, studies comparing ICG with reference methods have questioned the reliability of this approach. Electrical velocimetry (EV) provides a new algorithm to calculate cardiac output (CO). The purpose of this study was to validate the non-invasive CO measurement using EV (EV-CO) with Fick's Principle.

Method: Standard ECG electrodes were used for non-invasive EV-CO. These were placed in children under 2 years of age on the forehead respectively in children over 2 years of age on the left neck and the left thorax. Twenty three consecutive children mean age 2.9 years (25 days to 15.9 years), mean weight 12.6 kg (3.0 kg to 60.9 kg) who underwent hemodynamic evaluation for their cardiac anomaly in the cath lab were studied. During data acquisition for invasive CO measurement simultaneously VO_2 was measured (Vmax Encore®) and EV-CO (AESCULON®, Osypka Medical GmbH, Berlin, Germany) recorded. EV-CO was calculated using the Bernstein-Osypka equation. Invasive CO was calculated using the Fick's Principle with measured VO_2.

Results: A significant high correlation was found between EV-CO and invasive Goldstandard with measured VO_2 (r^2 0.89). Data were related linearly. The slope of the line (0.91 ± 0.06) was not significantly different from unity. Bland-Altman analysis revealed a bias of -0.19 litre min^{-1} with narrow limits of agreement (-0.95 to 0.56 litre min^{-1}).

Conclusion: The agreement between EV-CO and Fick's Principle with measured VO_2 is clinically acceptable. Therefore the non-invasive EV-CO method, a new ICG algorithm, provides a reliable continuous beat-to-beat estimation of CO over an arbitrarily long period in children with congenital heart disease.

P-33
The modified geometry of aortic arch after switch operation is associated with an early reflection of pulse wave pressure

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Background: Switch operation for transposition of the great arteries (TGA) translates the ascending aorta from anterior to posterior position. This results into a reduction of aortic arch curve and could potentially influence the pulse wave reflection.
Methods: From 1 January, 2005 to 31 June, 2005, we performed systematic aortography in 15 TGA children, with a median age of 5.5 years (range 5–6) and a median weight of 20 kilograms (range 15–25). Ten age and weight matched children undergoing patent ductus arteriosus closure constituted the control group. Aortic arch angle was measured in lateral view in all patients. All patients underwent tonometry of applanation to record pressure waveforms from the radial artery. Data were processed to produce estimated aortic pressure waveform, determine central pulse pressure (CPP = systolic-diastolic pressure) and augmentation index (AI), an estimate of the pulse wave reflection.
Results: Aortic arch angle was significantly more pointed in TGA than in control patients (54 ± 7.5° vs 68 ± 5°, $p < 0.001$). No difference of CPP was observed between the two groups. AI was significantly higher in TGA compared with control subjects (17 ± 10 vs 11 ± 9, $p = 0.03$). However, AI was not correlated with angle, age or height.
Conclusions: The reduction of the aortic arch angle after switch repair of TGA is associated with an early pulse wave reflection independently of the angle diameter, without modification of CPP. Long term effects of this data should be determined.

(For P-34, please see OP-21)

P-35
Pulmonary artery index as predictor of azygoz vein dilatation after hemi-Fontan paliation in children with hypoplastic left heart syndrome

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Background: In children after modified I-stage (RV–PA conduit) and hemi-Fontan paliation large disproportions of pulmonary artery branches (PAB) with left PAB hypoplasia have been observed. This can diminish pulmonary blood flow by drainage part of blood via dilated azygoz vein (AV).
Aim: To assess predictive value of pulmonary artery index (PAI) for dilatation of AV in children with HLHS after two stage treatment.
Material: Thirty five (31 male, 4 female) consecutive patients with men age equal to 26 months (SD:8.1) who underwent hemodynamic study between June 2003–November 2005.
Methods: Based on angiocardiograms of all patients the diameters of PAB with PAI as well as presence of dilated AV were assessed. Depending on presence of dilated AV the patients were divided into two groups: (1) AV absent (n = 27) and (2) AV present (n = 8).
Results: PAI, SaO_2 mSVCp, mLAp were significantly different between the studied groups. Values of PAI and selected hemodynamic parameters are summarized in the table (data are presented as mean ± SD).

Group (n)	Age at H-F (mo)	Age at cath. (mo)	BSA (m^2)	PAI (mm^2/m^2)	SaO2 (%)	mSVCp (mm Hg)	mLAp (mm Hg)	Qp/Qs	RVEDp (mm Hg)
1 (27)	6.8 ± 1.3	27.1 ± 6.8	0.52	199.5 ± 83	82.7 ± 8	12.7 ± 2	8.0 ± 2.1	0.67 ± 0.2	9.0 ± 3.3
2 (8)	6.3 ± 1.8	22.6 ± 12	0.51	106 ± 60.0	73.1 ± 15	17.9 ± 8	12.3 ± 6	0.58 ± 0.3	12.2 ± 6
p	ns	ns	ns	0.0046	0.025	0.038	0.029	ns	ns

Cut off point of PAI defined by ROC Curve analysis (StatsDirect Software) was 170 mm^2/m^2. Incidence of dilated AV was significantly higher in patients with PAI < 170 mm^2/m^2 compared with patients with PAI > 170 mm^2/m^2 (Relative risk: 8.94, 95% CI: 1.7–52.6).
Conclusions: Hypoplasia of pulmonary artery branches with significant decreased PAI (<170 mm^2/m^2) is a risk factor of dilatation of azygoz vein with decrease of systemic arterial oxygenation. All these findings can negatively affect right ventricular function in HLHS patients after hemi-Fontan operation.

P-36
The acute effect of anthracyclines on cardiac function assessed with strain rate imaging

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Background: Chronic cardiotoxicity is the main limitation for the use of Anthracyclines (Ant) to treat malignancies. However, the acute impact of Ant on cardiac function has not been studied due to the lack of sensitive non-invasive techniques. Strain rate imaging is a new technique which offers a more sensitive approach to quantifying early abnormalities in systolic function.
Aim: To describe the acute effects of Ant on left ventricular systolic function as assessed with strain rate imaging.
Methods: We prospectively studied 10 children (age range: 3–16) scheduled to receive Ant for different malignancies. They all underwent a tissue Doppler imaging echocardiographic exam before the first dose and within one hour after the completion of each of the first three doses of Ant (dose range: 40–75 mg/m^2). Peak systolic myocardial strain rate and strain were estimated both in the radial, from the infero-lateral wall, and in the longitudinal direction from the left ventricular (LV) lateral wall and interventricular septum.
Results: There was a significant decrease in deformation after the first dose; further less pronounced deterioration occurred after consecutive doses. See table. An inverse moderately strong correlation between cumulative Ant dose and radial strain was noted (r: 0.5, $p < 0.01$).
Conclusions: Our study shows that even low doses of Ant acutely reduce LV deformation. Strain rate imaging could potentially be used to monitor cardiac function since it detects abnormalities at an early subclinical stage. The predictive value of these findings needs further validation.

Table.

	Radial Strain Rate (1/s)	Radial Strain (%)	Longitudinal Strain Rate	Longitudinal Strain
Baseline	5.4 ± 1.1	75.3 ± 15.6	−2.0 ± 0.4	−28.7 ± 7.4
1st Dose	4.5 ± 0.8*	57.2 ± 15.2*	−2.1 ± 0.5	−26.7 ± 8.4
2nd Dose	4.8 ± 1.2$^{\#}$	52.1 ± 17.2*§	−1.8 ± 0.4	−23.2 ± 6.4$^{\#}$
3rd Dose	4.4 ± 0.7*	50.4 ± 15.6*§	−1.9 ± 6.5	−22.5 ± 7.8*§

$^{\star}p < 0.01$ vs. Baseline; $^{\#}p < 0.05$ vs. Baseline; $^{\S}p < 0.05$ vs. 1st dose.

P-37
Cardiac MRI is a valid non-invasive alternative to cardiac catheterization in selected patients candidate to Fontan intervention

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Introduction: Cardiac catheterization (CC) is routinely indicated for the evaluation of patients candidate to Fontan intervention. Magnetic resonance imaging (MRI) has recently gained widespread acceptance in the evaluation of mediastinal vessels and cardiac anatomy and function.
Aim: to test the accuracy of MRI in pre-Fontan patients, to compare MRI with cardiac catheterization and finally to select patient which can avoid routine catheterization before Fontan.
Methods: From June 2002 to November 2005, 26 patients (aged 9.5 ± 10 years) candidate to Fontan were evaluated by echo, MRI and CC. Before CC patients were divided in two groups according on clinical, echo and MRI findings: Group I (15): patients predicted to be suitable for Fontan procedure without CC. Group II (11): patients with indication to CC for interventional procedures, pulmonary resistances assessment or to detect more diagnostic details. CC and MRI findings were compared and surgical findings represented the gold standard against which MRI and CC were tested.
Results: In Group I (15 pts) one pt. was successfully operated without previous CC. In the other fourteen CC didn't add any useful information. All of them, but three still in waiting list, have been operated and surgical findings were always concordant with MRI and CC issues.

In Group II (11pts) MRI and CC findings were concordant in seven pts, and in two of them an interventional procedure was performed. In two other pts MRI highlighted a pulmonary venous collector mild stenosis not confirmed at CC, but assessed and corrected by the surgeon. In the last two pts MRI missed an azygos-portal vein collateral and a coronary fistula, both diagnosed by CC. About surgical issue seven pts have been successfully operated, one is waiting for the intervention, two refused the surgical indication and the last patient has been excluded from the Fontan procedure because of ventricular dysfunction.
Conclusion: In patients candidate to Fontan, MRI, in conjunction with clinical and echocardiographic assessment, can be a valid non invasive alternative to CC, which should be performed only when MRI is not exhaustive or when interventional procedures are indicated.

P-38

Assessment of time interval between onset of ventricular inflow and onset of early diastolic velocity by tissue Doppler in normal infants

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Introduction: Pulsed Doppler ventricular inflow velocities and tissue Doppler imaging of the annulus have been used to assess ventricular function in both children and adults. Recently, time interval between onset of transmitral early inflow (E) and onset of early diastolic (Ea) velocity of the mitral annulus (TEa-E) has been used to identify diastolic dysfunction in adults, but to date, no studies have been done in children, especially in infants. The purpose of our study was to evaluate the normal values of mitral and tricuspid TEa-E in infants under 1 year of age, and to assess the influence of age, heart rate and cardiac growth on this index.
Methods: Twenty five healthy children (mean ± SD age, 5.5 ± 2.2 months) underwent echocardiography at a single center. Mitral and tricuspid inflow and tissue Doppler velocities were obtained from the leaflet tips and lateral site of the annulus, respectively, in the apical four-chamber view. The time intervals between the peak of R wave and onset of mitral and tricuspid E velocity (T-E) and between peak of R wave and onset of Ea (T-Ea) velocity were measured. The differences between these time intervals were calculated as TEa-E, which were compared with demographic and echocardiographic variables.
Results: Mean heart rate during study was 125 ± 72 beats per minute. Mitral TEa-E was shorter than tricuspid TEa-E (mean ± SD, 24.6 ± 10.7 msec versus 33.1 ± 12.8 msec, $p < 0.05$). In all subjects, both mitral and tricuspid T-Ea were significantly greater than T-E (tricuspid: $p < 0.01$, mitral: $p < 0.01$). Mitral TEa-E tended to decrease with age, but the results were not statistically significant. Both mitral and tricuspid TEa-E did not correlate with heart rate. When correlated with echocardiographic variables, mitral TEa-E tended to decrease as left ventricular end diastolic dimension increased ($p = 0.035$). Tricuspid TEa-E tended to increase with tricuspid E/A ratio ($p = 0.024$).
Conclusions: In normal infants, TEa-E values differ between both ventricles, which suggest differences in ventricular adaptation in the first year of life.

(For P-39, please see OP-20)

P-40

Impaired subendocardial contractile myofibre function in healthy aged humans, as detected with MRI

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Objective: The ratio of left ventricular (LV) torsion to endocardial circumferential shortening (torsion-to-shortening ratio; TSR) during systole reflects the transmural distribution of contractile myofibre function. With aging several structural and functional changes occur in the myocardium without obvious impairment of systolic LV function. We investigated whether the transmural distribution of systolic contractile myofibre function changes with age.
Methods: Torsion and endocardial circumferential shortening were derived from displacements in parallel LV short axis sections, as measured with magnetic resonance tissue tagging (MRT). TSR was quantified in healthy young (age 23.2 ± 2.6 yrs, mean ± SD, n = 15) and aged volunteers (age 68.8 ± 4.4 yrs, n = 16).
Results: TSR and its standard deviation were significantly elevated in the aged group (0.47 ± 0.12 aged, 0.34 ± 0.05 young; $p < 0.0005$). Systolic and diastolic blood pressures and the ratio of LV mass to end-diastolic volume were also significantly increased in the aged group, whereas no significant differences were found in LV mass or ejection fraction.
Conclusion: The elevated systolic TSR in aged healthy subjects suggests that aging is associated with a decrease of contractile myofibre function in the subendocardium relative to the subepicardium.

P-41

Echocardiography and tissue Doppler Imaging of patients with Pompe disease under Enzyme Replacement Therapy

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Glycogenosis II (Pompe disease), is an autosomal-recessively inherited deficiency of the glycolytic lysosomal enzyme acid

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alpha-glucosidase (GAA). The infantile form is characterized by generalized myopathy and cardiomyopathy, leading to death in infancy, mostly of cardiac failure. Over the past years, an Enzyme Replacement Therapy (ERT) has been developed. Patients receive two-weekly infusions of rhGAA. The treatment is experimental.

We report two infants (M.C. and L.B.) who came to our department with neonatal heart failure due to hypertrophic cardiomyopathy. Both were diagnosed with Glycogenosis II and were enrolled in an ERT study at the age of 2 (L.B.) and 4 (M.C.) months respectively.

Echocardiographic studies including Tissue Doppler Imaging (TDI) were performed before initiation of therapy and during follow up exams under therapy over 5 and 9 months by now.

The echocardiographic images showed a change from hypertrophy to near normal of the interventricular septum and the left posterior wall. L.B. had initially a right ventricular intracavitary obstruction, disappearing under therapy. Surprisingly, the shortening fraction showed no changes, while the laboratory controls showed a considerable decrease of Brain Natriuretic Peptid.

The results of the TDI data showed regionally different courses. The left lateral wall showed no distinct changes, while the interventricular septum and the left inferior wall showed an increase in peak and mean systolic as well as diastolic velocities with increases from 25% to a doubling of the pre-treatment values. The right ventricle showed a remarkable increase in systolic (+75%) and diastolic velocity values for L.B. (ERT at 2 months), but a slight decrease for C.M. (start at 4 months).

We also noticed a distinct change of the curve pattern for the velocity curves for the Pompe patients versus normal curves.

To our knowledge, this is the first longitudinal echocardiographic and TDI examination of Pompe patients under ERT.

TDI might prove to be a more sensitive tool for general and regional improvement of cardiac function. It might be a diagnostic tool in the work up of differential diagnosis of hypertrophic cardiomyopathy. We would therefore suggest a detailed echocardiographic work-up of Pompe patients including TDI.

(For P-42, please see OP-23)

P-43

Does early pulmonary valve replacement in children with pulmonary regurgitation after repair of tetralogy of Fallot influence exercise capacity?

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Introduction: Severe pulmonary regurgitation after repair of tetralogy of Fallot (TOF) often causes right ventricular (RV) dysfunction and decreased exercise performance. The right timing of re-operation is still uncertain. The present study assessed the influence of PVR on exercise performance and compares it with the reduction of RV-size and function in magnetic resonance imaging (MRI).
Methods: We retrospectively analyzed 16 patients with TOF repair who had undergone PVR at a mean age of 14.1 ± 2.7 years. Indication for PVR was a right ventricular enddiastolic volume (RVEDV) >150 ml/m^2. Bovine xenografts (Contegra®) were used for PVR. Patients had exercise testing using a Bruce treadmill test at a median interval of 5.2 (range 1–20) months before and 7.5 (range 5–25) months after PVR. Clinical assessment, cardiac MRI and echocardiography were performed at the time of exercise testing to evaluate right ventricular size and function.
Results: Mean exercise capacity raised significantly after PVR (12.4 ± 1.6 vs 14 ± 2.4 METS, $p < 0.05$). MRI showed a significant reduction of RVEDV (193 ± 47 ml/m^2 vs 117 ± 28 ml/m^2, $p < 0.05$), but no significant changes in RV function.
Conclusions: Exercise capacity improves after PVR for pulmonary regurgitation in children with repaired TOF. There is no strong correlation between exercise capacity and RV function or RV size.

(For P-44, please see OP-22)

P-45

Intraoperative epicardial real-time 3D echocardiography to assist surgical repair of congenital heart defects

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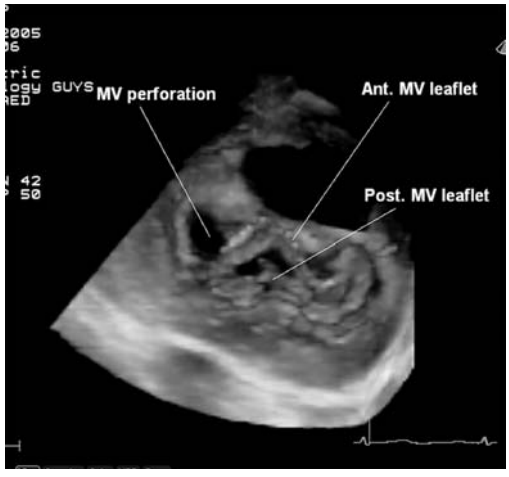
Introduction: Three-dimensional echocardiography (3D echo) has attractions for delineating congenital heart defects, thereby assisting surgical repair. An epicardial approach overcomes difficulties related to acoustic windows.
Methods: The use of epicardial 3D echo was reviewed retrospectively from January 2004 to end December 2005. Cases were selected for intraoperative 3D echo at surgical request as they were judged to be at high risk of residual lesions. All cases were evaluated using a Philips 7500 ultrasound system using the X4 matrix transducer. 3–4 volumetric datasets were obtained including both greyscale and colour flow Doppler. Ventilation was suspended during image acquisition to eliminate motion artrefact. TOE and 2D epicardial echo were in routine use throughout the study period.



Results: Twenty-one patients were evaluated. The lesions studied included supramitral membrane (n = 4), atrioventricular septal defect (n = 9), mitral valve chordal rupture (n = 2), mitral stenosis

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(n = 1), double orifice mitral valve (n = 1), mitral valve perforation (n = 1, see figure), subaortic VSD with aortic valve cusp prolapse (n = 1), subaortic stenosis (n = 1) and double outlet right ventricle (n = 1). Patient ages ranged from 2 months to 41 years and weight from 2.8 kg to 70 kg. In all patients, image acquisition was technically feasible. Echo data was available in less than 5 minutes for all cases, permitting discussion with the surgeon as surgical preparation continued. En-face views of AV valves could be constructed both from the ventricular or atrial sides, permitting accurate delineation of valve morphology and areas of AV valve regurgitation. In one case, a left AV valve described as "cleft" on echocardiography proved to be double orifice at surgery – the orifice nearest the aortic valve was slit-like, explaining the diagnostic error. Two patients died (10%) and 3 required reoperation within 30 days.
Conclusions: Important information relating to cardiac morphology and valvar function can be obtained with a high degree of diagnostic accuracy. Views can be constructed which are impossible using 2D techniques. Further work needs to be done to establish whether this technique reduces mortality or reoperation rates.

P-46
Usefulness of cardiovascular MRI and MRA in the post-operative assessment of the pulmonary arterial tree in cono-truncal malformations

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Introduction: Residual pulmonary artery (PA) anomalies after surgery for cono-truncal malformations are a diagnostic and therapeutic challenge. This study sought to assess the frequency and the therapeutic implications of residual PA anomalies with cardiac MRI/ MRA in the post-operative setting of these cardiac malformations.
Methods: Forty seven MRI/MRA studies were performed in 41 patients operated for cono-truncal malformations (23 tetralogy of Fallot, 8 pulmonary atresia with ventricular septal defect, 3 transposition of great arteries, 2 truncus arteriosus, 1 pulmonary stenosis and 4 complex anomalies). Thirty seven patients had corrective surgery (11 homografts, 9 transannular patches, 4 RV-PA non-valved tubes, 4 Contegra, 4 arterial switch operations and 5 complete corrections of Tetralogy of Fallot with infundibular resection and patching). MRI/MRA studies were performed with a 1.5 T machine and comprised spin-echo, cine, velocity-encoded and 3D Gadolinium-enhanced MRA sequences. Residual PA anomalies were searched in all patients; angiographic data were available in 13 patients and a comparison with MRI/MRA was made.
Results: 32/37 patients had postoperative anomalies of the pulmonary arterial tree. Left pulmonary artery (LPA) stenosis was the most common finding (14/32; the origin of the LPA was involved in 8/32), followed by stenosis of multiple sites (11/32). Isolated right pulmonary artery (RPA) stenosis was rare (2/32). The median time interval between MRI/MRA and angiography in the 13 patients undergoing both types of studies was 54 days. The findings between the two examinations were identical regarding stenoses and collateral vessels. In 4 patients, the MRI/MRA study allowed to plan interventional catheterisation with balloon dilatation and/or stenting of the obstructed arteries or coil-occlusion of systemic collaterals. Eleven patients had additional surgery based on MRI/MRA findings.
Conclusions: Post-operative anomalies of the PA in cono-truncal malformations can reliably be detected with MRI/MRA. Furthermore, this technique allows planning of the interventional or surgical procedure to correct the residual anomalies. As MRI/MRA is non-invasive and as reliable as angiography, it can replace or precede catheterisation during the follow-up of surgically corrected cono-truncal malformations.

P-47
Atrio-ventricular function and interaction in patients after Fontan operation compared to healthy volunteers: a magnetic resonance study

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Objectives: We quantified the interaction between atrial and ventricular function in patients after Fontan operation in comparison to that of healthy volunteers, using cine magnetic resonance imaging (MRI).
Methods: Twenty-four patients (15 female, 9 male, aged 5 to 44 years) and 24 age-matched healthy volunteers were studied using a 1.5 T Gyroscan ACS-NT (Phillips, Best, Netherlands) and cine MRI in short axis and transverse orientation for ventricular and atrial analysis. Patients were divided into 2 groups: group 1 (n = 19) with age at operation below 18 years; group 2 (n = 5) with age at operation above 18 years. The postoperative time interval was similar for both groups (5.7 ± 3.4 in group 1 and 6.7 ± 6 in group 2). The main ventricular morphology was left in all patients. Data from both groups were compared to those of the healthy controls.
Results: Ventricular volume and mass were similar to normal in group 1 but significantly larger in group 2 ($p < 0.01$). Ventricular ejection fraction (EF) was significantly impaired in group 1 compared to controls and was poorest in group 2 (controls: 65.5 ± 4.1, group 1: 55.8 ± 9.7, group 2: 44.4 ± 9.1; in %; $p < 0.03$). Atrial volumes were similar to normal in group 1 but significantly greater in group 2 ($p < 0.03$). Early diastolic (passive) function was significantly diminished in both group 1 and group 2 compared to controls (controls 19.5 ± 3.9, group 1: 10.1 ± 5, group 2: 7.8 ± 1.9; in ml/m^2; $p < 0.01$). Late systolic (active atrial) function was similar to normal in group 1 but significantly augmented in group 2 (controls 7.0 ± 3.4, group 1: 7.5 ± 3.6, group 2: 14.4 ± 2.9; in ml/m^2; $p < 0.02$).
Conclusion: Patients with Fontan operation performed before adulthood have normal ventricular and atrial volumes. Ventricular systolic function is reduced in all Fontan patients, with significant further reduction in adulthood. Early diastolic function is impaired in all Fontan patients, but late diastolic (active atrial) function shows a compensatory mechanism during evolving heart failure.

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(For P-48, please see OP-24)

(For P-49, please see OP-26)

P-50

Assessment of regional atrial and ventricular myocardial function in patients with hypertrophic cardiomyopathies using tissue Doppler imaging

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Introduction: Interaction between the atrial and ventricular function in patients with hypertrophic cardiomyopathies (HCM) plays an important role in the adaptation and compensation of the hypertrophic myocardium. We assess the regional and global ventricular and atrial function in patients with HCM using tissue Doppler imaging (TDI).
Patients and methods: The data of 20 patients with HCM (median age 12, range 2–18 years) who underwent conventional echocardiography and TDI were compared to those of 20 age-matched healthy subjects. In 4 apical views, TDI derived velocities, strain (S) and strain rate (SR), were measured in the ventricular and atrial walls. Isovolumic relaxation (IVRT) and contraction (IVCT) times were measured from the TDI spectral wall velocities.
Results: A significant reduction of the systolic and diastolic wall velocities at the basal and mid segments in HCM patients was found in the LV but not in the RV. Regional strain and strain rate values at the mid segments of LV and RV walls were significantly lower in HCM patients than in healthy controls ($p = 0.01$). Compared to normal subjects, patients with HCM had significantly reduced mean peak annular and mid segment IAS diastolic E velocity ($p = 0.001, 0.003$). Mean values of strain, systolic (SRS), early diastolic (SRE) and atrial (SRA) strain rate were found to be reduced in the left and right atrial walls ($p < 0.001$). Additionally, the left lateral annular early peak strain rate was negatively correlated to the left ventricular IVRT ($r = -0.7$, $p = 0.03$). Atrial velocities and strain rate parameters were not associated with thickness of the IVS septum or heart rate. However, late left atrial strain rate (SRA), representing the active atrial emptying, correlated negatively to the pressure gradient across the LVOT ($r = -0.5, p = 0.03$).
Conclusion: Altered regional atrial and ventricular function was found in HCM patients using TDI. The reduced TDI annular wall velocities may reflect the altered relaxation and systolic performance in these patients. Pressure overload in HCM patients may influence the atrial regional function and may have prognostic value with regard to atrial compensatory capacities and function.

(For P-51, please see OP-25)

P-52

Comparison of Tissue Doppler and echocardiographic wall-to-cavity ratios for screening for hypertrophic cardiomyopathy in the pediatric age range

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Introduction: It is well known that in patients with hypertrophic cardiomyopathy (HCM) the increase in wall thickness in many patients occurs in late childhood or early adolescence. The need for screening measures that helps to identify suspected gene carriers, that have not yet developed hypertrophy, but require continued monitoring is desirable. It has been reported in adult gene carriers that Tissue Doppler (TDI) pattern is abnormal before the hypertrophy develops. We therefore compared distributions of TDI velocities and conventional echo parameters in children with a 50% risk of inheriting HCM.
Methods: Twenty five children with a parent with clearcut HCM were screened, median age 12y (range 0.5 to 16 y), with a male/female ratio of 0.52. They were compared with age- and when possible sex-matched controls (n = 17).
Results: Thirteen of the 25 screened children (age ranged 0.5 to 15 y) had clinically apparent HCM (52%), and three had borderline hypertrophy above 95% prediction limit, without clearcut HCM; male/female ratio 0.69. In many measurements there was a large overlap between normal controls and identified HCM. Comparing clinically overt cases with normal controls, the individual parameters with best discrimination of pathological cardiac phenotype were: systolic posterior wall-to-cavity ratio > 0.5 (sensitivity = 92%, specificity = 93%, positive predictive value (PPV) = 92%, negative predictive value (NPV) = 93%), diastolic septal-to-cavity ratio > 0.24 (sensitivity = 62%, specificity = 100%, PPV = 100%, NPV = 75%), Fractional Shortening (FS) $> 40\%$ (sensitivity = 53%, specificity = 100%, PPV = 100%, NPV = 71%), mitral valve E/TDI e ratio > 8.8 (sensitivity = 54%, specificity = 75%, PPV = 64%, NPV = 67%), TDI e/a < 2.0 (sensitivity = 46%, specificity = 81%, PPV = 67%, NPV = 65%) and isovolumetric relaxation time (IVRT) > 78 ms (sensitivity = 31%, specificity = 100%, PPV = 100%, NPV = 66%). The test positive rate among all screened children vs percentage false positives among controls for: systolic posterior wall-to-cavity ratio > 0.5 (52% vs 7%), diastolic septal-to-cavity ratio > 0.24 (32% vs 0%), (FS) $> 40\%$ (28% vs 0%), mitral valve E/TDI e ratio > 8.8 (36% vs 25%) and TDI e/a < 2.0 (32% vs 19%) and IVRT > 78 ms (21% vs 0%).
Conclusion: In pediatric age ranges, wall-to-cavity ratios indicating systolic hypercontractility appear to have a higher sensitivity for the detection of mutation carriers than TDI-parameters. For clinically overt cases they also have a higher positive predictive value. There is a male preponderance in clinically overt cases in this age range.

P-53

Evaluation of the univentricular performance early and late after modified Fontan operation using tissue Doppler and magnet resonance imaging

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Introduction: Patients after modified Fontan operation (mod Fontan) are known to have diastolic dysfunction, whereas the role of systolic function remains unclear. This study aimed at assessing univentricular function in patients after mod Fontan as compared to healthy subjects with tissue-Doppler echocardiography (TDI) and MRI measurements.

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Patients and Methods: 45 patients with a mean age of 14 years (range 2–36 years) after mod Fontan were studied prospectively. All patients, as well as a control group of 45 healthy individuals matched for age and heart rate underwent conventional pulsed wave (PD) and TDI.

Isovolumetric contraction time (IVCT) was determined by PD and used for calculation of the Tei index. For the regional ventricular function, the mean longitudinal TDI-derived strain at the right anterior and at the left posterior walls (SRAW and SLPW) was recorded from an apical four chamber view and the sum calculated as the common longitudinal univentricular strain (SUNI = SRAW + SLPW).

20 patients underwent MRI examination with estimations of muscle mass (MM), end diastolic and end systolic volumes (EDV, ESV), ejection fraction (EF) and stroke volume (SV). Patients were then divided into two different groups with respect to a univentricular EF superior or inferior to 50%.

Results: 9 patients had univentricular systolic dysfunction (EF-MRI $< 50\%$) and 11 patients had normal univentricular systolic function (EF $> 50\%$). MRI derived MM, EDV and ESV were significantly larger in patients with reduced systolic function ($p = 0.01$, $p = 0.002$, $p < 0.0001$, respectively).

There was significantly prolonged IVCT in patients with systolic dysfunction as compared to patients with normal systolic function and to controls ($p < 0.05$ and $p = 0.004$, respectively), which resulted in an abnormally increased Tei index.

Univentricular strain correlated highly significant with MRI derived univentricular EF, ESV, and EDV ($r = 0.85$; $p < 0.0001$). Patients with EF $< 50\%$ had a univentricular strain $\leqslant 30\%$.

Conclusions: TDI derived strain is readily available and delivers reproducible quantitative information on systolic univentricular function. It shows high correlations with MRI assessing global univentricular performance, a global univentricular strain $\leqslant 30\%$ reflecting a reduced systolic function (EF $< 50\%$) as determined by MRI. This novel diagnostic tool might improve the medical management of patients after mod Fontan.

P-54

Diagnostic reference level and effective dose in paediatric cardiac catheterization

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Aim: According to national and international rules diagnostic reference levels (DRL) have to be observed for human x-ray examinations. However, up to now there are no DRLs for paediatric cardiac catheterization.

Methods: We have evaluated the dose area products for fluoroscopy (DAPF) and cine acquisition (DAPA) as well as the total time of fluoroscopy (FT) and the total number of acquired images (AN) of all paediatric catheterizations during a time span of six years. As the used equipment (Philips Integris 5000 BH) does not provide exact dose area products in infants and short cine runs only long runs ($>1\,Gy\,cm^2$ per run) were analysed.

Results: DAPA/AN ($r = 0.896$, $n = 1346$) and DAPF/FT ($r = 0.84$, $n = 2138$) are directly proportional to body weight (BW) over two orders of magnitude. The mean proportional constants were DAPA/AN/BW = 0.107 (± 0.064 SD) $mGy\,cm^2/kg$ and DAPF/FT/BW = 0.025 (± 0.03 SD) $Gy\,cm^2/min/kg$. We observed several examinations with very small total DAPs. For comprehensive diagnostic and interventional catheterizations we got an average total DAP (DAPA + DAPF) of 0.49 (± 0.58) $Gy\,cm^2/kg$. The third quartile, considered as DRL, was $0.58\,Gy\,cm^2/kg$. The difference between interventional and diagnostic examinations was not significant. The effective dose was derived from DAP/BW by use of a constant conversion factor ($9.26\,kg\,mSv\,Gy^{-1}\,cm^{-2}$), independent of the patient's age.

Conclusion: Our results correspond to a DRL of $40\,Gy\,cm^2$ of a 70 kg patient. This value lies significantly below the DRL for coronary angiography of adults (German Bundesamt für Strahlenschutz: $60\,Gy\,cm^2$), likely due to the heavy Cu-filtration (0.4 mm) we use as well as to institutional experience and to the specific needs in diagnostic and therapeutic catheterization of patients with congenital heart disease. We suggest a DAP/BW = $0.58\,Gy\,cm^2/kg$ as appropriate DRL for paediatric catheterization and recommend it instead of using different DRLs for age groups.

P-55

Effects of cardiopulmonary bypass on coronary flow in children – a Transthoracic Doppler Echocardiography study

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Background: Coronary flow abnormalities contribute to myocardial pathology such as arrhythmia and myocardial dysfunction. Previous invasive studies with positron emission tomography and intra-coronary Doppler guide-wire suggest decreased coronary flow reserve following cardiopulmonary bypass (CPB) surgery. The coronary blood flow assessment before and after CPB and off-pump surgery was investigated noninvasively by aid of transthoracic Doppler echocardiography (TTDE).

Methods: Peak flow velocity in diasole (PFVd) and Basal blood flow (BBF) in the left anterior descending coronary artery (LADCA) were assessed by TTDE before and 5 ± 1 days after surgery in 30 children. Eighteen children underwent CPB surgery for atrioventricular septal defect (mean age at surgery: 6 months) while off-pump surgery (aortic coarctectomy) was undertaken in the remaining 12 children (mean age: 10 days). All data are mean $\pm$ SD.

Results: After CBP surgery, LADCA's mean diameter increased markedly from 1.7 ± 0.3 to 2.1 ± 0.4 mm ($p = 0.001$). Coronary peak flow velocity in diastole (PFVd) increased from 36 ± 10 to 49 ± 16 cm/s ($p = 0.05$), the overall velocity time integral (VTId + s) from 9 ± 3 to 14 ± 5 cm ($p = 0.008$), and BBF from 27 ± 8 to 63 ± 18 ml/minute ($p = 0.004$). In contrast, all these measures decreased in children following off-pump coarctectomy: PFVd from 42 ± 14 to 30 ± 10 cm/s ($p = 0.008$), VTId + s from 11 ± 4 to 8 ± 2 cm ($p = 0.05$) and BBF from 44 ± 20 to 22 ± 14 ml/min ($p = 0.001$). The decrease in both BBF and PFVd correlated with the reduction in diastolic arterial pressure after coarctectomy ($r = 0.52$, $p = 0.08$, for both).

Conclusion: The reduced coronary flow reserve after CPB surgery could be due to increase in basal coronary flow. Off-pump surgery, at least in coarctation children seems, to have little or no impact on coronary flow as the post surgical decline in flow here relates to the reduction in cardiac pressure afterload.

P-56

Adverse effects of C – reactive protein on coronary microcirculation in children after cardiac surgery

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Background: Systemic inflammation has been suggested to underlie in part the elevated risk of arrhythmias and myocardial dysfunction during the first weeks after cardiac surgery. Recent transthoracic Doppler echocardiography (TTDE) studies from our centre indicated increased basal coronary flow in children 4–6 days after cardiopulmonary bypass (CPB) surgery. In these children, we investigated whether the inflammatory mediator C-reactive protein (CRP) could influence the postsurgical increase in coronary flow.
Methods: The peak flow velocity (PFV) and velocity time-integral in diastole (VTId) in the left anterior descending coronary artery (LADCA) and its diameter were assessed by TTDE 4–6 days after CPB surgery in 17 children (mean age at surgery 6 months) with atrioventricular septal defects. Plasma CRP was also measured.
Results: All children had CRP levels under the detection limit prior to surgery (<0.8 mg/L). CRP rose markedly by day 2 (median: 25, range: 4 to 142 mg/L), and remained elevated on day 4–6 after surgery (median: 11, range: 3 to 20 mg/L). At this time-point, both PFV and VTId significantly and inversely correlated with log CRP ($r = -0.74$, $p < 0.001$, and $r = -0.65$, $p < 0.01$, respectively). The association remained significant ($r = -0.67$, $p < 0.05$) after adjustment for body weight, perfusion and clamping duration, left ventricle's shortening fraction, and blood pressure.
Conclusion: The previously reported postsurgical increase in coronary flow velocity in children seems to be inversely associated with CRP levels, suggesting possible constrictive effects of CRP on coronary microcirculation. This could be one mechanism of the adverse effects of inflammation on myocardial physiology during the first week after cardiac surgery.

P-57
Toll-like receptor expression in circulating monocytes is correlated with the development of coronary artery lesions in Kawasaki disease patients

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Introduction: Kawasaki disease (KD) is a systemic vasculitis, and monocytes have been suggested to play a role in the inflammatory process. To date, precise mechanisms leading to coronary artery lesions in KD are unknown. We hypothesized that expressions of Toll-like receptor (TLR)2 and TLR4 in monocytes are related to the development of coronary artery lesions (CALs) in patients with KD.
Methods: We collected whole blood samples from 23 patients with KD before intravenous immunoglobulin (IVIG) treatment and 1 week afterward. Ten age-matched febrile children were compared as controls. Expressions of TLR2 and TLR4 on circulating monocytes were compared using flow-cytometry. Regulation of TLR2 expression by TLR4 activation was also analyzed after lipopolysaccharide (LPS) stimulation of monocytes.
Results: Expression of TLR2 on CD14 + monocytes were higher in patients with KD than in controls ($1.53 \pm 2.3\%$ vs.$1.18 \pm 1.09\%$). However, TLR4 expression on CD14 + monocytes were lower in patients with KD than in controls ($0.44 \pm 1.00\%$ vs. $0.71 \pm 1.41\%$). No significant change in TLR2 and TLR4 expression were observed 1 week after IVIG therapy. Among patients with KD, both TLR2 and TLR4 expression levels were higher in patients who developed coronary artery lesions (CAL+) than those who did not (CAL−). After stimulation with LPS, TLR2 expression levels decreased by 3% in CAL + group, but decreased by 19% in CAL − group.
Conclusion: TLR2 expression in circulating monocytes is correlated with the development of CALs in patients with KD. Modulation of TLR2 expression may be beneficial in preventing the occurrence of CALs in patients with KD.

P-58
Long-term follow-up of acute changes in coronary artery diameter caused by Kawasaki disease

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Introduction: In the acute stage of Kawasaki Disease (KD) coronary ectasia or aneurysm formation may occur. Despite a high incidence of regression of coronary dilatation, the risk of thrombotic occlusion and sudden cardiac death remains for some. This study assesses the long-term outcome of initially dilated coronary arteries in KD and tries to define risk factors for significant myocardial ischemia during follow-up.
Methods: Since 1985, 38 children with coronary changes due to KD have been identified. This reflects an incidence of 21% in our patients. These patients had a mean age of 2,4 years (0.01–12.8) and were mainly male (male:female 24:14). LCA was more often dilated than RCA. In 37 patients therapy with iv immunoglobin (IVIG) was initiated within a mean of 12 d (1–30) after begin of KD, in 1 patient 1 year after onset of the acute symptoms. All received Aspirin (ASA) and 3 also steroids. Mean follow-up is 5,6 years (0.2–16).
Results: We defined two groups – A aneurysm/ectasia ⩽ 4.5 mm ($n = 18$) and B aneurysm >4.5 mm ($n = 20$). Most of the aneurysms showed "spontaneous" regression in size. 6 patients developed severe coronary stenosis at the proximal and/or distal end of the aneurysm and needed an intervention (3 × endovascular balloon dilation or stent-implantation and 3 × bypass surgery) after a mean interval of 7,2 years (0,1–15,5 years). They all belonged to group B ($6/20 = 30\%$). 5 ($5/6 = 83\%$) of them had ECG changes preceding the intervention, whereas only 1 patient of those without stenosis had this. Using cox-regression only the maximum aneurysm size had a significant influence on the development of stenotic lesions ($p < 0.05$), but not age, sex or therapy delay.
Conclusions: Patients after KD with a maximum size of their coronary aneurysms >4.5 mm need close follow-up assessments because of an elevated risk for the development of coronary stenotic lesions. In case of new and even unspecific ECG changes coronary imaging modalities (angiography, MRI) have to be considered.

P-59
HMG-CoA reductase inhibitors (statins) is effective in the prevention of acute coronary arteries injury in a rabbit model for Kawasaki disease

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Backgrounds: Recently, therapy with HMG-CoA reductase inhibitors (statins) has been shown to significantly reduce major coronary events. In addition, recent observations suggest that some of the clinical benefits associated with statin therapy may be

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pleiotropic; that is, independent of their cholesterol-inhibiting action. In this study, we tried to evaluate the availability of statin mediated through its inhibitory and suppressive effect on inflammation in Kawasaki disease (KD) model rabbits
Methods and Results: To address this question, an animal model of allergic coronary arteritis was established by intravenous administration of horse serum (10 ml/kg) twice to juvenile rabbits. Rabbits were further divided into 3 groups: no treatment group (A), those given fluvastatin (B) and those pravastatin (C) at 3rd day before the second administration of horse serum. In group A, histological examination demonstrated marked mononuclear cell infiltration in intima and adventitia, disruption of internal elastic lamina, and intracellular edema in medial smooth muscle cell layer, which are similar to histopathological features of KD. These changes were most prominent at the day 3 and persisted until after the day 14. On the other hand, in both group B and C, those given statins were initially expressed mononuclear cell infiltration, but had fewer tendencies to develop cellular infiltration. These changes were peaked at the day 3 and 7. Treatment of statins improved the vascular thickening at the day 7 and 14.
Conclusions: These results suggest that statin is effective in the prevention of the acute phase of panvasculitis in the animal model for KD.

P-60
Linear shadow in coronary aneurysm on two-dimensional echocardiography in Kawasaki disease
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Introduction: The most common and serious clinical feature in Kawasaki disease is the involvement of the coronary arteries resulting in aneurysmal changes. Further, it has been revealed that a large or moderate aneurysm could often cause stenotic lesions or myocardial ischemia associated with late mortality. Therefore, it is essential to understand the gradual structural changes in coronary aneurysmal lesions (CALs) using some imaging method. Recently, we have detected linear shadows in large or moderate coronary aneurysmal lesions (CALs) on high-resolution two-dimensional echocardiography (2DE) in Kawasaki disease (KD) patients.
Methods: In this study, we investigated the origin and clinical significance of these linear shadows compared with findings on coronary angiography (CAG), magnetic resonance imaging (MRI), and intravascular ultrasound (IVUS) in 9 KD patients. The linear shadows detected on 2DE were along both the walls in the cavity of the aneurysms and were relatively straight. The spaces between the linear shadows and the coronary arterial wall had low echogenicity.
Results: The outer diameters of the CALs on 2DE were 6.90 ± 2.16 mm, and the inner diameters between the linear shadows were 4.06 ± 1.72 mm. The diameters of the CALs on CAG were 4.15 ± 1.94 mm and were almost equal to the inner diameters between the linear shadows on 2DE. There was a statistically significant positive correlation ($y = 0.99x - 0.10, r = 0.88$) between the diameter of the CALs on CAG and the inner diameter between the linear shadows on 2DE. On MRI and IVUS, a thickened intima was revealed in the same regions showing linear shadows on 2DE.
Conclusions: From these results, it was confirmed that the linear shadows in the CALs on 2DE reflected the existence of a thickened intima. Therefore, we believe that linear shadows on 2DE can act as a possible useful marker in the evaluation of the regression of the CAL or its development into a stenotic lesion occurring in the process of negative remodeling of coronary aneurysms.

P-61
Persistent left superior vena cava associated with cardiac and extra-cardiac congenital anomalies
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Introduction: A persistent left superior vena cava (LSVC) and its association with congenital heart disease (CHD) have long been recognised. There are few reports on the association between LSVC and congenital extra-cardiac anomalies.
Methods: We carried out a retrospective analysis of 4426 consecutive patients in our paediatric cardiology department who had echocardiograms in a seven year period. 4187 patients underwent screening echocardiograms for CHD and 239 patients had echocardiograms to exclude side effects of chemotherapy in the course of anticancer treatment. Echocardiographic findings were entered prospectively in a computerized database with the use of European Paediatric Cardiac Codes. The database was searched for the recording of a LSVC in the two groups. Of all patients diagnosed with a LSVC, additional data on congenital extra-cardiac anomalies was acquired by searching the medical records.
Results: A LSVC was found in 94 studies (91 cardiology patients and 3 oncology patients). The diagnosis of a LSVC was strongly associated with CHD (87%, OR 10.0, 95% CI 5.4 to 18.4, $p < 0.001$). The most commonly diagnosed cardiac anomalies were a ventricular septal defect in 34% and atrial septal defect in 20% of LSVC patients. Furthermore, the presence of congenital extra-cardiac anomalies was clearly associated with a LSVC (59%, OR 24.0, 95% CI 15.6 to 37.0, $p < 0.001$). Confirmed syndromes were frequently present (39%), including trisomy 21 (6%), 22q11 (6%), 45XO (4%), VACTERL association 9% (Vertebral defects, Anal atresia, Cardiac malformations, Tracheo-esophageal fistula with Esophageal atresia, Radial and Renal dysplasia, and Limb anomalies) and CHARGE association 4% (Coloboma, Heart defects, Atresia of choanae, Retardation, Genital and Ear anomalies). Of the LSVC patients 19% had multiple anomalies in different organ systems and a strong suspicion of a syndrome diagnosis, however not confirmed.
Conclusions: A persistent left superior vena cava is more commonly encountered in congenital heart disease than in the general population. The persistence of this vein also appears to predict the presence of congenital extra-cardiac anomalies. A persistent left superior vena cava may be a marker for anomalous development in general.

P-62
The inbred Brown-Norway rat as a novel animal model of persistent ductus arteriosus
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Introduction: Persistent ductus arteriosus (PDA) is a common cardiovascular anomaly in children caused by the pathological persistence of the sixth pharyngeal arch artery. The inbred Brown-Norway (BN) rat has been shown to differ significantly from other rat strains with respect to aortic elastin content and elastic lamina rupture leading to increased vascular fragility. In a pilot study a strikingly high prevalence of PDA in BN-rats had been observed.

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Objective: Characterization of the BN-rat as novel animal model of PDA.
Methods: Morphological study of PDA in 12 consecutive adult BN-rats from an inbred strain (BN/Rij) with conventional histology and immunohistochemistry. Neonatal and adult Wistar-rats served as controls.
Results: In all adult BN-rats a PDA was observed macroscopically, whereas a ligamentum arteriosum was found in adult controls. The macroscopic appearance of the PDA was tubular (n = 2), stenotic (n = 8) or diverticular (n = 2). The PDA had the structure of a muscular artery with intimal thickening. In the normal closing ductus of the neonatal control rats, the media consisted of layers of smooth muscle cells intermingled with layers of elastin. The intima was thin and poor in elastin. By contrast, the media of PDA in BN-rats elastin lamellae were absent and the intima contained many elastic fibers.
Conclusions: The abnormal distribution of elastin in the PDA of BN–rats suggests, that impaired elastin metabolism is related to the persistence of the ductus. The combination of a persistent ductus arteriosus in the present study and aortic pathology in adult BN-rats implicates a genetically determined factor that may be related to both: the PDA and the aortic fragility.

P-63
Right aortic arch with double vascular ring: atretic distal left arch and a left ligamentum arteriosum: not uncommon!
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Background: Double aortic arch is the most common form of vascular ring. The ring may assume many different anatomical patterns. In some patients transsection of 2 fibrous structures was required in order to relieve constriction. This study was set to investigate this morphologic variant.
Methods: The clinical data, imaging studies and surgical reports were reviewed in all 29 patients that were treated for vascular ring at our institution since 1984. We identified 8 patients (5 males, 3 females) with double aortic arch: dominant right arch, with both atresia of the distal left arch and left ligamentum arteriosum, thereby creating a double vascular ring.
Results: There were no associated cardiac malformations. Our diagnostic approach emphasised echocardiography along with bronchoscopy. Additional studies to evaluate the aortic arch anomaly included barium swallow (n = 7), angiography (n = 4), and computed tomographic scanning (n = 3). Differentiation from other aortic arch anomalies was made by the absence of flow in the left arch, the symmetric appearance of the subclavian and carotid arteries originating from both arches, the proximal downward course of the left subclavian artery, and the presence of an aortic diverticulum of Kommerell at the site of attachment of the fibrous cords.

The mean age and body weight at the time of operation were 10.2 ± 2.7 months (1.8 to 37.3 months) and 8.1 ± 2.1 kg (3.5 to 16.7 kg). Left thoracotomy was used in all patients. Both the atretic fibrous distal left arch and the ligamentum arteriosum were divided. There has been no operative or postoperative mortality. Complications occurred in 2 patients including pneumonia and chylothorax. At a mean follow-up of 3.3 years (range 10 days to 13.2 years), 4 (50%) patients showed signs of residual airway malacia.
Conclusions: Double vascular ring formed by a dominant right aortic arch, an atretic distal left arch and a left ligamentum arteriosum represents a type of double aortic arch accounting in our series for 20% of vascular rings. Correct preoperative diagnosis allows the surgeon to minimize dissection and to successfully relieve the patient's symptoms: both the atretic fibrous cord as well as the ligamentum arteriosum need to be divided.

P-64
Anomalous proximal origin and hypoplasia of the left pulmonary artery associated with monosomy 22q11 – a rare but clinically relevant anomaly
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Objectives: Deletions within chromosome 22q11 are thought to impair the normal migration and/or development of the cardiac neural crest cells and to interfere with the formation the outflow tracts and embryonic pharyngeal arches. The majority of these patients therefore present with conotruncal anomalies and/or anomalies of the 4th aortic arch derivates.

Isolated anomalies of the proximal pulmonary arteries are a rare abnormality found in conjunction with chromosomal deletions diagnosed in association with various cardiovascular lesions. Anomalous origin of the branch pulmonary arteries has been described as malposition with or without crossing of the pulmonary artery branches. In this situation however severe proximal hypoplasia or stenosis of the LPA has not been described before in detail.
Patients: We report 2 neonates with diagnosis of 22q11 deletion. Pt 1 had IAA type B with normal left-sided ductus, aberrant right subclavian artery, doubly-committed VSD and dysplastic aortic valve. In addition he had anomalous origin of the small, severely stenotic LPA, originating from the proximal pulmonary artery immediately distal to the pulmonary valve. Pt 2 had right aortic arch with aberrant left subclavian artery, perimembranous VSD and right-sided ductus. This patient also had an extremely short main pulmonary artery and anomalous proximal origin of a small LPA with mild stenosis. Pt 1 underwent intracardiac repair by a Norwood-Rastelli procedure at the age of 2 weeks with reconstruction and reanastomosis of the LPA. Pt 2 is still under clinical surveillance.
Conclusions: Pathogenetic considerations in our patients suggest additional maldevelopment of the proximal sixth pharyngeal arches which have been found to be inhabited by neural crest cells in the same way as the third and fourth pharyngeal arches. Although anomalous proximal stenotic origin of the LPA from the MPA has not been described yet, our patients confirm previous findings of proximal pulmonary artery anomalies in children with monosomy 22q11. This peculiar anomaly can be diagnosed by echocardiography as demonstrated in our patients. Preoperative recognition of this anomaly is mandatory, since it excludes the possibility of a banding procedure and may require patch-enlargement or reanastomosis of the stenotic LPA.

P-65
Assessment of fetal atrio-ventricular time intervals by tissue doppler and pulse doppler echocardiography: normal values and correlation with fetal electrocardiography
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Introduction: Accurate assessment of AV conduction is crucial for the early detection of evolving fetal heart block. By pulse wave (PD) and tissue (TDI) Doppler imaging, it is possible to study the

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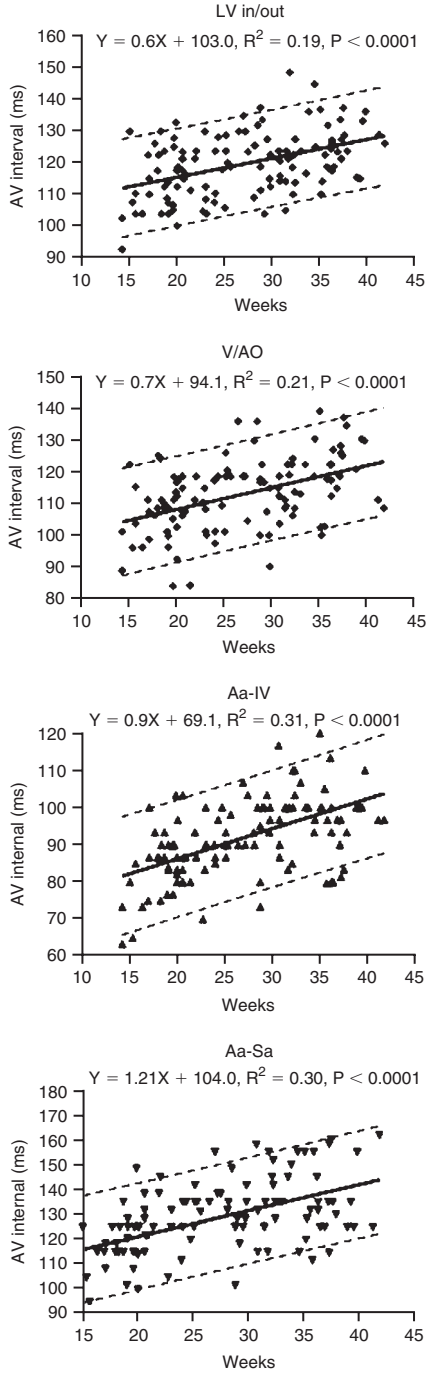
chronology of atrio-ventricular electrical events indirectly by their mechanical consequences.
Objectives: To 1) compare PD- and TDI-derived AV time to electrical PR intervals, and 2) to establish gestational age-specific reference values.



Methods: 131 women with a normal singleton pregnancy (14–42 weeks) underwent 196 fetal echo-cardiograms and 158 fetal ECGs. TDI-derived AV intervals were measured as 1) Aa-IV and 2) Aa-Sa at the lateral right ventricular wall. PD-derived AV intervals were studied on 3) left ventricular inflow/outflow (In/Out) and 4) SVC/aorta (V/Ao) recordings as previously reported. Averaged fetal ECG signals to measure PR intervals were obtained by a commercially available system (FEMO).

Results: AV interval measurements of good quality were possible by ECG in 61%, TDI in 100%, In/Out in 100%, and V/AO in 97% of exams. Aa-IV correlated better with PR intervals ($R^2 = 0.15$; $P < 0.0001$; bias: 8 ± 13.9 ms) than In/Out ($R^2 = 0.1$; $P = 0.002$; bias: 18.7 ± 14.8 ms), V/Ao ($R^2 = 0.06$; $P = 0.02$; bias: 12.4 ± 15.8 ms) and Aa-Sa ($R^2 = 0.11$; $P = 0.001$; bias: 32.8 ± 17.2 ms). There was a strong positive correlation between gestational age and AV intervals with all modalities, as shown in the figures (lines denote regression and 95% confidence intervals).
Conclusion: This study demonstrates the feasibility of fetal AV interval measurements by TDI. Reference data were established for TDI and PD methods that may prove useful for the diagnosis of first degree AV block. Aa-IV intervals correlated significantly better with PR intervals when compared to other imaging approaches, and should be preferred to measure fetal AV intervals in the surveillance of fetuses at risk of heart block if electrocardiogram or magnetocardiogram are unavailable.

(For P-66, please see OP-5)

(For P-67, please see OP-3)

P-68
Outcome after prenatal diagnosis of tetralogy of Fallot

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Objectives: To describe the phenotypic features of fetal tetralogy of Fallot (ToF) and to determine accuracy of prenatal diagnosis to predict surgical outcome.
Patients and methods: Over a period of 7 years, 202 cases of ToF (133 without and 69 with pulmonary atresia ToF-PA) were included. Fetal karyotyping was performed in 191 cases and screening for 22q11 deletion in 169 cases. Information on the pulmonary arteries (PA – main PA, PA branches and MAPCAs) were noticed. After birth or termination of pregnancy, available data on PA anatomy and on 1 year outcome were retrieved from the data base. Finally, prenatal anatomy of the PA was correlated with reparability of the defect at 1 year.
Results: Chromosomal anomalies were detected in 45 fetuses including 22q11 deletion. Extracardiac anomalies were present in 42% of the fetuses. Pregnancy was terminated in 72 (36%) of the cases. Reasons for termination were severity of the cardiac defect in 33%, chromosomal anomalies in 40.5% and extracardiac malformations in 26.5%. 127 (63%) neonates were born alive. Neonatal death occurred in 8 cases and was never related to the cardiac defect. Precise anatomy of the PA could be assessed in 163 fetuses. Prenatal description correlated with postnatal anatomy in 81% of the cases for the PA branches, 83% for the main PA, and 86% for the MAPCAs. Finally, when the fetal scan could describe 2 PA of normal size for the term, 81% of the patients had undergone complete repair at one year; conversely, when the PA were either hypoplastic or absent, only 51% of the patients had undergone complete repair at one year.
Conclusion: Prenatal diagnosis of ToF leads to a high rate of termination of pregnancy mostly because of extracardiac anomalies. A third of the terminations is related to the supposed severity of the defect. Precise scanning for the anatomy of the PA is mandatory as it correlates with postnatal anatomy in more than 80% of the cases and its reliability to predict outcome has a good positive predictive value.

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(For P-69, please see OP-6)

P-70

Functional pulmonary atresia or systolic pulmonary regurgitation – phenomenon of severe fetal or neonatal right ventricular failure

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Introduction: Functional pulmonary atresia (funPA) has been reported in fetuses and newborns with severe Ebstein anomaly. Few anecdotal reports described this condition with normal heart anatomy. The aim of this study was to analyze fetal and neonatal echocardiographic exams with funPA and try to explain pathophysiology of this phenomenon.
Methods: Retrospective evaluation of echocardiographic exams in cases with funPA and follow-up analysis.
Results: 8 fetuses and one neonate had funPA. There was regurgitant flow through the pulmonary valve during systole, and tricuspid insufficiency indicated low right ventricular pressure in all of them. 4 fetuses and 1 newborn (who developed funPA after pulmonary valvuloplasty) had Ebstein anomaly. 2 died, 3 are alive. 3 fetuses with funPA had normal heart anatomy. Two of them were hydropic recipients in twin-to-twin transfusion syndrome. Both had severe TR, impaired heart function, abnormal ductus venosus and umbilical vein flow. They were born in 27 and 29 weeks respectively. One recipient died, three others are alive. It was normal flow in the PA in living recipient. In the last fetus severe arrhythmia (intermittent bradycardia and SVT) appeared at 25th weeks of pregnancy. Fetal condition deteriorated despite various methods of treatment. In 32 weeks funPA appeared and three days later emergency CC was performed due to fetal compromise (psudo-normalization in middle cerebral artery flow). There were signs of myocarditis with severe arrhythmia and hypertrophic cardiomyopathy, with normal forward flow in the pulmonary artery in the newborn.
Conclusion: FunPA is probably a result of severe right ventricular failure in fetuses and neonates. In cases with Ebstein anomaly it was due to abnormal RV, which was unable to pump the blood against high resistance pulmonary circulation. FunPA in fetuses with normal heart anatomy could be compared to severe left ventricular insufficiency in adult patients in whom diastolic mitral regurgitation is diagnosed. In all fetuses with impaired RV function careful examination of pulmonary flow should be performed during each exam, as it can change and deteriorate during prenatal life. It should be kept in mind that funPA is in fact "systolic pulmonary regurgitation", what is important information for neonatal treatment.

P-71

Fetal volumetry: validation of the method using 3-dimensional ultrasound of heart, brain and liver

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Introduction: Three- and four-dimensional ultrasound have opened new aspects of fetal examination. Besides "babyfacing" and differentiated visualisation of external anomalies, volumetry of fetal organs has become possible. Aim of this prospective interdisciplinary study was the validation of this method with respect to the fetal myocardial mass, the liver and the brain.
Methods: One hundred and four fetuses between 14 and 38 weeks of gestation were prospectively studied. In 39 of them, a structural defect of the heart could be diagnosed, in 65 of them it was excluded. The sonographic examination was performed with a Voluson 730 Expert machine GE, Germany. Three- and 4D data sets were acquired for the heart, brain and abdomen and were stored for offline analysis. Using the 4D-view-software (GE, Germany) measurement and calculation of the following parameters was established twice each: epi- and endocardial area, myocardial thickness, ventricular length and myocardial mass of each ventricle, liver and brain volume. Descriptive statistics, coefficients of correlation and variation as well as Bland Altman analyses were performed.
Results: Acquirement of volumetric datasets lasted approx. 10 minutes, offline analysis took up to 45 minutes per patient. The latter was possible in 75% of the stored volumetries. For the fetal liver and brain volumetries, coefficients of variation ranged between 2.57 and 3.48, respectively ($r2 = 0.98$). For the myocardial parameters, coefficients of variation were between 5.96 and 12 (r2 between 0.88 and 0.94) in the left heart, and between 6.59 and 16.4 (r2 between 0.74 and 0.9) in the right heart. Fetal liver and brain volumes were positively associated with gestational age; also fetal myocardial mass showed a significant increase with advancing gestational age. Neither fetal liver nor brain volumes were different in fetuses with or without congenital heart disease.
Conclusions: Prenatal volumetry of organs and heart is time consuming, but reliable depending on the examined parameter. Basing on our data, which present fetal reference values for liver and brain volume as well as for myocardial mass, clinical application becomes more realistic and further scientific programs can be performed.

P-72

Cardiac problems in the offspring of the diabetic mother

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Introduction: The group of pregnant patients with maternal diabetes is expanding rapidly mainly because of the increase in woman with type II (insulin resistant) and gestational diabetes. The meticulous control of the glycaemia of future mothers is presumed to diminish the incidence of complications due to the maternal diabetes, including prevention of congenital heart disease (CHD) and hypertrophic cardiomyopathy (HCM). In order to assess the risk of the foetus for CHD and/or HCM we studied a well controlled population of mothers with diabetes passing through our perinatal unit between 2003–2005.
Methods: A retrospective study of all comers with diabetes was performed. Data were acquired regarding prenatal echocardiographic evaluation, delivery, and postnatal condition, including sequential echocardiographic follow-up.
Results: Included in the study were 77 pregnancies, 78 neonates, of which 22 mothers with type I, 1 with type II and 54 with gestational diabetes, of which 37 insulin dependant. 3 neonates had congenital heart disease (CHD), 1 DORV, 1 Pulmonary Stenosis, 1 VSD all diagnosed prenatally. HCM was encountered in 11 and 64 neonates had a normal intracardiac anatomie and no signs of HCM. 1/11 pt with HCM died immediately post-natally and 1/11 required premature delivery because of pericardial effusion, RV dilatation and bradycardia. Postnatal follow-up showed regression of the HCM in all patients in a period of 5 months (range 3–7).

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The 22 Type I pregnancies resulted in 2 neonates with CHD (PS, VSD), 1 HCM mortality, 1 HCM delivered prematurely and 6 HCM with spontaneous regression. The 55 Type II and Gestational diabetes pregnancies resulted in 1 pt with DORV and 3 with HCM and spontaneous regression.
Conclusion: The presumed solution of diminishing the incidence of cardiac complications by the meticulous control of glycaemia of future mothers seems to have a limited impact. This more rigorous control resulted in better glycaemic values, but not necessarily in the prevention of CHD and late development of HCM. This leads us to the conclusion that although no definite predictive parameters for foetal demise or malignant outcome can be presented, close monitoring of these pregnancies may prevent perinatal catastrophes.

P-73
Outcome of cases with tricuspid valve anomaly presenting in fetal age
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Objectives: To analyse retrospectively the characteristics and outcome of cases with tricuspid valve anomaly diagnosed at fetal echocardiography.
Material and methods: Between 1995–2005 23 fetuses were diagnosed in our center to have Ebstein (Ebst) or non Ebstein anomaly (NEbst) of the tricuspid valve, at 21 to 36 week's gestation (wg), median 30: 11 had Ebst and 12 NEbst, one with associated mitral dysplasia. One fetus with Ebst had associated labiopalatoschisis, 2 with Ebst had familial history of congenital heart disease: one had a parent with transposition of great arteries and a previous sybling with Prune-Belly syndrome and the father of the second one had Robertsonian translocation; 2 women were on drugs in pregnancy: 1 lithium, 1 gardenal. Echocardiographic features, course in utero and after birth were analysed.
Results: Echocardiographic features: 9/11 fetuses with Ebst had a severe displacement of the TV and severe TR, 2 had moderate-severe pulmonary stenosis (PS); the case with lithium therapy was of a mild entity. 9/12 cases with NEbst had severe TR, 3 moderate; 2 had moderate organic PS and 3 had functional PS evolving to atresia in 1.

Fetal hydrops (FH) was present at presentation at 29–31 wg in 6/23 cases (22.7%) − 5 Ebst, 2 NEbst.
Outcome: 2 cases opted for the termination of pregnancy, 2 Ebst died in utero at 31 w. both, (1 with FH, 1 in supraventricular tachycardia). The remaining cases were delivered all but one by cesarean section at 31–38 wg. Five cases died in severe conditions at 1–7 days. Three infants with Ebst and 3 with NEbst were operated and 2 survived, one being reoperated; the case with associated mitral dyplasia improved the TR but required a mitral valve plasty and is alive at 6 yrs. Total mortality was 10/21 cases that continued pregnancy (47.6%), 4/6 with FH. Eleven cases are alive at 6 m–7 yrs, stable or improved (3 Ebst, 8 NEbst).
Conclusions: Our data confirm a variable spectrum of tricuspid valve anomalies presenting in utero. Severe variants with heart failure have a poor outcome, milder forms of both variants may improve after birth.

(For P-74, please see OP-4)

P-75
N-terminal Pro-B-type natriuretic peptide as a predictive risk factor in fontan operation
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Objectives: To investigate the correlation between the plasma level of N-terminal pro-B-type natriuretic peptide (N-BNP) and several known risk factors influencing outcomes after Fontan operations, and to assess whether the N-BNP levels can be used as a predictive risk factor in Fontan operations.
Methods: We studied 35 patients (21 men and 14 women, mean age 4.7 years [range: 4 month–16 years]) with complex cardiac anomalies who were admitted to the Sejong Hospital for the catheterization between June 2004 and February 2005. Underlying diagnosis included univentricular heart (n = 11), double outlet right ventricle (n = 9), heterotaxy syndrome (n = 4), tricuspid atresia (n = 3), and others (n = 8). Plasma N-BNP concentrations were measured before catheterization. Cardiac catheterization was performed in all subjects. Mean right atrium pressure, ventricular end-diastolic pressure (EDP), mean pulmonary artery pressure (PAP), pulmonary venous pressure, systemic arterial pressure, and oxygen saturations were obtained. Cardiac output and pulmonary vascular resistance were calculated by Fick method.
Results:

1) Plasma N-BNP levels exhibited statistically significant positive correlations with mean PAP ($r = 0.70$, $P < 0.001$), pulmonary vascular resistance ($r = 0.57$, $P < 0.001$), RVEDP ($r = 0.63$, $P < 0.001$), LVEDP ($r = 0.74$, $P < 0.001$), and cardiothoracic ratio ($r = 0.71$, $P < 0.001$).
2) The area under the ROC curve using N-BNP level to differentiate risk groups in Fontan operation was high: 0.868 (95 percent CI, 0.712–1.023, $P < 0.01$).
3) The cutoff value of N-BNP concentrations for the detection of risk group in Fontan operation was determined to be 332.4 pg/ml (sensitivity 83.3 percent, specificity 82.7 percent).

Conclusions: These data suggest that plasma N-BNP levels may be used as a predictive risk factor in Fontan operations, and as a guide to determine the mode of therapy during follow-up after Fontan operations.

P-76
Unilateral exclusion of the pulmonary artery
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Isolated congenital unilateral exclusion of a pulmonary artery (UEPA) is a rare malformation.

Most cases are presented as case-reports in literature. The clinical presentation and spectrum is not well defined yet.
Methods: A retrospective data-base study in forty-thousand patients between September 1976 and September 2004. Search criteria were absent or excluded unilateral PA associated with normal heart or with simple associated defects (L-R shunts, single valvular stenosis). Data were collected about age, symptoms at diagnosis, treatment and outcome.
Results: Our retrospective study presents the data, treatment and outcome of fourteen patients with UEPA during study period.

Age at diagnosis ranged between 2 days and 12 years. Presenting symptoms at diagnosis were dyspnoe and exercise intolerance in

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eight, recurrent pulmonary infections in five and lung infarction in one patient. Some had combinations of symptoms. One patient was asymptomatic at the time of diagnosis. In all patients the affected lung was on the opposite side of the side of the aortic arch. Pulmonary hypertension was present in nine at diagnosis. Surgical procedures were performed in six patients. At follow-up five are asymptomatic. There was no mortality in our group.
Conclusions: Our study proves that isolated UEPA is a rare malformation. Clinical symptoms, like dyspnea or exercise intolerance or recurrent respiratory infections, are almost always present and are often age related. Our angiographic data support the embryologic mechanism described in the development of UEPA. The missing part is a consequence of early embryologic involution of the ventral proximal part of the sixth aortic arch while the distal sixth aortic arch part (which become the ductus) remains connected to the ipsilateral intrapulmonary PA. Our study remains inconclusive about the positive effect on PA patency after early restoration (surgical or interventional) of blood supply to the affected lung. Long term survival in our group seemed to be determined by absence of pulmonary hemorrhage and heart failure due to severe pulmonary hypertension.

P-77

Silent pericardial effusions in anorectic girls

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Introduction: The frequency of Eating Disorders (obesity, boulimia and anorexia nervosa (AN)) is rapidly increasing in western societies. It is now estimated that up to 2 to 5% of adolescent school age girls have AN to some degree and are referred to psychiatric units, where it is not always recognized that AN may affect both body and mind.
Aim: To evaluate the impact of caloric deprivation upon somatic functions.
Methods: 89 girls with AN were referred between 01/09/2002 and 01/11/05 to our tertiary eating disorder unit. The diagnosis of AN was confirmed according to the criteria of the Diagnostic and Statistical Manual of Mental Disorders (DSM IV). None of the patients had clinical symptoms or signs of cardiovascular disease and were anamnestically negative for known cardiac or any systemic disease involving the cardiovascular system. All patients underwent a complete clinical examination, serum biochemistry for electrolytes, albumin, thyroid hormones, vitamins ADEKB, folic acid and minerals including iron, zinc, iodium and selenium.

All subjects underwent a 2-D echocardiogram and 12 lead ECG.
Results: In 12 cases pericardial effusions were observed at admission. The mean age of the whole group was 14,1 ± 1,74 years (8,0–17,0 years). The average body mass index was 14,9 ± 2,07 kg/m^2 (11,0–20,0). The mean LVM (left ventricular mass) is 84,6 ± 21,7 gr.
Summary: 12/89 AN girls (13,5%) had a pericardial effusion. This is probably multifactorial: in all those patients there was always a very severe and rapid weight loss (29,0 ± 10,3%), BMI (13,0 ± 1,48 kg/m^2),in 7/12 girls there was an association with secondary hypothyroidism. After 12–18 weeks of realimentation there was a disparition of the effusion in 10/12 patients , linking this abnormality most probably to the malnutrition.
Conclusion: It is mandatory that every severely malnourished AN patient has a complete somatic check-up including ECG and echocardiography.

P-78

Natural medium-term course of rhabdomyomas in children

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Objectives: To evaluate clinical presentation, echocardiographic findings and morbidity and mortality rates for cardiac rhabdomyoma (CR) in children registered from three university hospitals.
Methods: Medical files of all children registered at three university hospitals with the diagnosis of CR in a 10-year-period (1986–2005) were reviewed. The diagnosis CR was made by echocardiography, based on the presence of tumour(s) with or without a history of tuberous sclerosis (TS). The location and number of the tumours were determined. Analysis was made of the clinical manifestation, further medium-term follow-up, serial ECG and echocardiographic studies.
Results: 29 children with CR were identified; two were excluded from the study because lack of follow-up. 15 boys and 12 girls were included with a median age of 180 days (range 0 days to 11.4 years). In 24 out of 27 patients CR was associated with TS, and in 5 a positive family history of TS was present. In 6 patients the diagnosis was made in utero, in 12 in infancy, in 9 later. Cardiac signs were present in two patients, six patients manifested with rhythm disturbance (1° AV-block (n = 1), supraventricular tachycardia (n = 3) and ventricular tachycardia and/or ventricular fibrillation (n = 2). In 19 patients cardiac signs were absent. All patients were treated during a mean follow-up of 3.6 years. 8 children had single and 19 multiple tumours. Tumour sites were ventricular, atrial, the interventricular septum and combined in 24, 3, 9 and 9 patients, respectively. Outflow tract obstruction was present in 3, valvar regurgitation in 3 patients. During follow-up, 2 patients achieved complete and 21 partial regression of the tumours, four remained unchanged. Progression of the tumour was not observed. All patients were treated conservatively. No cardiac surgery was required. One patient died of unrelated skin malignancy. Of the 6 patients with rhythm disturbances 5 improved, one recent patient has symptomatic arrhythmias (ventricular tachycardia and ventricular fibrillation) despite treatment.
Conclusions: CR is diagnosed usually fetally or in infancy. Most patients remained asymptomatic during medium-term follow-up and spontaneous complete or partial regression occurred frequently. A conservative approach is indicated.

P-79

Myocardial infarction as first manifestation of pulmonary arteriovenous fistula in a 15 year old girl

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Background: Pulmonary arteriovenous fistula (PAVF) is an abnormal vascular connection between a pulmonary artery and a pulmonary vein. We present a young patient with myocardial infarction as first manifestation of PAVF, most probably due to paradoxic embolism.
Case report: A previously healthy 15 year old girl presented with a 90 minute history of sudden-onset chest pain associated with breathlessness. Based on typical ECG changes and raised enzymatic markers acute inferior myocardial infarction was diagnosed. After successful thrombolysis with recombinant tissue plasminogen activator her clinical condition improved quickly and cardiac enzymes normalised. Repeated echocardiograms demonstrated a hypokinetic segment of the inferior wall. Subsequently coronary angiography showed normal appearances of the coronaries. Routine coagulation studies revealed normal results. Screening for autoimmune and metabolic disorders was negative. The most outstanding clinical feature was a slightly reduced oxygen saturation between 88% and 92% in supine position, whereas oxygen saturation decreased to 85% in upright position. A contrast transoesophageal echocardiography demonstrated significant intrapulmonary right-to-left shunting in the presence of an intact interatrial septum. The presumed diagnosis of multiple PAVF was confirmed by means of a computed tomography of the chest. Three weeks after admission cardiac catherisation was performed. A right and a left-sided PAVF were successfully closed with 10 mm vascular plugs and a detachable 5 mm coil. The postprocedural recovery was uncomplicated and the girl was discharged on peroral medication with ramipril and aspirin. In a subsequent genetic testing a heterozygote genotype for the G20210A prothrombin mutation was found.
Conclusion: To the best of our knowledge this is the first report of myocardial infarction as a first manifestation of PAVF. In young patients with myocardial infarction paradoxic embolism has to be considered, especially in patients with prothrombotic disorders. Orthodeoxia may be a valuable hint to the diagnosis of PAVF and transoesophageal echocardiography should be performed to exclude not only persistent foramen ovale but also PAVF.

P-80
Exercise improves surrogate parameters of early atherosclerosis and cardiovascular risk profile in obese children

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Introduction: Atheroslerosis is a disease that begins in childhood and primary prevention strategies are likely to be most effective if instituted early. Impaired flow mediated vasodilation (FMD) and increased intima-media thickness (IMT) are surrogate parameters of early atherosclerosis and correlate with a number of known cardiovascular risk factors (RF) in children. We investigated whether an exercise programm improves vascular changes and RF in obese children.
Methods: Seventy obese subjects (14 ± 1.7 years) were randomly assigned to a six month exercise or non-exercise protocol. We examined the influence of exercise training (1 hour, 3 times per week) on FMD, carotid IMT, and risk profile.
Results: At baseline obese children showed significantly ($p < 0.001$) impaired FMD (4.92 ± 2.5% vs. 10.1 ± 2.6%) and increased IMT ($p < 0.001$) compared to 55 lean control children (0.49 ± 0.07 mm vs. 0.36 ± 0.05 mm). Multivariate risk analysis showed that vascular changes were independently associated with distinct obesity (BMI > 30 kg/m^2), reduced physical fitness (Wmax < 2.5 w/kg), and elevated levels of fibrinogen (>3.5 g/l), insulin resistance (HOMA > 4) and triglycerides (>1.2 mmol/l). After a six month exercise training significant improvements were observed in the exercise group for FMD (+3.8% (+77%), $p = 0.001$), IMT (−0.05 mm (−8.1%), $p = 0.02$) and risk parameters like body mass index (−2.6 kg/m^2 (−8.4%), $p < 0.001$), mean ambulatory systolic blood pressure (−6.9 mmHg (−5.6%), $p = 0.041$), Wmax (+0.35 w/kg (+16.6%), $p = 0.019$), fasting insulin (−2.4 pmol/l (−17.5%), $p = 0.005$), triglycerides (−0.35 mmol/l (−9.2%), $p = 0.02$), fibrinogen (−0.32 g/l (−8.9%), $p = 0.05$) and C-reactive protein (−2.72 mg/l (−57%), $p < 0.001$). The group of obese control children showed no significant differences of vascular status and a aggravation of risk profile after the six month non-exercise protocol.
Conclusions: Early vascular wall changes are associated with a number of obesity related RF. Regular exercise restors endothelial dysfunction and improves IMT and cardiovascular risk profile in obese children. This study supports the value of an exercise program in the treatment of obese children in a primary prevention setting.

P-81
Cardiac outcome of children with 22q11 microdeletion syndrome and congenital heart defect

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Objectives: Congenital heart defects (CHD), particularly conotruncal (cCHD) are a major feature of the 22q11 microdeletion syndrome (22q11). The prevalence and the spectrum of clinical symptoms of 22q11 are well known but information about the clinical course and outcome are rare. The aim of the study was to evaluate the clinical course, postoperative period and early results of cardiosurgical treatment in children with cCHD and with and without 22q11.
Methods: Among 310 pts 22q11 microdeletion was diagnosed in 17 (5,5%) children. Out of 17 in 15 (88%) pts cCHD were observed (TOF n = 6, AP + VSD n = 2, TOF/absent PA valve n = 1, DORV n = 2, TAC n = 3, IAA type B n = 1). Two groups of children were evaluated: group A consisted of 11 pts with cCHD (TOF group) and 22q11 and group B consisted of 111 pts with the same cCHD, but without 22q11. The both groups were compared according to the clinical course, age at the surgical correction of cCHD, postoperative complications, duration of intubation, length of hospital stay (LOS) and deaths.
Results: Clinical cardiological status of pts before surgical repair of cCHD was comparable in both groups. Primary correction of cCHD was done in 36% vs 35% of pts in group A and B, respectively. The mean age at surgical correction was 27 months in group A compared with 19 months, in group B. In group A the postoperative period was more often complicated (n = 10/90,9%) and the intubation period was longer (mean 8,1 days) comparing with group B (n = 45/40,5%; mean 3,8 days). The length of stay (LOS) was mean 28,8 days in group A and mean 27,5 days in group B. Mortality rate in group A was 17,6%, in group B 3,4%.
Conclusion: (1) In children with cCHD and 22q11 microdeletion the postoperative period after surgical correction was more often complicated and length of hospital stay was longer comparing to children with the same CHD but without 22q11. (2) Mortality rate in group of patients with 22q11 was significantly higher than in children without microdeletion.

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P-82

Predictors of embolization in paediatric infective endocarditis: experience from a developing country

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Introduction: Infective endocarditis (IE) remains a serious disease despite several advances in its diagnosis and treatment. The complication of embolization is an important contributor to the morbidity and mortality associated with IE. Predicting which vegetation will embolize has been difficult in spite of several publications on this issue. We describe our experience in 19 children diagnosed to have IE over the last 2 years.
Methods: Hospital records of 19 consecutive children (age 6 months to 13 years, mean 7 ± 4.1 years) were analyzed for clinical and echocardiographic data, hospital course, complications if any, type of treatment received and the outcome.
Results: Congenital heart disease was the underlying cause in 17/19 cases, 10 of these had undergone palliative or corrective surgery more than 6 months ago. History of fever was of >4 weeks duration in all. Blood cultures were positive in only 11/19 cases, 4 cases had fungal IE. Transthoracic echocardiography showed vegetations in all, the size was >10 mm in 8, 3–10 mm in 6 and <3 mm in 5. A total of 9 cases (47%) developed embolization, 5 of these had vegetation size of >10 mm. Embolization was acutely fatal in 5, 4 of these had vegetation of >10 mm. 3 of 4 patients with fungal endocarditis embolized. Interestingly 4 embolizations occurred after over two weeks of antibacterial/antifungal therapy. Failure to demonstrate reduction in size of vegetation on therapy was also predictive of embolization (8/9). No embolization occurred in those with vegetation size <3 mm. A total of 6 patients underwent surgery, 4 in large vegetation group (one post embolization), and 2 in small vegetation group due to aortic root abscess, all these survived. In the non-surgical group, 5 of 13 patients (38%) expired.
Conclusions: The incidence of embolization was much higher in our experience possibly related to late presentation and thereby large sized vegetations. Vegetations >10 mm in size and not decreasing on treatment were more likely to embolize. Embolization can occur even after weeks of treatment in an otherwise stable case. An early aggressive surgical approach is helpful in reducing mortality in this group.

(For P-83, please see OP-10)

(For P-84, please see OP-7)

P-85

Assessment of abdominal aorta stiffness in children with bicuspid aortic valve

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Introduction: Bicuspid aortic valve is the most common congenital cardiac malformation, with a prevalence of 1–2% in the overall population. Associated aortic wall abnormalities due to cystic medial necrosis have been reported in the subjects with bicuspid aortic valve. The aim of this study was to evaluate the stiffness of the abdominal aorta in children with bicuspid aortic valve.
Methods: Thirty-five children with normally functioning or mildly regurgitant bicuspid aortic valve and 35 healthy children as a control group were enrolled for the study. The mean ages were 10.3 ± 2.3 years (22 male) and, 10.7 ± 1.9 years (21 male) in the study and control group respectively ($p > 0.05$). Systolic and diastolic blood pressures were measured at the brachial artery by sphygmomanometer. All subjects 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 β index (β) and, aortic distensibility (DIS) were calculated using the measured data.
Results: Diameter of the aortic annulus was found significantly larger in the children with bicuspid aortic valve than the control group (18.1 ± 3.0 vs. 16.6 ± 2.5 mm, $p < 0.05$). Although there was not significant difference in abdominal aorta systolic diameter between two groups (10.7 ± 2.4/10.4 ± 1.6 mm, $p > 0.05$); in the children with bicuspid aortic valve, S and DIS (1.04 ± 0.2/1.4 ± 0.4, $p < 0.001$) parameters were found significantly lower and, Ep (200 ± 39/153 ± 47, $p < 0.001$), Ep* (3.42 ± 0.9/2.5 ± 0.9, $p < 0.001$) and β (1.1 ± 0.3/0.84 ± 0.3, $p < 0.001$) were found significantly higher compared to control group.
Conclusions: Aortic annulus diameter was larger in the children with bicuspid aortic valve compared to the control group. Although there was not any difference in abdominal aorta systolic diameter between the study and control group, abdominal aortic stiffness was found to be increased in the children with bicuspid aortic valve compared to control group.

P-86

Cardiological symptoms and final diagnosis – multicentre study

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Objective: To evaluate a relation between symptoms at admission to tertiary pediatric cardiology centre and final diagnosis.
Methods: Data were collected prospectively using form completed on-line in Internet for each new patient in 8 outpatient clinics over the year.
Material: 14554 patients (mean age 5,8 ± 5,8 years, median 3,3 years). They were divided into 6 groups: newborns – 9,6%, infants <6 mths – 16,3%, infants >6 mths – 7,8%, young children (2nd and 3rd year of life) – 14,9%, pre-school children (4–7 years) – 12,3%, school children (8–14 years) – 24,3%, adolescents (14–18 years) – 14,8%.
Results: Heart murmur was the main cause of referral (55,8%) in each group, especially in children before 7th year of life. Cyanosis was observed in newborns and infants (1,8–4,4%). Arrhythmias (7,2%), syncope (6,4%) and chest pain (3,4%) were the important causes of referral in patients older than 7 years. Normal heart was found in 61% patients. Congenital heart defects (CHD) were diagnosed in 23,6% patients, mostly in children in 1st year of life, arrhythmias were recognized in 7,8% cases, especially in school children (12,8%) and adolescents (19,0%). In more than 70% patients with murmur, failure to thrive, recurrent infections and chest pain normal hearts were found. Murmur coexisting with failure to thrive and/or cyanosis, and/or poor exercise tolerance was a symptom of CHD in more than 50% cases. In 74% patients referred to cardiologist because of arrhythmias the diagnosis was confirmed. Syncope was caused by arrhythmias only in 9% cases.
Conclusions: Heart murmur was the main cause of referral to pediatric cardiologist, in older children (>7 years) the frequency

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of arrhythmias, syncope and chest pain increased. More than half of patients referred to pediatric cardiology centers had a normal heart. Isolated heart murmur was rarely a symptom of cardiac malformation, but murmur coexisting with failure to thrive and/or cyanosis, and/or poor exercise tolerance in more than half of cases was a symptom of CHD. The suspicion of arrhythmias was confirmed in most of children while syncope was caused by arrhythmias only in 9% patients.

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P-87

Congenital cardiovascular defects in children with intestinal malrotation: a 25 years cohort study of a serious entity

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Introduction: Intestinal malrotation (IM) is a congenital malformation that is frequently seen in patients with heterotaxia, an abnormality usually accompanied by major cardiac defects. We investigated the co-occurrence of different types of congenital cardiovascular defects (CCVD) in children with IM.
Methods: Retrospective study of patients born between September 1980 and September 2005, registered for IM and CCVD at a single university medical center. Records were reviewed for major and minor CCVD, age of CCVD and IM diagnoses, other congenital abnormalities, syndromes or associations, morbidity and mortality.
Results: In a total of 284 patients with IM, 78 were also diagnosed with CCVD (27.5%). In this study-group, major heart defects were diagnosed in 37 (47.4%) patients, e.g., HLHS, TOF, left or right isomerism, AVSD, CAT, or TAPVC. Forty-one patients (52.6%) had minor CCVD, e.g., ASD, (sub)valvar aortic stenosis or isolated VSD. Median age at time of IM diagnosis was 10 days (range 0–1357). Major and minor CCVD's were diagnosed at median of 4 (0–98) and 7 (0–210) days after birth, respectively. Other congenital defects included: diaphragmatic hernia (7/78), omphalocele (7/78), anorectal malformation (6/78), esophageal atresia (5/78) and heterotaxia (9/78).

Forty-seven (60.3%) patients were not diagnosed with any known syndrome or association; others had Down syndrome (9.0%), VACTERL (6.4%), Ivemark (6.4%), and several other syndromes were seen once. Major CCVD's were more frequently seen in patients with a syndrome than minor CCVD's (58.1% vs. 41.9%).

Forty-five patients (57.7%) underwent elective IM surgery. Fifteen patients (19.2%) died at a median age of 46 (range 3–3010) days, due to cardiovascular complications (n = 7), extensive intestinal necrosis (n = 2), sepsis (n = 2), unexplained death at home (n = 1), respiratory failure (n = 1) and withdrawal of medical care (n = 2). The morbidity was high, including ileus and re-operations because of IM complications, of which 38.5% (5/13) died thereafter.
Conclusions: Minor and major CCVD's are frequently diagnosed in children with IM. Most patients have other congenital abnormalities too. Surgical intervention in these patients with IM should be carefully considered, since morbidity and mortality are higher than reported in the literature.

P-88

Complex cardiological examination after liver transplantation

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Tacrolimus (Tac) and cyclosporine A (CsA), the major immunosuppressant drugs used in patients (pts) after liver transplantation (LTx), may cause various side-effects such as arterial hypertension (AH) and left ventricular hypertrophy (LVH).

Study evaluate results of complex cardiological exams after LTx in 100 pts aged 9 mths-22 yrs, treated with Tac (78 pts, during mean 1,4 yrs) or CsA (22 pts, mean 5,7 yrs). Ecg, Holter ecg and echo were performed in all pts post LTx. LV mass index (LVMI) was calculated by echo (g/BSA, g/height2.7). Post LTx LV ejection fraction (LVEF) was estimated by radionuclide angiography (87 pts), LV myocardial perfusion by 99 mTcMIBI SPECT (53 pts).
Before LTx: 38/100 pts had echo, in 6/38 pts LVH (LVMI above 38,6 g/m2.7) was found both prior and after LTx.
After LTx: In ecg LVH had 22 out of 100 pts, ST-LV changes 8. Decreased LVEF had 6 pts. In echo: mean LVMI was 78,8 g/m2 (41,5 g/m2.7), LVH had 44 pts (15 pts had significant LVH: LVMI above 51 g/m2.7). There was no difference in mean LVMI between children on CsA/Tac, with/without AH; 23/26 pts with AH received antihypertensive drugs. Longer mean QTd time in ecg had pts with decreased LVEF (36 vs 22 ms, $p = 0,04$), LVH in ecg (29 vs 21 ms, $p = 0,03$) or in echo (31 vs 22 ms, $p = 0,03$). Arrhythmias had 11 pts, more frequently pts with ST changes (25 vs 9,8%, $p = 0,04$). LAHB had 13 pts, more frequently with significant LVH (26 vs 9,8%, $p = 0,02$). Perfusion defects in SPECT had 19 pts, especially with ST changes (83 vs 30%, $p = 0,019$). Lower LVEF had pts with abnormal SPECT (51 vs 62%, $p < 0,05$), late graft rejection (28 vs 5%, $p = 0,07$). Post LTx LVH in echo had pts with higher (>10 ng/ml) Tac blood levels (77 vs 43%, $p = 0,007$), post LTx follow-up below 6 months (71 vs 44%, $p = 0,045$). Higher post-LTx LVMI had pts with ST changes (110 vs 76 g/m2, $p = 0,01$), Tac levels above 10 ng/ml (46 vs 41 g/m2.7, $p = 0,06$), LTx at age above 2 yrs (91 vs 57 g/m2, $p = 0,001$) (43,7 vs 27,9 g/m2.7, $p = 0,05$). Higher blood levels of Tac (>10 ng/ml) had pts examined during first 6 months after LTx.
Conclusions: In patients after LTx long-term immunosuppression may cause various cardiovascular side-effects and the most common abnormality is LVH. Ecg and echocardiogram are proposed to be a part of the routine pre and post liver transplant examination. After LTx early tacrolimus blood concentration reduction may be beneficial regarding LVH.

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P-90

Assessment of the need for elective balloon atrial septostomy in patients with tricuspid atresia: a 25 year experience at the yorkshire heart centre

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Introduction: Significant variation exists in the indications for balloon atrial septostomy in children with tricuspid atresia (TA). Many units advocate early, elective balloon atrial septostomy due to potential progressive narrowing of the interatrial opening, especially after the insertion of an arterial shunt. Our practice is to perform an atrial septostomy only in children with clinical and/or echocardiographic features of a restrictive interatrial communication.

The aim of the study was to assess our practice by retrospective evaluation of patients born with tricuspid atresia, with particular reference to the clinical fate of the atrial septum.
Materials and Methods: The study was a retrospective review of the data on 54 live-born patients in our departmental database with a diagnosis of TA. 2 patients were excluded from the study. By reviewing the notes of the remaining 52 patients we collected data

on indications, and timing of balloon atrial septostomy, subsequent complications and long term outcome.
Results: The median age to follow up was 11.3 years (0.7–23.7). 5 patients required balloon atrial septostomy for a clinically and/or echocardiographically restrictive atrial septum. 3 procedures were performed within the first 24 hrs of life and 2 within the first month. One of the patients developed NEC following the septostomy and subsequently died of overwhelming sepsis. 1 patient required delayed surgical septectomy at 6 years of age, during a bidirectional Glenn operation.

Of the remaining 46 patients there were 9 deaths but none were attributed to a restrictive atrial septum. 27 patients required palliation with an arterial shunt. None of the patients developed a restrictive atrial septum post procedure or during long term follow-up.
Conclusion: We conclude that elective balloon atrial septostomy at presentation is not necessary in patients born with tricuspid atresia. Accepting the limitations of retrospective data collection we also conclude that progressive narrowing of the interatrial septum at a later date is an unusual occurrence, even after an arterial shunt operation.

We recommend that atrial septostomy should only be performed in patients who show obvious clinical and/or echocardiographic evidence of restrictive interatrial communication.

P-91
Exercise performance in patients with pulmonary atresia and intact ventricular septum (PA-IVS)
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Objective: The aim was to explore the exercise performance in patients with PA-IVS, compared to healthy controls. We also compared the results between patients with biventricular repair and those with univentricular heart palliation.
Methods: From 1980–1995, 66 children with PA-IVS were born in Sweden. Thirty-nine of these patients are alive and thirty-two were offered to participate in the study. Twenty-five completed cardiopulmonary exercise performance on Sensor Medics bicycle ergometer. Peak oxygen consumption (VO_2peak) was measured using the breath to breath method. We examined fifteen (4 males/11females) healthy controls (age 6–20, median 12 years).
Results: Seven male patients (age 9–20, median 16 years) with biventricular repair had a VO_2peak of median 41.8 ml/kg/min (range 25.9–43.6), compared to seven patients (age 10–15, median 14 years) with univentricular palliation having a VO_2peak of median 38 ml/kg/min (range 20.2–46.3) (n.s). The VO_2peak in the male control group was median 41 ml/kg/min (range 34.5–46.8) (n.s).

Eight female patients (age 9–24, median 12.5 years) with biventricular repair had a VO_2peak of median 36.8 ml/kg/min (range 22.6–46.9), compared to three patients (age 12–22, median 12 years) with univentricular palliation having a VO_2peak of median 29.5 ml/kg/min (range 21.6–29.8). The VO_2peak in the female control group was median 32 ml/kg/min (range 29.6–40.9) (n.s versus patients with biventricular repair).

Six male patients with ventriculocoronary artery connections VCAC (age 10–20, median 12 years) had a VO_2peak of median 33 ml/kg/min (range 20.2–39.8) compared to 40.9 ml/kg/min (range 36–46.3) in eight patients without VCAC (age 9–19, median 14). As only two female patients had VCAC they were not included in the analysis.
Conclusions: In this cohort of patients with PA-IVS exercise performance was similar to that of healthy controls. In addition we found no significant difference between patients with biventricular repair and those with univentricular heart palliation. Male patients with ventriculocoronary artery connections had lower exercise capacity than that of the control group.

P-92
Efficacy of angiotensin converting enzyme (ACE) inhibitors in pediatric patients with mid-to-severe aortic regurgitation
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Background: Aortic regurgitation may cause left ventricular dilatation due to volume overload. The systemic afterload reduction by ACE inhibitors theoretically decreases volume overload. There is little information about the efficacy of ACE inhibitors in pediatric patients with aortic regurgitation. Therefore, the aim of this study was to analyse the effect of ACE inhibitors in pediatric patients on volume overloaded left ventricles due to aortic regurgitation.
Methods: Clinical and echocardiographic data were retrospectively analysed in 24 patients with hemodynamically isolated mid-to-severe aortic regurgitation. ACE inhibitor therapy was established in 19 patients while 5 patients received no therapy due to various reasons. Median (quartiles) age at start of ACE inhibitor was 8.7 (5.3 to 10.0) years, follow-up under ACE inhibitor was 2.4 (0.9 to 5.5) years. As ACE inhibitors captopril was given at a dose of 1 mg/kg/die for infants and enalapril at a dose of 5–10 mg/m^2/die for children. Left ventricular enddiastolic diameter (LVEDD), shortening fraction (SF), left ventricular posterior wall diameter (LVPWD), and grade of aortic regurgitation were measured echocardiographically. To adjust for body surface, measurements were expressed as differences from body-surface-adjusted normal values.
Results: There was no significant change of ACE inhibitor therapy in respect to reduce LVEDD (median change = −2, p = 0.26), increase SF (median change = −1, p = 0.92) or reduce LVPWD (median change = −0.05, p = 0.64). Individual regression slopes for repeated measurement on each patient (either without or during treatment) were computed to assess individual changes over time. No significant difference between untreated (n = 5) or treated (n = 19) patients was seen in changes (slopes) for LVEDD (median slope 0.14 for untreated vs. 1.1 for treated, p = 0.36), SF (median slope −0.22 for untreated vs. 0.15 for treated, p = 0.97) or LVPWD (median slope 0.17 for untreated vs. −0.08 for treated, p = 0.15).
Conclusions: No ACE effect was seen in this study in terms of left ventricular dimensions and function in pediatric patients with mid-to-severe aortic regurgitation. Due to the limited case load and retrospective study design, a prospective randomized double blind study of pediatric patients with aortic regurgitation is recommended.

P-93
Prevalence and fifteen-year trends in prehypertension, hypertension, lipid disorders and overweight among adolescents in Siberia (1989–2003)
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High blood pressure, overweight and lipid disorders are known risk factors of coronary heart disease. Control of the factors since adolescence may reduce cardiovascular morbidity and mortality in adulthood. Aim of the study was to assess trends in prevalence of overweight, lipid disorders, arterial prehypertension and hypertension among adolescent population in Novosibirsk during the reforms (1989–2003).

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Methods: Four cross-sectional surveys of representative samples of school children aged 14–17 in 1989 (656), in 1994 (620), in 1999 (626) and in 2003 (667) were carried out. Body mass index (BMI, kg/m^2), systolic (SBP) and diastolic blood pressure (SBP), serum total cholesterol (TC), high-density lipoprotein cholesterol (HDL-C) and low-density lipoprotein cholesterol (LDL-C) were measured. Diet was estimated using 24-hour dietary recall.
Results: According to the 4th Report of the NHBPEP Working Group (2004), prevalence of arterial prehypertension during the period was high in both gender groups with decreasing from 58% to 36% ($P < 0.01$) in boys and from 31% to 23% in girls; arterial hypertension also decreased from 12% to 6% in boys ($P < 0.05$) and from 16% to 5% in girls ($P < 0.05$).

According to NCEP-peds criteria (1992), prevalence of high TC (200 mg/dl and more) during the period (1989–2003) significantly decreased from 22% to 8% ($P < 0.01$) in males and from 32% to 17% ($P < 0.05$) in females. The similar trends were found in prevalence of high LDL-C (130 mg/dl and more) and low HDL-C (<40 mg/dl).

Frequencies of overweight (BMI $\geqslant 23\,kg/m^2$) decreased from 18% to 11% in boys and from 22% to 10% ($P < 0.05$) in girls. The most decreasing of the parameter was revealed in 1999.

Trends in diet during the period showed significant decreasing of total energy intakes (from 3021 to 2340 kcal in boys and from 2300 to 1640 kcal in girls) and basic nutrient intakes (proteins, fats and carbohydrates).
Conclusion: The data from Novosibirsk indicate parallel trends to decreasing in classical CVD risk factors in adolescents following the period of socioeconomic reforms in Russia.

P-94
Infective endocarditis after repair of congenital heart disease

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Objective: Surgical repair of congenital heart diseases (CHD) is supposed to prevent infective endocarditis (IE). The aim of this retrospective study was to evaluate the occurrence of IE early and late after repair in adult and pediatric patients with CHD.
Material and Methods: Of 153 IE episodes diagnosed from 1966 to 2001, 38 (25%) occurred early (<3rd month postoperative: 17 cases) or late (>3rd month postoperative: 21 cases) after surgical repair; 9 were adults. Correction of CHD consisted in the following: VSD patch closure (4), regular Tetralogy of Fallot (4), repair + valvular prosthesis or pace maker (14), repair + residual high velocity turbulence (8: residual VSD, aortic or mitral valvulopathy, mild isthmic stenosis), Rastelli conduit (5) and repair + residual low velocity turbulences (Senning, Switch operation, ASD + pulmonary artery patch).
Results: Early post-repair episodes (17 cases) were more frequent in children (16/29 = 55%) than in adults (1/9 = 11%) and were: 4/4 VSD patch, 1/3 Fallot, 7/14 prosthesis or pace maker, 3/8 high velocity residual lesions, 2/5 Rastelli. The remaining 21 late episodes occurred from dental (7), ENT (5), cutaneous (4) or other infections (5), and included: 3/4 Fallot, 7/14 prosthesis or pace maker, 5/8 high velocity residual lesions, 3/5 Rastelli and 3/3 residual low-velocity lesions. Staphylococcus was commoner in early postoperative and cutaneous cases, streptococcus in dental and ENT episodes. Surgery as a treatment of IE was required in 9 patients (24%). Six deaths occurred (16%); mortality was higher in cases observed in the early postrepair period (29%) than in the late follow-up (19%). Six-month survival was 85% (74% in early post-repair cases and 95% in late cases, $p = 0.1$).
Conclusion: Infective endocarditis can occur early and late after complete repair of CHD, because of prosthetic material and/or high or low-velocity residual lesions. The absence of any residual lesion only can suppress the risk of late but not early postrepair IE. Prophylaxis should be recommended despite previous repair, with special attention toward dental, ENT and cutaneous sources of infection.

P-95
Clinically important differences in accuracy of different pulse oximeters in the assessment of children with cyanotic heart disease

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Background: Movement artefact and poor peripheral perfusion are well-known sources of difficulties in obtaining satisfactory pulse-oximetry (Pox) readings with conventional pulse oxymeters (C-Pox). It is also well known that saturation measurement in cyanotic patient is less accurate in cyanotic patients. New generation pulse oxymeters (Pox-SET) with signal-extraction technology can give readings in spite of movements or low perfusion. During clinical work we have observed that even during optimal measurement conditions very large differences in values were present in cyanotic children using different oxymeters. The purpose of this study was to determine if there were clinically important differences in C-Pox and Pox-SET values to saturations measured on arterial blood gases.
Methods: 44 children spontaneously breathing children with cyanotic heart defects had simultaneous measurements with C-pox, pox-SET and an arterial bloodgas.
Results: The results from blood gas-analysis included 44 children with SpO_2 <95%. Both pulse oxymeters tended to over-estimate the saturation, but the discrepancies found with C-pox were larger than those with Pox-SET ($p = 0.01$). Among patients with saturations <95% C-pox saturations deviated >10% from arterial blood gas-saturation in 15.9%, and with Pox-SET in 11%. In some cases the differences between simultaneous Pox-recordings were as large as 25%, which in patients with eg. transposition could significantly influence management. On the linear part of the haemoglobin oxygen saturation curve a regression analysis describes the correlation between saturation and arterial pO_2 with a correlation coefficient of 0.90, slope 7.31, intercept 39.52. Identical regression analysis for Pox-SET shows correlation coefficient 0.80, slope 6.77, intercept 47.34 and that for C-pox showed correlation coefficient 0.59, slope 4.7, intercept 59.75. Significant deviation from the 95th prediction limits occurs below 6 kPa with pox-SET and 6.5 kPa with C-pox. The more hypoxic child the larger the systematic error.
Conclusion: In cyanosed patients conventional pulse oxymetry is not always a reliable way of monitoring arterial oxygenation, particularly not for profoundly hypoxic patients. In patients with marked hypoxia and arterial pO_2 below 6.0–6.5 kPa pulse oximetry monitoring systematically overestimates true saturation levels. Underestimation of the levels of cyanose may leed to failure to implement needed therapeutic measures such as septostomy.

P-96
Clinical characteristics of Kawasaki disease in patients under 6 months of age

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Introduction: Kawasaki disease is acute febrile vasculitis and the most common cause of acquired heart disease in children. More than 80% of patients with Kawasaki disease occur in age group under 4 years of age. Incomplete Kawasaki disease is known to be more frequent in young infant, which makes a difficulty in early diagnosis and treatment. We analyzed the clinical characteristics of Kawasaki disease in patients younger than 6 months in order to make early diagnosis and to decrease coronary complications.
Methods: Total 410 patients were diagnosed as Kawasaki disease in our hospital between March 1995 and July 2005. We reviewed the medical records of 30 patients of study group (under 6 months) and 54 patients of control group (13–99 months).
Results: Mean age was 3.9 ± 1.4 months (1–6 months range) in study group and 34.2 ± 18.8 months in control group. Incomplete Kawasaki disease was more frequent in study group (4; 13% vs. 2; 4%) but not statistically significant. Less frequent cervical lymphadenopathy (15; 50% vs. 40;74%) and more frequent BCG site erythema in study group (18; 60% vs. 2; 4%) were statistically significant. Male to female ratio (1.7:1), fever duration, conjunctival injection, oral changes, skin rashes, extremity changes were not statistically significant. WBC and platelet count were significantly higher in study group. Coronary arterial dilatation was more common in study group but not statistically significant.
Conclusions: This study shows that BCG site erythema was more frequent and cervical lymphadenopathy was less frequent in younger than 6 months. WBC and platelet count were higher in this age group. These findings may be of help in early diagnosis and treatment in this age group.

P-97

Dural ectasia in children with Marfan syndrome – a prospective, multicenter, patient-control study

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Objectives: The clinical diagnosis of Marfan syndrome in childhood is difficult, because symptoms may not have developed to their full expression until adulthood. The Ghent nosology for the diagnosis of Marfan syndrome classifies dural ectasia as a major diagnostic criterion. More than two thirds of adult patients with Marfan syndrome show dural ectasia, while the frequency in childhood is unknown. This prospective multicenter observational patient-control study was performed to identify pathologic changes of the lumbosacral spine in young patients with Marfan syndrome.
Methods: Design: Prospective clinical trial, multicentric, cross sectional.
Setting: MRI of the lumbosacral spine.
Patients: 20 patients with proven Marfan syndrome, 20 patients suspicious for Marfan syndrome and 38 healthy controls.
Outcome measures: vertebral body diameter (VBD) from L1 to S1, dural sac diameter (DSD) from L1 to S1, dural sac ratio (DSR), qualitative assessment of the lumbosacral spine.
Results: DSD and VBD in different age groups were higher in patients with proven or suspected Marfan syndrome than in healthy controls (DSD: L1, 6 to 8 years, $p < 0.05$). VBD related to body height showed a similar growth related increase in patients with proven or suspected Marfan syndrome and controls. DSD related to body height was elevated in patients with proven or suspected Marfan syndrome at different levels of the lumbar spine. DSD at levels L1, L5 and S1, and DSR at levels L5 and S1 of patients with proven Marfan syndrome were significantly higher ($p < 0.05$) than in controls.
Conclusions: Even during childhood pathologic changes inside the lumbosacral spine of patients with Marfan syndrome can be observed. Dural ectasia, which occurs at different levels of the lumbar spine, can be detected at levels L5 and S1 in up to 40% of patients with Marfan syndrome.

P-98

The valvular aortic stenosis diagnosed in neonatal period – clinical course and the prognosis

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The aim of the Study:

1) To evaluate the clinical picture and to analyze the results of laboratory tests as well as the kind and effect of performed invasive treatment in newborns with aortic stenosis (AS).
2) To find the prognostic factors for newborns with AS.

Materials and Methods: The study group was composed of 30 newborns with AS recognized in neonatal period hospitalizes in the Department of Pediatric Cardiology between 1986–2005. The group was divided into 2 subgroups – the newborns who presented symptoms of heart failure (HF) and the newborns who did not present the symptoms of HF at the recognition. The physical examination, electrocardiography (ECG), chest X-ray (RTG), echocardiography (ECHO) was performed in both groups. Based on available medical documentation the data concerning invasive treatment in the study group were collected. The results were compared between the groups.
Results: 74% of newborns with AS presented the symptoms of HF. ECHO revealed higher systolic gradient between left ventricle and aorta (LV-Ao) in the group of newborns with AS and HF and normal LV systolic function (89.5 mmHg vs 50.8 mmHg). There was not good correlation between LV-Ao gradient and AS severity in neonates with AS and LV systolic dysfunction. The balloon valvuloplasty (BVP) was performed in 95% of newborns with AS and HF and in 37% newborns with AS with no HF. In postneonatal period the surgical treatment was necessary in 30% patients with HF at average age 5.5 month and in 50% with no HF; average age 24 month. 7 patients died during follow up period, including 6 from the group with HF at the recognition.
Conclusions:

1. The majority of newborns with severe AS diagnosed after birth present symptoms of HF.
2. The systolic gradient LV-Ao is not objective parameter of AS severity in neonates with AS and LV systolic dysfunction.
3. In children after invasive treatment of AS in neonatal period, the surgical treatment is necessary sooner in the group of patients who presented HF at recognition than in patients without HF.
4. The poor prognostic factor in newborns with AS is HF at the recognition.

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P-99

Long term results of 69 cases of valvular bioprotheses implanted in children at the institut de cardiologie d'abidjan (Côte d'Ivoire)

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Introduction: The choice of valvular protheses remains problematic in developing countries since it is difficult and expensive to follow the patients after the operation.

This difficulty is even more evident in subsaharian part of Africa as the rheumatismal valvulopathies are frequent and severe for children from low-income families.

Objective: The objective of the study was to evaluate the long-term behaviour of 69 pig protheses implanted from 1978 to 1995 in Abidjan into 68 adolescents below 20 years and followed between 1978 and 2004.

Material and Methods: Sixty-nine pig protheses were implanted into 68 patients (40 females and 28 males).

The mean age was 11 years with extremes at 4 and 17 years.

Fourty-four protheses (63.8%) were of Angel Shiley type, 19 (27.5%) of Carpentier Edwards type and 6 (8.7%) were Hancok protheses.

Fourty-nine (72.05%) were mitral position, 18 (26.5%) tricuspid and 2 (3%) were in mitrotricuspid position.

Sixty patients (88.2%) were followed up; 8 (11.8%) died directly after the operation.

Fourty-two patients (70%) of the 60 survivors have been regularly followed during the whole period. 18 patients (30%) did no longer show up after 3 years.

The follow-up included ECG, chest X-ray and cardiac Doppler echocardiography.

Results: The 42 protheses (100%) failed: 15 (36.6%) before 5 years and 26 (63.4%) between 5 and 10 years.

The mean age of prothesis failure was 4.8 years.

All 42 patients were reoperated after 5 to 10 years and were implanted mechanical protheses in mitral position or Carpentier-Edwards bioprotheses in tricuspid position.

Conclusion: The most significant advantage of valvular heterografts is that you do not need coagulation tests. But they result in severe complications such as calcifications and degeneration. This fact has led us to adopt a new policy which is that since 2000 heterografts are only used in tricuspid position.

P-100

Reduced quality of life and physical activity in obese children

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Introduction: Childhood obesity is associated with poor quality of life, due to reduced psychosocial health, self-esteem and physical functioning. Physical activity is a major determinant of cardiovascular health and may also improve components of quality of life in this population. The aim of this study was to assess health-related quality of life and physical activity in obese children.

Methods: This was a cross-sectional including 2 groups: 41 obese prepubertal children and 41 non-obese controls aged 6 to 11 years (9.0 ± 1.5 years). Both groups were matched for age, height and pubertal stage. The health-related quality of life was assessed by the Child Health Questionnaire, a parent report tool that provides information on children's social roles, emotional health, physical health and family functioning. Other measures included: 7-day physical activity count by Actigraph MTI accelerometer; past 12 months physical activity level by the Modifiable Activity Questionnaire for Adolescents; body composition by dual-energy x-ray absorptiometry and anthropometric measures.

Results: Health-related quality of life total score (73.0 ± 10.9 vs 82.4 ± 7.3, $p = 0.001$) was significantly decreased in obese children compared to controls, specifically subscales related to psychosocial health, self-esteem, physical functioning, and impact on parental emotional well-being. Obese children had lower 7-day activity count (309.3 ± 42.4 vs 394.4 ± 99.5 cpm, $p = 0.04$) and past 12 months physical activity (0.8 ± 1.3 vs 3.9 ± 3.2 hours/week, $p = 0.001$) than controls, whereas body mass index (25.2 ± 4.8 vs 15.8 ± 1.5 U, $p = 0.001$) and percentage of body fat (42.5 ± 7.4 vs 19.4 ± 6.6%, $p = 0.001$) were significantly increased.

Conclusion: Our study demonstrates that health-related quality of life is reduced in prepubertal obese children compared to non-obese children, particularly psychosocial health and physical functioning. In addition, their physical activity level is lower than controls, suggesting decreased sports participation and physical conditioning. We conclude that obese children may benefit from adapted physical activity interventions to improve health-related quality of life and prevent cardiovascular complications associated with body fatness.

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P-101

Vasoactive intestinal peptide (VIP) is diminished in pediatric patients after Fontan procedure

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Background: The Fontan circulation is correlated with a loss of cardiac pulsatility in the pulmonary vascular bed, pulsatile flow however is important for maintaining a low resistance in the pulmonary vasculature. Plasma cytokines seem to exert an important effect on postoperative Fontan hemodynamic status, recent studies have shown abnormal values of vasoactive substances (ANP, BNP, angiotensin II, norepinephrine, adrenomedullin).

VIP is a neuropeptide with potent vasodilatory effects. It was originally isolated from intestinal extracts, where it induces smooth muscle relaxation and it was shown to be significantly reduced in patients with pulmonary hypertension.

The aim of our study was to evaluate VIP levels in patients with Fontan circulation. We hypothesized that the unique low flow in the central veins and the pulmonary system may have an influence on VIP levels.

Patients: Up to now 8 patients (6 with extracardiac and 2 with intracardiac Fontan) aged from 8 to 16 years (median: 13.5y) were investigated. Their median oxygen saturation was 93% and median time after operation was 4.6 years. All but one patient were anticoagulated with phenprocoumon.

Methods: VIP serum concentration was determined by radioimmunoassay (RIA), with a detection threshold at 5 pg/ml.

Results: All patients had reduced VIP levels with 6 of them showing levels below the detection threshold of 5 pg/ml, in the rem-aining two patients VIP levels were in the low normal range (18 pg/ml and 13 pg/ml, respectively) (VIP normal range:10–60 pg/ml).

Conclusion: Our preliminary results show that Plasma VIP levels are diminished in patients with Fontan circulation. VIP may have an

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important role in the pathogenesis of elevated pulmonary vascular resistance late after Fontan procedure.

It remains unclear whether the low VIP levels in Fontan patients are due to the low flow in the pulmonary circulation or to the altered flow in the splanchnicus territory.

To our knowledge this is the first report on VIP in patients after Fontan procedure.

(For P-102, please see OP-9)

(For P-103, please see OP-8)

P-104
Vascular endothelial dysfunction and oxidative stress late after the onset of Kawasaki Disease

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There have been some important reports that some endothelial damage might present even in the case of late after Kawasaki Disease (KD) patients, but clinical evidence or the mechanism for causing the endothelium dysfunction of systemic arteries has not been fully understood. We investigated the vascular endothelial dysfunction in KD patients, from the viewpoints of the correlations with oxidative stress.

The study group consisted of 33 KD patients [Group A: 11 patients with coronary artery lesions, Group B: 5 patients with transient dilatation, and Group C: 17 patients without coronary artery lesions] in adolescence and adulthood (mean age, 25.6 years) and 28 age- and sex-matched healthy subjects without any risk factors for vascular endothelial damage. We measured von Willebrand factor, t PA-PAI-1, TAT, urine nitrites and nitrates (NOx;μM/creatinine), and flow mediated endothelium-dependent vasodilatation (%FMD) to assess the vascular endothelial function. Urine 8-isoprostane (pg/ml · creatinine) and 8OHdG (ng/ml · creatinine) were also evaluated as an oxidative stress marker. [Results] Significant increase in TAT was noted in KD group. %FMDs in the KD group were significantly less than those in the control group (11.2 ± 3.4 to 15.1 ± 3.0, $p < 0.01$), and %FMD in 8 patients (24%) of KD patients were less than 2SD of the mean values in control. The increases in 8-isoprostane and 8OHdG values were remarkable in KD patients, but there was no significant correlation between oxidative stress markers and %FMD. *Conclusions:* This study revealed that vascular endothelial dysfunction could exist in the KD patients with CAL and without CAL, even in the chronic stage. It was also suggested that the KD patients were under oxidative stress. Further study is inquired to confirm any correlation between the endothelial dysfunction and oxidative stress.

P-105
Involvement, education and activities of nurse specialists in adult congenital heart disease programmes in Europe

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Introduction: Children with congenital heart disease are often transitioned to adult-centred health care when they grow up. The increasing number of adults with congenital heart disease (ACHD) has prompted the development of recommendations for the management of these patients and for the organisation of their healthcare. One of the recommendations dictate that specialist centres should employ nurse specialists who are trained and educated in ACHD care. In this study, we surveyed the involvement, education and activities of nurse specialists in the care for ACHD in Europe.
Methods: As part of the Euro Heart Survey on ACHD, data on delivery of care from 48 specialist centres throughout Europe was obtained, using a 20-item web-based survey tool. An additional survey tool was designed, comprising 15 items, covering issues such as the extent of involvement of nurse specialists, the activities undertaken, education, involvement in research, collaboration with other health care professionals, and financing of the position.
Results: Twenty out of 48 specialist centres (42%) have nurse specialists affiliated with their ACHD programme. These centres had a median number of 2 nurse specialists on staff, corresponding with 1 full-time equivalent. The involvement of nurse specialists was not related to the caseload of inpatients and outpatient visits. Most nurse specialists had a Bachelor/Polytechnic degree (53.4%) as highest earned degree, followed by in-hospital nursing training (33.3%) and Masters degree (13.3%). Physical examination was the most prevalent activity undertaken by nurse specialists (93.3%), followed by telephone accessibility (86.7%), and patient education (86.7%). Patient education covered mainly prevention of endocarditis (100%), cardiovascular risk factors (92.3%), sport activities (92.3%), the type and characteristics of the heart defect (92.3%), the definition and aetiology of endocarditis (84.6%), cardiac risk in case of pregnancy (84.6%), and heredity (84.6%). Insurability (38.5%) and nonadherence with the prescribed regimen or lifestyle (30.8%) were the least addressed issues.
Conclusion: This survey revealed that less than half of the centres have employed nurse specialists in ACHD. There is room for improvement with respect to their involvement, education, and activities.

P-106
Cardiopulmonary exercise test for the evaluation of exercise capacity of adults after surgical repair of coarctation of the aorta related to residual stenosis of the descending aorta and arterial hypertension

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Introduction: Despite effective surgical repair of coarctation of the aorta (CoA) range of patients (P) presents arterial hypertension (AH) and early onset coronary artery disease which may result in heart failure.

The aim of the study was to evaluate exercise capacity in adults after surgical repair of CoA with cardiopulmonary exercise test and to determine their relationship with AH and residual descending aortic stenosis.
Material and Methods: We studied 74 P (45M) aged 19–61 years (mean 31.2 ± 9.8 years) operated on at the age of 0.5–34 years (mean 10.4 ± 6.8 years) which was 5–34 (mean 21.4 ± 6.2) years ago. All P-NYHA class I. Controls: 30 subjects (18M) aged 26–46 years (mean 32.2 ± 6.6 years). Echocardiography was used for evaluation of severity of stenosis of the descending aorta (significant $\geq$25 mmHg). The groups with (AoD+, n = 32) and without residual stenosis (AoD−, n = 41) and without (AH−, n = 32) and with exercise induced arterial hypertension (HTex, n = 10) and persistent arterial hypertension (HT+, n = 32) were selected.

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Maximal exercise treadmill test was performed acc. to modified Bruce's protocol: forced vital capacity (FVC), forced expiratory volume in 1 second (FEV1), peak oxygen consumption (maxVO_2), and VE/VCO_2, heart rate at peak exercise (HR max) were analyzed:.
Results: Comparison of control and study group showed: maxVO_2 (mL/kg/min): 26.77 ± 6.75 vs. 35.27 ± 7.6 (p = 0.0001); maxVO_2%: 70.09 ± 13.39 vs. 90.3 ± 15.35 (p = 0.0001); VE/VCO_2: 33.14 ± 5.33 vs. 29.73 ± 3.32 (p = 0.001); HRmax: 167.91 ± 21.25 vs. 180.83 ± 12.93 bpm (p = 0.0001), FVC (L): (4.12 ± 0.95 vs. 8.03 ± 0.53 p = 0.02), FEV L(L): (3.30 ± 0.80 vs. 3.76 ± 0.86 p = 0.02).

No differences were observed between AoD+ and AoD− group with respect to cardiopulmonary parameters.

Comparison of AH+ and AH− group: VO_2max (mL/kg/min): (26.30 ± 15.08 vs. 28.05 ± 7.9, p = 0.01); VO_2max%: (66.17 ± 14.58 vs. 74.75 ± 12.16, p = 0.02); VE/VCO_2: (34.06 ± 4.78 vs. 28.31 ± 4.56, p = 0.003); HRmax (bpm): (162.46 ± 21.76 vs. 181.00 ± 13.05, p = 0.0001). Comparison of AH+ and AH- groups: VE/VCO_2: (33.89 ± 5.02 vs 28.31 ± 4.56, p = 0.01); HRmax (171.24 ± 15.76 vs. 181.00 ± 13.05, p = 0.01). There were no differences in respect to spirometry parameters.
Conclusions:

1. Exercise capacity of adults after surgical repair of CoAo is reduced.
2. This reduction is more severe in patients with persistent and exercise induced arterial hypertension.
3. Significant residual stenosis in the AoD not affect physical performance of these patients.

P-107
"Structural and electrical reverse remodeling after transcatheter closure of atrial septal defects in adults"

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Objectives: To determine the effects of transcatheter closure of atrial septal defects (ASD) on structural and electrical remodeling during short and long term follow-up.
Methods: Thirty-seven consecutive patients after successful device closure of ASDs were included into this analysis. Patients (mean age: 40 ± 17 years, male 24%), were assessed by transthoracic echocardiography and by means of standard 12-lead electrocardiography (ECG). Data were obtained before the intervention, at 1 month and 12-month follow-up. The following echocardiographic and ECG parameters were collected using conventional measurements: right ventricular dimensions (M-mode, 2D echocardiography: end-diastole diameters), right atrial area, left atrial area, right ventricular/left ventricular ratio; and the pulmonary arterial pressure; maximal duration of the P wave and QT interval, P dispersion and QT dispersion, the amplitude of second R′ in lead V1.
Results: At one-month and one year follow-up significant reduction were observed in the following parameters: right atrial area, right ventricular dimension (M-mode measurements), right ventricular/left ventricular ratio, P max, QT max and R′. Pulmonary arterial pressure decreased significantly at the early follow-up. There was no significant change in P dispersion and left atrial area during follow-up. Six patients (16%) had documented paroxysmal atrial tachyarrhythmias before the procedure and four of them became arrhythmia-free during the follow-up. Only atrial dimensions were different between patients with and without arrhythmias.
Conclusions:

1. Structural and electrical reverse remodeling starts early after defect closure and continues at one year follow-up.
2. Pulmonary arterial pressure decreases promptly after the procedure, and remains unchanged during the follow up period.
3. Our data suggest that structural reverse remodeling may play a role in the reduction of tachyarrhythmia recurrences.

P-108
Atrial fibrillation and impaired left ventricular function are risk factors for atrial septal defect closure in patients over 30 years of age

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Introduction: Today percutaneous or surgical closure of secundum atrial septal defects (ASD II) in adults is considered an effective and safe treatment. However, left ventricular dysfunction, may lead to a prolonged intensive care unit stay, morbidity or even to an early death.
Methods: Records of 281 adult patients who underwent surgical closure of an ASD II between 1974 and 2001 at an age over 30 years were reviewed retrospectively for demographic, hemodynamic, and anatomic predictors and outcomes. Median age at operation was 42.9 years (range from 30 to 76 years).
Results: Patients with atrial fibrillation had a significantly longer stay at the intensive care unit (p < 0.001), and needed significantly more inotropic support postoperatively (p = 0.007). Patients with impaired left ventricular function also had a significantly longer stay at the intensive care unit (low cardiac index, p = 0.037), and needed significantly more diuretics postoperatively (low cardiac index, p = 0.011; low left ventricular ejection fraction, p = 0.019). Patients who presented in NYHA functional class II or more needed significantly more inotropic support postoperatively (p = 0.043). Thirty days mortality rate was 0.7% (2 patients). One 36-year-old patient died intraoperatively due to low cardiac output, the other 31-year-old patient died at the 6th postoperative day due to low cardiac output. Both patients presented in NYHA functional class III preoperatively.
Conclusions: Surgical ASD closure in adults is usually safe and effective. However a small subgroup of patients with left ventricular dysfunction is at risk for significant morbidity and even mortality. We found atrial fibrillation and impaired left ventricular function to be risk factors for increased morbidity.

P-109
Conversion of atriopulmonary Fontan to extracardiac total cavopulmonary connection improves cardiopulmonary function

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Background: Experimental studies showed that extra-cardiac total cavo-pulmonary connection provides superior hemodynamics than atrio-pulmonary Fontan.
Methods: We prospectively assessed the impact of conversion of atrio-pulmonary Fontan to extra-cardiac total cavo-pulmonary connection on exercise capacity and cardiac function in 6 consecutive patients.

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Results: Six months after conversion to extra-cardiac total cavo-pulmonary connection, we observed an increase in peak oxygen uptake in all patients ($p = 0.01$; +17%). This improvement was associated to an increase of peak O_2 pulse ($p = 0.01$; +16%), but no change in peak heart rate, arterial oxygen saturation at peak exercise, and pulmonary function. Ventricular ejection fraction did not change significantly after surgery. Conversion was associated with an improvement in heart failure symptoms as assessed by the New York Heart Association classification. Patients who had undergone additional anti-arrhythmia surgery for atrial fibrillation had no recurrence of arrhythmia at follow-up.
Conclusion: Data indicates that conversion to extra-cardiac total cavo-pulmonary connection is associated with an improvement of cardiopulmonary function and heart failure symptoms. Improved exercise capacity is due to an increase in O_2 pulse and may reflect an improved cardiac stroke volume after the operation.

P-110
B-Type natiuretic peptide (BNP) correlates to clinical status and fractional shortening in paediatric heart failure
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Background: Paediatric heart failure is difficult to assess particularly as echocardiographic findings do not always correlate to symptoms. We investigated BNP in the setting of a paediatric heart failure and transplantation service to assess its usefulness as a clinical marker of heart failure.
Method: Clinical and echocardiographic data were correlated to 126 BNP samples. Patients were 3 weeks to 16 years of age. Clinical status was defined using Ross and NYHA scores. 34 samples were from patients with normal ventricular function. Of those with poor ventricular function majority of samples were from patients with idiopathic dilated cardiomyopathy (38), anthracycline cardiotoxicity (15), congenital heart disease (25), viral myocarditis (6) and restrictive cardiomyopathy (6).
Analysis: BNP was correlated to parametric data (fractional shortening (FS%)) with pearsons correlation coefficient. For non-parametric data (NYHA and Ross score), spearmans correlation coefficient was used.
Results: Rising BNP levels correlate to deteriorating clinical status with significance to the 0.01 level (Ross and NYHA). There was also correlation, significant to the 0.01 level to FS%. Mean BNP: in ventricular dysfunction 634 pg/ml (S.E.M. = 80), in normal function 11.9 pg/ml (S.E.M. = 1.6).
Conclusions: There is a strong correlation between BNP and clinical status as defined by the NYHA and ROSS scoring. BNP also correlates well to left ventricular systolic function as estimated by FS%. This suggests a useful role in assessment of children with heart failure.

P-111
Stiffness of the abdominal aorta in Beta-thalassemia major patients in relation with body iron load
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Objectives: Increased iron stores have been implicated in the association with increased risk of cardiovascular events. We evaluated whether abdominal aortic stiffness was altered in patients with beta-thalassemia major in relation with body iron load.
Methods: Sixty-two (32 males and 30 females) ß thalassaemia major patients aged 16,47 ± 4,8 years were enrolled into study. Healthy 52 subjects matched for age, gender recruited as controls. In all subjects, hemoglobin, fasting glucose, cholesterol, HDL-cholesterol, and LDL-cholesterol levels were measured. The average serum ferritin level and liver iron concentration (LIC) were assessed in thalassemia patients. Left ventricular function and mass were evaluated echocardiographically. Aortic strain (S), pressure strain elastic modulus (Ep), and normalized Ep (Ep*), aortic distensibility (DIS) and beta stiffness index (ß index) were calculated in all subjects.
Results: There was not statistically significant difference between the study and control groups in gender, mean age, body mass index, heart rate, systolic and diastolic blood pressure. However, pulse pressure and left ventricular mass index were found higher in thalassaemia major patients compared to the control group. In thalassaemia major patients S and DIS were significantly lower compared to the control group. However, Ep, Ep* and β index were significantly higher in thalassaemia patients than controls. There was a statistically significant negative correlation between LIC and S, DIS. There was also negative correlation between LVMI and S. However, there was a statistically significant positive correlation between LIC and Ep, Ep*.
Conclusions: Increased abdominal aortic stiffness was detected in beta thalassaemia major patients and this increase in arterial stiffness correlated with liver iron concentration and left ventricular mass index. The alteration in mechanical properties of abdominal aorta may be another factor responsible for heart failure in thalassaemia patients.

P-112
Restrictive cardiomyopathies in children: a series of 33 patients
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Restrictives cardiomyopathies (RCM) account for 2 to 3% of cardiomyopathies in children. The prognosis is considered poor and their causes unknown.
Patients: Over a period of 22 years, we found in our data base 33 cases of apparently primitive RCM. Post-operative RCM (n = 3), endomyocardial fibrosis and endocardial fibrosis related to hypereosinophiliae (n = 3), and 1 case of RCM due to thoracic irradiation were excluded. Clinical, echocardiographic and hemodynamic characteristics as well as outcome of all patients were reviewed. For the most recent patients of the series (21/31) a complete metabolic investigation, endomyocardial biospsy and spectrophotometric study of the respiratory chain were performed.
Results: Mean age at diagnosis was 4.2 ± 3.8 years. Sex-ratio was 1.6 female/male. Family history of RCM was found in 8/33 patients (4/28 families). A known cause could be found only in 3/31 cases: 2 desminopathies and 1 defect of the complex III of the mitochondrial respiratory chain. In 8 cases, pathologic studies found a myofibrillar disarray without desmin anomaly; lipids droplets in myocytes were observed in one case in whom no metabolic defect could be identified.
During follow-up, we observed 3 strokes related to proven left atrium thrombus, 1 AV-block and 1 atrial tachycardia. Seventeen patients were listed for heart transplantation and 7 are alive at last follow-up. Actuarial survival was 58 at 1 year, 47% at 5 years and 40% at 10 years.
Conclusions: RCM in children is a rare entity of poor prognosis. The decision to transplant should be taken early and prevention

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of complications particularly stroke is mandatory. Extensive investigations have a low efficiency in identifying the causes but the high prevalence of familial forms justify first degree relatives examination.

P-113
Initial experience with levosimendan in the treatment of acute heart failure in children

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Levosimendan is a new inodilator that improves myocardial contractility by raising the sensibility of myofilaments to calcium and by promoting vasodilatation through the opening of the vascular smooth muscle's potassium channels. Multicenter studies in adults have demonstrated more efficacy when compared to conventional inotropic drugs and a better arrhytmogenic profile than dobutamine. The experience in paediatric age is scarce.

The objective is to report our experience with levosimendan in children with acute heart failure refractory to conventional treatment.

Eight patients (pts) were included, aged two months to 15 years (median: 4.5 years). Three pts were in acute phase of miocarditis; one had congenital heart disease. Two pts had dilated cardiomyopathy. Two pts were in the immediate post-operative period.

An initial dosage of 0.05 μg/kg/min during two hours, followed by a 24-hour perfusion of 0.1 μg/kg/min without a charging dose of levosimendan was used. Laboratorial, echocardiographic and continuos invasive pressure monitoring was used in all pts.

In the first child, treatment was interrupted at second hour due to sinus tachycardia and supraventricular ectopic beats. In three pts a slight increase in heart rate (HR) and a slight decrease in blood pressure (BP) were noticed. The two-month-old child had a decrease in HR and an increase of BP to levels of a better haemodynamic stability. Monomorphic ventricular ectopy occurred during perfusion and resolved spontaneously. Three pts had persistent hypokaliemia. In seven pts that accomplished the protocol an improvement in clinical and echocardiographic signs of LV function was observed (namely an increase in aortic VTI: basal 9.4 ± 1.9 cm; final 13.7 ± 1.1 cm). Four pts could be discharged from the intensive care unit in 72 hours. One child died four days after treatment with levosimendan due to severe heart failure aggravated by a respiratory infection.

In our experience levosimendan was superior to conventional treatment for acute heart failure. It reduced the length of hospitalisation in the intensive care unit. There were no major complications. Further and larger studies are needed to confirm the efficacy of this treatment in children.

P-114
Hypertension in heart and heart-lung transplanted children: does impaired baroreceptor function play a role?

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Background: Baroreceptor control of beat-to-beat blood pressure in heart and heart-lung-transplanted children is impaired. Time-related trends of baroreceptor function recovery are studied and a possible correlation of baroreflex impairment and systolic hypertension may give evidence for supplemental medical treatment of hypertension.

Methods and Patients: 17 patients (6 female) 6.1 ± 3.7 years (range 0.8–13.0 y) after heart (n = 14) and heart-lung (n = 3) transplantation (TX) were studied. 12 healthy children and 10 children after liver and bone marrow TX taking Cyclosporine A (CyA) served as control group 1 and 2, respectively. Baroreceptor sensitivity (BRS) was calculated from non-invasive systolic beat-to-beat blood pressure (sBP) measurement during a resting phase and a tilt-table test.

Results: BRS was significantly impaired in the study group at rest and during tilting; mean sBP was slightly elevated. Significant difference between patients on CyA and healthy controls was not observed. Discrete recovery of BRS occurred after four years post-TX with decreased sBP (n = 12 pts, BRS 6.78 ± 7.44 msec/mmHg, sBP 116.2 ± 12.4 mmHg) when compared to a post-TX time course of less than four years (n = 5 pts, BRS 4.02 ± 4.21 msec/mmHg, sBP 122.0 ± 6.7 mmHg, n.s.).

Conclusions: BRS is disturbed after TX in children; after four years post-TX, a minimal recovery of BRS and a discrete reduction of sBP seem to occur. Those patients with a persistent low BRS and elevated sBP may profit from pharmacological influence in sympathovagal imbalance.

P-115
Chronic respiratory complications of cardiac transplantation in children

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Introduction: Both acute and chronic respiratory problems are common following cardiac transplantation in children. By far the most common acute problem is lower respiratory tract infection, often with pneumonic consolidation. The frequency of chronic problems is not well documented.

Methods: A retrospective case note review was performed of all children transplanted in this regional centre.

Results: 125 transplants have been performed on 123 patients over the last 18 years. The mean age at transplant was 7.4 years (range 0.1–17.0 years) with mean length of follow up 6.8 years. 23 patients have died and 36 have been transferred to adult follow up, leaving 64 patients under paediatric follow up. Chronic respiratory problems have been documented in 33 children. Bronchiectasis has been identified in 10 patients; this has most frequently related to the well described problem of polysaccharide antibody deficiency occurring in children transplanted and receiving immunosuppressant therapy in the first two years of life (Gennery AR, Lancet 1998;351:1778–81). Six of these children have been managed with long term intravenous immunoglobulin (IVIG). Bronchiectasis has subsequently resolved in three patients, of whom two were managed with IVIG. Ten further children have had recurrent lower respiratory tract infections without bronchiectasis. Obstructive sleep apnoea related to lymphoid hyperplasia and requiring adenotonsillectomy has occurred in five patients. Subglottic stenosis has occurred in three children, but surgical intervention was not required. Significant compression of the left main bronchus related to a large donor heart has occurred in two children.

Conclusions: Chronic respiratory problems are therefore very common in this complex group of patients. The respiratory prognosis is usually good, but long term follow up by both respiratory paediatrician and immunologist is required.

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P-116

Induction therapy with Daclizumab in pediatric heart transplantation

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Daclizumab, a humanized monoclonal antibody, is a new immunosuppressive drug which binds with high affinity to the Tac subunit of the II-2 receptor complex. Effective immunosuppression with Daclizumab in adult patients encouraged the initiation of the administration of Daclizumab as induction therapy in pediatric heart transplantation.

Sixteen patients (9 boys, 7 girls, age 8.7 yrs, BMI 1.7 m^2), received Daclizumab as induction therapy in a dosis of 1 mg/kg intravenously perioperatively and on day 7 and 21 after orthotopic heart transplantation. Additional immunosuppression was cyclosporine (CsA, n = 14) or tacrolimus (TAC, n = 2), mycophenolate mofetil (MMF) and prednisolone. Prednisolone was tapered rapidly in the first six months after heart transplantation.

The administration of Daclizumab was not associated with any side effect. Owing to the blockade of the IL-2-receptor the dosage of calcineurin inhibitors could be reduced leading to less renal and hepatic toxicity. Instead of aiming at CsA trough levels of 350–400 ng/ml/TAC trough levels of 12–15 ng/ml in the first weeks after transplantation we reduced to 250 in the CsA group and to 10 in the TAC group. CD25 + T-lymphocytes began to be re-expressed after 2–3 months after administration of Daclizumab.

In a mean follow-up time of 2611.5 months no acute or chronic episode of rejection could be experienced. The incidence of opportunistic infections was not elevated (5 bacterial, 4 viral and 3 fungal infections which responded well to adequate treatment). No de novo malignancies, especially no lymphoproliferative disease (PTLD) was noticed. Actually patient and graft survival is 100%.

Our results show that immunoprophylaxis with Daclizumab induction therapy in pediatric heart transplantation is safe, effective and well tolerated and does not lead to increased opportunistic infections or malignancies. The reduction of calcineurin inhibitors led to less calcineurin related side effects and raised the quality of life of transplanted patients.

P-117

Clinical relevance of heart rate response to exercise in children after heart transplantation

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Background: Reinnervation of the transplanted heart has been described in adults and may be recognized by an immediate increase in heart rate upon exercise loading and unloading ("on" and "off"-phase). This response – so far not well described in children – has been observed in our pediatric heart transplant recipients. We examined whether the type of heart rate response to exercise was advantageous for physical exercise performance in our patient group of transplanted children.

Patients and Methods: 34 patients (18 male, 16 female, transplanted at 11,4 years of age, now 17,8 (range: 11,0–27,0) years old) were included in this study 8,2 years (range: 0,3–16,3) years after heart transplantation. All received a clinical examination including vital parameters. A symptom limited bicycle exercise test was performed according to a standardized protocol with an initial workload of 20 W. The work load was subsequently increased by 16 W every minute. Heart rate response was defined as "delayed, type 1" when the change was less than 5 bpm within 30 seconds when exercise was commenced ("on") or was stopped after reaching the patient's maximum ("off"), and was defined as "immediate, type 2", when these values were >= 5 bpm.

Results: The peak of O_2-uptake (max VO_2) during cardiopulmonary exercise during non-steady-state-cycloergometric exercise was significantly better in heart transplant recipients with an immediate (reinnervated) heart rate response (24,0 (21,4/27,6) ml · kg^{-1} · min^{-1} versus 20,0 (18,9/24,4) ml · kg^{-1} · min^{-1}, p = 0.027). The occurrence of the immediate type 2 heart rate response was associated with longer time post transplantation (p = 0.043) and younger age at transplantation (p = 0.014).

Conclusion: In pediatric heart transplant recipients, patients with an immediate heart rate response to exercise (indirectly suggesting partial reinnervation of the transplanted organ) profit from this normal response with a significantly higher maximum oxygen uptake during exercise.

P-118

Acute interventions following modified Norwood operation due to right ventricle to pulmonary artery (RV-PA) conduit stenosis

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Introduction: The outcome for neonates with hypoplastic left heart syndrome and its variants have improved consistently since the introduction of staged reconstructive surgery in the early 1980s. Recent modifications in surgical technique for Norwood stage 1 procedure, by replacing the Blalock-Taussig shunt with Right Ventricle-Pulmonary artery (RV-PA) conduit, have shown further improvement in the immediate survival.

Objective: To study the acute interventions (surgical and transcatheter) post modified Norwood Stage1 procedure with respect to RV-PA conduit stenosis/obstruction.

Methods: This was a retrospective study in tertiary referral centre. A total of 95 Modified Norwood operations (RV-PA conduit) were performed between April 2002 and May 2005. Six patients had acute interventions (surgical and transcatheter stent) for severe desaturation secondary to RV-PA conduit stenosis.

Age range at the time of intervention was 2 months to 4.5 months (median: 3months) and weight range 3.5 kg to 6.3 kg (median: 4.6 kg).

In 4 patients, the RV-PA conduit stenosis was demonstrated by angiography, however two patients presented extremely unstable with saturations of 30%, and had emergency surgery without cardiac catheter.

Results: Two patients had emergency transcatheter interventions with Liberte Coronary Stent (Boston Scientific) implantation with improvement in saturations and haemodynamics. This allowed planned surgical interventions, 2 weeks following the stent placement in first patient (Replacement of RV-PA conduit and Tricuspid valve repair) and 5 weeks post stent in the second patient (Cavopulmonary Shunt) with good recovery.

Four patients underwent urgent surgical intervention in the form of Bidirectional Cavopulmonary Shunt with good outcome.

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Four out of the above 6 patients had blood culture positive sepsis with infected RV-PA conduits.

No mortality or procedural complications were encountered.

All 6 patients await further palliative surgical procedures leading to completion of Fontan.

Conclusions: Acute RV-PA conduit stenosis can be a serious complication following Modified Norwood 1 procedure. This can be successfully treated either by transcatheter approach to stabilize the patient until further surgery is planned with more stable haemodynamics or by emergency surgical approach, with good outcome.

P-119

Growth characteristics of the aortic arch after the Norwood operation in patients with hypoplastic left heart syndrome (HLHS)

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Aim: The Norwood operation of HLHS includes the reconstruction of the aortic arch with homograft or bovine pericardium. We sought to characterize the growth potential of the reconstructed aortic arch compared to normal values.

Methods: We investigated retrospectively echocardiograms and angiograms of 109 patients who had their Norwood operation and subsequent Fontan palliation between 02/1996 and 10/2004. The diameter of the reconstructed aortic arch was measured at nine defined points. There were 43 reconstructions using homograft and 66 reconstructions using bovine pericardium. The follow-up interval was up to nine years.

Results: The morphology of the reconstructed aortic arch after the Norwood operation is characterized by a significant dilatation compared to the normal values ($Z_{echo} = +2.4 \pm 1.9$) respective the diameter of the descending aorta, measured next to the diaphragm (ratio aortic arch/descending aorta 1.7 ± 0.5). The root of the neo-aorta was dilated as well ($Z_{angio} = +4.4 \pm 1.7$; $Z_{echo} = +1.9 \pm 1.8$). There was no significant change in diameter and no sign of increasing dilatation in the follow-up to one year after the completion of the Fontan circulation. There was no difference in the results regarding the material used for the reconstruction of the aortic arch.

Conclusions: The root of the neo-aorta and the reconstructed aortic arch after the staged palliation showed persistently a considerable dilatation with an area of change in caliber to the descending aorta. We assume a growth of the native parts of the reconstructed aortic arch. It needs further investigations to evaluate the influence of the dilatation on haemodynamic effects.

P-120

Arrhythmias after palliative surgery of hypoplastic left heart syndrome (HLHS) – midterm follow up

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Aim: To study the prevalence of rhythm disturbances with special consideration of sinus node dysfunction (SND) in patients with HLHS arising along their palliative surgical treatment with modified Norwood–Hemifontan operation and total cavopulmonary connection (lateral tunnel).

Methods: From 1996 to 2005 70 patients were examined at the age of 0 to 113 months by a total of 267 Holter-recordings (3 ± 2.3 recordings per patient). 56 patients already completed their surgical palliation, 14 have still a Hemifontan circulation. We paid attention to any rhythm disturbance, but especially to SND, which was defined as occurrence of sinus bradycardia accordant to age and/or junctional rhythm >20 minutes, and noticed the time of development.

Results: In 33 patients (47%) a SND was diagnosed at a mean age of 37 ± 17 months, mostly after the total cavopulmonary connection. SND occurred as bradycardia in all these patients with additional junctional rhythm in only 3 of them. When two groups of patients born before and after the year 2000, respectively were compared until the age of 48 months, the younger group showed a significantly lower prevalence of SND. Other rhythm disturbances as AV block, SVES, VES and paroxysmal tachycardias did not play a significant role in our analysis.

Conclusions: Mid term follow up of patients with HLHS after surgical palliation reveals a high prevalence of SND, mostly presenting as isolated sinus bradycardia. Its haemodynamic effect on size and function of the right ventricle has to be investigated. Therefore, a permanent pacemaker treatment does not seem to be justified for clinically asymptomatic patients.

P-121

Closure of extracardiac Fontan fenestration by using the covered CP stent

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Background: Closure of extra-cardiac Fontan fenestration has been performed so far by using different devices designed to close intra-cardiac or aorto-pulmonary communications. These devices necessitate the introduction of a guide-wire and a long sheath trough the fenestration into the pulmonary atrium. This approach can be made difficult by the fenestration's angle and can be burdened by the risk of systemic embolism.

Methods: We closed the extra-cardiac fenestration of 8 patients having had a total cavo-pulmonary connection by using the covered CP stent. Median age and weight of patients were 9 years and 26 Kg, respectively. The stent was mounted on a BiB balloon, having the same diameter of the extra-cardiac conduit and deployed at fenestration's origin. The long sheath diameters was 12 or 14 French.

Results: Mean procedural and fluoroscopy time were $41 + 9$ and $9 + 1.7$ minutes, respectively. Mean central venous pressure non significantly increased from $10.7 + 2.7$ to $12.2 + 2.5$ mmHg and oxygen saturation significantly increased from $90.8 + 3.3$ to $98.2 + 1.7\%$ ($p = 0.008$). Femoral approach was used in all but 1 patient having bilateral thrombosis of femoral veins. No procedural or intra-hospital complications occurred. In particular, no arrhythmias or systemic embolism was observed. Residual shunt was never present. At a median follow-up of 6 months all patients have a normal oxygen saturations, all are symptom-free.

Conclusions: Closure of extra-cardiac Fontan fenestrations by using the covered CP stent is an easy and safe procedure that allows short procedural and fluoroscopy times. This technique could be preferred in relatively old children, due to the need of large sheaths. The potential risks of entering the pulmonary atrium trough the fenestration are avoided.

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P-122

Development of the MAGIC congenital heart disease catheterization database for interventional outcome studies

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As the field of catheter based therapies for congenital heart disease continues to expand, we lack the evidence based data to make appropriate therapeutic decisions in the catheterization laboratory, specifically because of our inability to simply and reliably share outcome data across multiple centers. We investigated whether a commonly used (> 100 centers) international congenital heart disease catheterization database software program (PedCath™) could be modified to become an automatic catheterization data submission tool to a central database for outcome analysis of registry type data (minimal supplemental questions, 7–10) and clinical studies (with in depth supplemental questions, 18–20). We designed data templates for registry type data collection on pulmonary and aortic valvuloplasty and clinical study type data collection on ASD closure and coarctation of the aorta angioplasty and stenting. To test the feasibility of such a tool we formed a national group of 7 congenital heart disease centers, the Mid-Atlantic Group of Inter-ventional Cardiology (MAGIC), to warehouse and analyze catheterization data. We successfully modified PedCath™ to transfer over a 13 month pilot period, the results of 256 therapeutic procedures for ASD closure (151 procedures), coarctation of the aorta angioplasty and stenting (39 procedures), and pulmonary (47 procedures) and aortic (19 procedures) balloon valvuloplasties. Short term follow up within the 13 month period was received on 31 patients. Analysis of immediate results demonstrated a high rate of success for the recorded procedures with low complication rates. The common platform for local storage of clinical catheterization data and transfer to the central data warehouse minimized data entry with high data accuracy. In addition, automated report generation and transfer by email from the database allowed monthly feedback on progress to the investigators. Based on the activity of the participating 7 MAGIC centers, we predict data on >1200 therapeutic procedures can be collected each year for analysis. In conclusion, this pilot study demonstrated the successful development of a collaborative, simple process, requiring minimal data entry, which would allow >100 congenital heart centers from around the world to immediately share cardiac catheterization data for long term outcome determinations of catheter based therapies for congenital heart disease.

P-123

Long-term results of percutaneous balloon angioplasty of recurrent aortic coarctation

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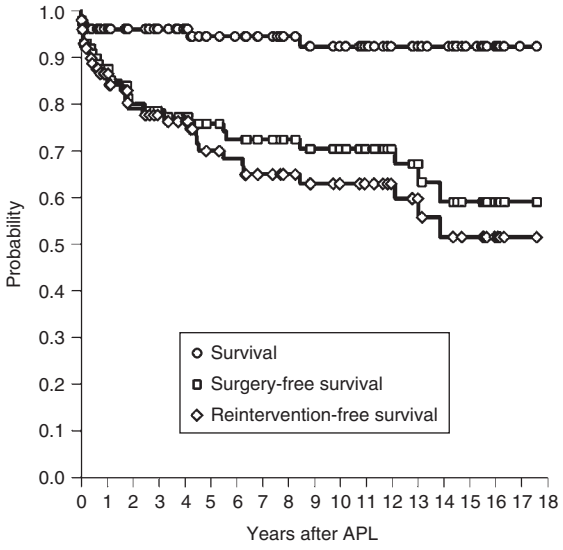
Objective: To assess efficacy, safety, and long-term results of the balloon angioplasty of recurrent aortic coarctation.

Patients and Methods: In the years 1986 to 2005, recoarctation angioplasty was performed in 102 patients aged 36 days to 32.6 years (median 268 days), 32 days to 15.8 years (median 251 days) after a surgery for aortic coarctation (N = 95) or aortic arch interruption (N = 7). In 59 patients the coarctation was isolated and 43 had concomitant intracardiac diseases. Balloons with diameter ⩽ descending thoracic aorta diameter and ⩽three times the recoarctation diameter were used. Patients were regularly seen at outpatient clinic and studied by complete echocardiography. Follow-up period ranged up to 17.6 years after the angioplasty (median 6.4 years, 725 total cumulative patent years).

Results: Due to the angioplasty systolic pressure gradient dropped form 37.3 ± 18.3 (±SD) to 17.8 ± 12.9 mmHg and mean gradient from 14.6 ± 9.5 to 6.6 ± 5.6 mmHg (both $P < 0.001$). Angiographic recoarctation diameter increased from 43.4 ± 12.2 to 67.2 ± 14.9% of the descending aorta diameter ($P < 0.001$). Peak Doppler gradient was reduced from 55.3 ± 8.8 to 31.6 ± 16.2 mmHg ($P < 0.001$) and did not increase significantly over the follow-up period (33.7 ± 21.2 mmHg at latest follow up, $P = 0.240$). Repeated interventions at the recoarctation site were necessary in 29 patients (28.43%): surgery in 21, repeated angioplasty in 5, and surgery after a repeated angioplasty in 3. In 1 patient (0.98%) an intimal tear necessitated a surgical revision and 1 patient (0.98%) died of sudden aortic bleeding that occurred 28 hours after a stent implantation during a repeated angioplasty. Other 4 patients (3.92%) with a good result of the angioplasty died because of a concomitant intracardiac disease. 17.6 years after the angioplasty the actuarial probability of survival was 92.3 ± 3.2% (±SEE), of surgery-free survival 59.0 ± 7.5%, and of reintervention-free survival 51.5 ± 7.7%.



Conclusions: In long-term perspective, percutaneous balloon angioplasty of recurrent aortic coarctation without a stent deployment is effective in more than a half of patients. Serious procedural complications may be expected in 1 to 2% of cases.

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(For P-124, please see OP-28)

P-125

Application of Amplatzer Muscular Ventricular Septal Occluders for closures of perimembranous VSD

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Introduction: Perimembranous ventricular septal defect (PmVSD) is nowadays frequently closed interventionally by asymmetric Amplatzer device. Our experience in such a treatment with Muscular VSD Occluder (MVSO) is presented.
Material & Method: Seven patients (pts) in mean age 14.1 (from 3.2 to 40) y with body weight 31 (14–56) kg were included. There were 5 females and 2 males. All had PmVSD with mean diameter 4.5 (from 4 to 5.5) mm. VSDs were extended toward outflow tract with aortic rim 5 (from 4 to 6) mm. In all VSD lower border of the defect was formed by muscular part of ventricular septum. Mean pulmonary artery pressure was 23 (15–30) mm Hg and Qp/Qs 1.7 (1.6–2.0). In all pts MVSO were applied in routine (transvenous) fashion and in 1 pt by retrograde implantation. In 5 pts 6 mm MVSO was used and in 2 pts − 8 mm. Fluoroscopy time was 25.4 (from 9 to 49) min.
Results: Procedures were performed without any complications in all pts and complete closure of VSD was achieved. In 3 patients trivial tricuspid incompetency appeared after closure. It did not increased and no other adverse effects of such therapy were observed in 2.0 (0.2–3.2) years of follow up.
Conclusion: Applications of Amplatzer Muscular Ventricular Septal Occluders for closure of selected perimembranous VSD seems to be a safe and effective option of treatment.

(For P-126, please see OP-36)

P-127
Transvenous permanent pacing in patients with venous baffle occlusion or obstruction following the Mustard procedure

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Introduction: The atrial switch operation (Senning or Mustard) was the surgical treatment of choice for patients with d-transposition of the great arteries for decades. Sinus node dysfunction and bradyarrhythmia requiring pacemaker implantation is a common sequel of that operation. Some patients also develop stenoses or occlusion of the systemic venous pathways, which makes standard transvenous pacing difficult. We report on our experience with percutaneous recanalization and/or stent implantation, followed by concomitant transvenous permanent pacing in a cohort of children and adolescents following the Mustard procedure.
Patients: Seven patients with symptomatic bradyarrhythmia after Mustard operation (one following combined Mustard and arterial switch operation for congenitally corrected transposition) were considered for transvenous pacing. Six of them had a stenosis in the superior caval pathway, and one a complete occlusion of the superior vena cava. Their median age was 14.1(range 5–19) years.
Methods: Six of the 7 patients underwent stent implantation (Palmaz P308 or 4014 stent mounted on a 15 mm or 20 mm diameter balloon), which abolished the stenosis, and allowed pacemaker implantation. In 1 patient, recanalization of the atretic segment of superior vena cava was performed by transseptal needle perforation, followed by stent implantation (P308 on a 20 mm diameter balloon). Thereafter, transvenous pacemaker implantation could be performed. Four patients received a DDDR system, and 3 a single lead (AAIR) system. In five patients the pacemaker was implanted during the same procedure, in the other 2 the pacemaker was implanted after an interval of 6 weeks.
Results: The median follow up was 52 (2–120) months. One pacemaker system had to be explanted because of pacemaker pocket infection. He is currently awaiting reimplantation of a pacemaker system. In two others the pacemaker generator was electively replaced due to end-of-life indication. None of the patients has clinical evidence of superior caval stenosis or obstruction, and none of the chronically implanted leads (>6 months) has required readvancement or removal.
Conclusion: Stenting of the superior caval baffle followed by transvenous pacemaker implantation is an elegant alternative to epicardial pacemaker implantation in patients with symptomatic bradyarrhythmia following the Mustard procedure.

P-128
Radiofrequency catheter ablation of septal hypertrophy in children with hypertrophic cardiomyopathy: initial experience

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Objectives: To describe the initial results and early follow-up data after radiofrequency catheter ablation (RFA) of septal hypertrophy in children with hypertrophic obstructive cardiomyopathy (HOCM).
Methods: 8 children (5 female, age range 5–17 years; weight range 17–52 kg) with symptomatic HOCM underwent RFA for septal reduction. After initial LV angiography to delineate the anatomical extent of LVOT obstruction, sequential AV pacing was instituted to see whether septal gradient could be influenced by pacing techniques. If this was not the case, RFA was performed. The His bundle was initially plotted and marked using the Localisa mapping system (Medtronic, USA). Using an 8 F cooled tip catheter (Sprinklr, Medtronic,USA) with an infusion rate of 300 ml/minute during RFA, ablation was commenced at the most apical extent of the hypertrophied septum, and 3 lines of contiguous lesions were applied upto the aortic valve. Care was taken to stay away from the His bundle during RFA. The number of RF lesions ranged between 10 and 50.
Results: The LVOT Doppler gradient changed from median of 80 (range 50–112) mm Hg pre-RFA to 30 (20–50) mm Hg at 72 hours post-RFA. Cardiac troponin T (5.1–8.6 micrograms/L; normal value < 0.1) and CK-MB (45–397 units/L; normal < 20) were considerably elevated at 24 hours, confirming significant myocardial necrosis. All patients had abolition of symptoms at 6 weeks' follow-up. Recurrence of obstruction, or incomplete relief was seen in 3 patients, within 6 months of RFA; 1 underwent redo RFA with success; one is awaiting surgery, and a third has been lost to follow-up.
Conclusions: RFA for HOCM produces relief of LVOT obstruction with resolution of symptoms. It avoids AV block, and can be repeated. Further studies are warranted.

(For P-129, please see OP-35)

P-130
Newborns and infants with acute complications after surgical creation of systemic-pulmonary shunt: interventional stent implantation – an alternative to reoperation?

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Objective: Shunt operation is still the treatment of choice in newborns with duct dependent pulmonary circulation or duct dependent systemic blood flow when performing Norwood stage I procedure. Acute and chronically complications after shunt surgery are described. Acute complications as stenosis, thrombosis or kinking are life threatening. These complications need surgical treatment with revision or exchange of the shunt immediately. Interventional reopening and implantation of stents into the shunt is presented in this report as an alternative to early reoperation.
Method: Four patients, three with complex cardiac malformations and duct dependent lung circulation and one patients with hypoplastic left heart syndrome after Norwood Stage I procedure, had acute shunt complications and were treated with stent implantation. The median patients age at time of intervention was 15 days (range 6 to 62 days) and median patients weight was 3.3 kg (range 2.6 to 4.6 kg). Two patients had no additional blood flow via the pulmonary artery. One of these had kinking of a sano-shunt; the other had thrombosis with subtotal closure of a central aortopulmonary shunt. The other two patients had kinking of a central aortic-pulmonary shunt or thrombosis of a right-sided modified Blalock-Taussig shunt.
Results: In four patients a total of six stents was implanted during five procedures. Two patients needed two stents to stabilise the whole shunt. These two patients had thrombosis causing severe obstruction or subtotal occlusion of the shunts. In one Patient two stents were implanted during one procedure. In the other patient kinking of the shunt was treated first with one stent. A second procedure with implantation of a second stent was necessary due to thrombosis. Increase of oxygen saturation represented the improved pulmonary blood flow after the intervention in all patients. Further surgical procedures could be performed at the right time.
Conclusion: Interventional treatment of acute shunt complications appears to be a save alternative to surgery. Diagnosis and therapy can be done at the same procedure. In addition, the number of individual surgical procedures can be reduced particularly in patients with complex cardiac malformations.

(For P-131, please see OP-32)

(For P-132, please see OP-34)

P-133
Interventional catheter preparation for Fontan procedure

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Background: The low-risk successful completion of a Fontan procedure is largely dependant on near-perfect anatomic and haemodynamic features in patients with univentricular hearts.
Aim: This study was conducted to evaluate the role of interventional catheter techniques in the preparation of patients with univentricular hearts who were considered for completion of a Fontan procedure.
Methods: We retrospectively reviewed our database for Fontan procedures performed over the last 5 years. We identified patients who underwent interventional cardiac catheterisation procedures within 2 years prior to completion of the Fontan procedure.
Results: A total of 180 Fontan operations were performed at our institute from Jan 2000 to Sep 2005. All patients had cardiac catheterisation performed either at Birmingham Children's Hospital or the referring hospital. Forty patients (22.2%) had transcatheter interventions performed within 2 years prior to operation to improve haemodynamics and or treat anatomical abnormalities.

Interventions	Number	(%)
EPS and RFA	2	1.1
Stenting of pulmonary artery	14	7.8
Balloon angioplasty of pulmonary artery	6	3.3
Occlusion of forward pulmonary flow	3	1.7
Balloon angioplasty of aorta	1	0.6
Occlusion of venous collaterals	15	8.3

There were no catheter related deaths. In 16 patients (8.9%) the Fontan procedure could be delayed for >6 months due to clinical improvement post catheter intervention. Two patients (1.1%) who previously acutely failed a Fontan procedure underwent successful Fontan completion after catheter intervention. Cumulative post Fontan mortality was 5/180 (2.8%).
Conclusion: Interventional cardiac catheterisation techniques contribute significantly to the timing and successful outcome of the Fontan procedure.

P-134
The type of initial treatment for congenital aortic valve stenosis has no impact on long-term re-intervention free survival

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Introduction: Over recent years transvascular balloon dilation (BD) replaced surgical valvotomy (SV) as first-line treatment for congenital aortic valve stenosis (AS). Whether, the type of initial intervention influences long-term outcome is still the subject of intense debate. We hypothesized that surgical valvotomy may lead to a higher procedural mortality but will result in a longer lasting effect.
Methods: We conducted a retrospective analysis on AS in children less than 1 year, treated in our centre between 1964 and 2004. Factors influencing survival and re-intervention free survival were tested by Cox regression for different variables. Evolution of aortic insufficiency and ventricular function were tested using a proportional odds model and a linear mixed model, respectively.
Results: 62 children with AS have undergone either SV (n = 29) or BD (n = 33) at a median age of 39 days (range 0 to 364 days). Median follow-up was 7.1 years (range 0 to 35 years). The overall survival at 10 years was 80%. There was no evidence for a difference in survival between SV and BD ($p = 0.7$). No risk factors were identified that influenced overall survival.

Re-intervention for AS was needed in 25 children (SV: n = 12, BD: n = 13) after a median interval of 3.3 years (range 0 to 23 years). There was no evidence for a difference in re-intervention free survival between SV and BD ($p = 0.38$). Age at time of intervention is a significant risk factor for re-intervention free survival ($p < 0.01$; Hazard ratio 0.91) [Figure].

We found no difference in evolution of aortic insufficiency ($p = 0.98$) and fraction of shortening ($p = 0.75$) after SV or BD.

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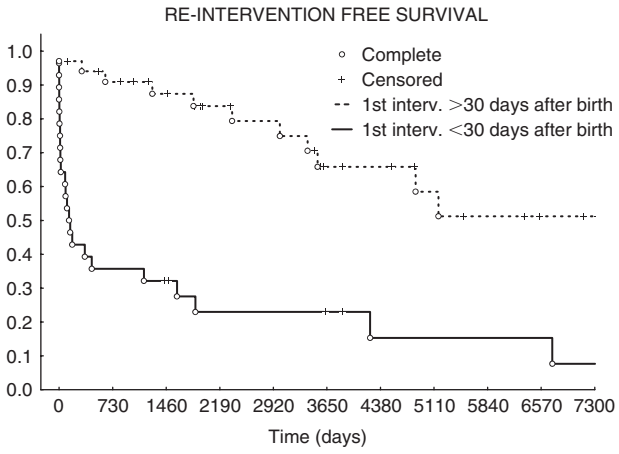

Conclusion: BD as well as SV are effective treatment options for AS and long-term results are encouraging. Overall survival was not influenced by the type of first intervention for AS. Re-intervention is common but did not affect survival. The only predictor for re-intervention free survival was age at time of intervention. There was no difference in evolution of aortic insufficiency or fraction of shortening after SV or BD.

(For P-135, please see OP-31)

P-136

Late fenestration of any atrial septum by sequential stent flaring, including the extra-cardiac conduit in a Fontan circuit

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Background: A fenestration can be life-saving in patients with low cardiac output due to a failing Fontan circuit or obstructive pulmonary hypertension. Delivery of a short stent in diabolo across the atrial septum can be problematic. Since the use of extra-cardiac conduits in Fontan circuits, the wall between systemic and pulmonary venous blood consists of the conduit tissue, a virtual pericardial space and atrial muscular wall, which are usually separated during the puncture; this complicates the fenestration procedure.
Technique: The stent is prepared by the Stumper technique: a loop of 3–4 mm diameter is created using a set of temporary epicardial pacing wires. The resultant loop is placed over the mid-portion of a standard 12 mm valvuloplasty balloon catheter. A standard short stent is then mounted onto the valvuloplasty balloon over the knot.
Stent Deployment: The mounted stent is delivered through a 9 French Mullins sheath. The stent is advanced through the sheath into the atrium. The distal half of the stent is uncovered from the sheath and inflated, flaring the distal half of the stent well within the atrium. The balloon-stent-sheath complex is pulled back, allowing perfect centering or reapproximation of the atrial wall against the extra cardiac Fontan conduit. The balloon is then fully inflated, opening the stent in diabolo across the fenestration. The balloon (with the metal knot-wire) is removed. Further dilation of the mid-section of the stent is performed with a 5 or 6 mm non compliant balloon until the desired arterial desaturation.
Patient and Results: This procedure was performed with success in 4 patients: one 5.7 year old patient, 28 months after Fontan completion because of protein losing enteropathy after Fontan completion, and in 3 adults with obstructive pulmonary hypertension awaiting lung transplantation.
Conclusion: Sequential opening of stent across the atrial wall allows accurate positioning, reapproximation of different layers if required, and a stable limited right-to-left shunt. This technique is currently our technique of choice when creating a fenestration in a simple or complex atrial wall.

(For P-137, please see OP-33)

P-138

Pulmonary vein stenosis presenting as pulmonary hypertension: short and medium term outcome after stent implantation

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Background: Pulmonary vein stenosis (PVS), though rare, is a challenging and persistent disease, and its treatment has long been an enigma. Both transcatheter and open surgical approaches have been technically difficult, often with poor outcomes. We report our experience with isolated PVS (not associated with total anomalous pulmonary venous return) presenting to a tertiary pulmonary hypertension (PH) center and its treatment by stent implantation as a prelude to pharmacological manipulation of the pulmonary vascular bed.
Methods: Retrospective review of all patients with native PVS presenting as PH who were treated by stent implantation between December 2000 and August 2005 at The Children's Hospital Denver, Colorado. Clinical, echocardiographic and cardiac catheterization data, including follow-up data, were collected for analysis.
Results: 5 patients had PVS amongst the approximately 200 patients with PH and a total of 11 stents were implanted. Ages ranged from 1.5 to 16 years (median 8 years), and weight ranged from 8.8 to 51 kg (median 24.8 kg). Two patients had a prior surgical attempt to repair stenotic pulmonary veins. All but one patient had bare metal stents and one had a sirolimus drug eluting stent implanted. The patients were maintained on coumadin and antiplatelet therapy in addition to individualized PH medication. Follow-up data at 18 to 61 months show improved pulmonary hemodynamics. Mean pulmonary artery pressure decreased in all patients (47 ± 18 mmHg to 23 ± 6 mmHg, $p < 0.05$) following stent placement. Eight stents required re-intervention for growth and in-stent stenosis, and one stent was completely occluded from intimal growth. Two patients required further implantation of a second stent in the same vein. Three stents have not required further intervention to this point.
Conclusion: Isolated PVS is significant cause of PH. Based on our institutional experience, transcatheter stent placement in stenosed pulmonary veins is safe, feasible and provides short term hemodynamic relief. All stents but one placed in this series have remained patent up to 53 months and one-quarter have not required any further intervention following implantation.

P-139

Magnetic guidewire navigation in congenital heart disease – a feasibility study

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Introduction: Cardiac catheterization continues to be important in the diagnosis and treatment of systemic to pulmonary collaterals, persistent ductus arteriosus and pulmonary artery stenosis. Targeting a vessel with the guidewire can be difficult, especially in pediatric patients. This study addresses the feasibility of navigating a guidewire that is equipped with a magnetic tip by an external magnetic field.

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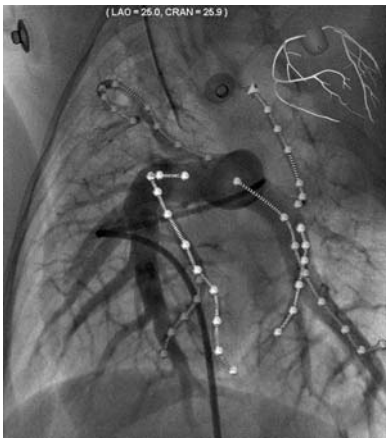

Methods: Six piglets with an average weight of 12.9 kg were catheterized using magnetic guidewire navigation. Two pulmonary arteriograms were taken from different angles. From these, a computer interface calculated coordinates for the selected vessels in space (Fig). Two external magnets on both sides of the animal created a manoeuvrable magnetic field between them and within the piglet's thorax. The magnetic vector was directed along the three dimensional vessel course. The magnetic guidewire tip bent in the orientation of the external field and target vessels were entered by advancing the guidewire. Aortic and renal branches were addressed in an analogue fashion. Emphasis was given to vessels that seemed difficult to enter because of their peripheral position, their sharply angled origin off the main vessel or because of their tortuous course.
Results: 35 out of 37 arteries (94.6%) were reached and entered successfully. Branches at angles of less than 90° were entered. Peripheral arteries of small diameter stabilized the floppy guidewire and facilitated magnetic navigation as compared to the large central arteries.
Conclusion: This is the first report on vascular magnetic guidewire navigation using a manoeuvrable magnetic field. With this method, tortuous and angled arterial branches could be entered. We expect it to help in reaching anomalous vessels and peripheral stenoses, thus shortening procedure times and radiation exposure. The current need for two separate angiographies is disadvantageous and may be overcome by biplane imaging or the incorporation of MRI or CT data for vessel identification.

Vascular magnetic guidewire navigation is feasible. It can be used successfully to enter small arterial branches at sharp angles and may be an adjunct in cardiac catheterization of pediatric patients.

P-140

Use of premounted stents delivered "naked" without guiding catheters in pulmonary conduits and branch pulmonary artery stenosis in pediatric patients

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Introduction: In pediatric interventional procedures, stents are typically delivered protected via long delivery catheters. Data regarding safety, feasibility and effectiveness of delivery of premounted stents without guiding catheters is limited. We report our experience in delivery of premounted stents "naked" over a wire in selected patients with pulmonary conduit and pulmonary artery branch stenosis.
Methods: All patients selected (n = 10) had discrete pulmonary conduit stenosis and/or pulmonary artery branch stenosis in the context of Tetrallogy of Fallot with Pulmonary valve atresia (TOF, PA) (n = 4), ventricular septal defect (VSD) and PA (n = 1), Truncus arteriosus (n = 2), Hypoplastic left heart syndrome (HLHS) s/p Sano conduit (n = 1) and single ventricle physiology s/p Fontan with isolated LPA stenosis (n = 2).
Results: 10 pts received 15 stents in 13 vessels. Age range 2 mo. to 19 yrs (mean 4.0 yrs), Wt was 3.7 to 60.3 kg (mean 18.6 kg), Ht was 58 to 160 cm (mean 98.9 cm). Gradient decreased from 34 to 5 mmHg ($p < 0.001$), minimum diameter increased from 3.2 mm to 9.2 mm ($p < 0.001$). Fluoroscopy time was 21.9 to 81.3 min (mean 44.5 min), French size was 5 to 7 Fr. There were no procedure failures. All stents were successfully deployed. One pt had transient bradycardia and oxygen desaturation during stent deployment that resolved upon deflation of the balloon. The pt with HLHS s/p Sano conduit and RPA stenosis underwent successful completion of the Glenn shunt with removal of the stents from the Sano conduit and RPA stenosis two months later. Another patient with PA, VSD underwent conduit revision with stent removal four months later. Both pts were 2 and 3 mo. of age respectively when stents were placed. All other pts remain stable at f/u 2 mo. to 2.5 yrs (mean 14 mo.).
Conclusion: The delivery of "naked" premounted stents was feasible, safe and effective in selected pts postponing additional surgery. There were no cases of stent embolizations. The stents were easily removed at time of further surgical palliation or repair.

P-141

Covered cp stents for occlusion of intracardial tunnel leaks in children after total cavopulmonary connection

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Introduction: Leaks in intraatrial tunnels may complicate the postoperative course of patients after TCPC. They are often the cause for an increasing cyanosis and exercise intolerance. Frequently, device closure of such leaks is technically demanding and may lead to obstructions and less favourable flow dynamics. We report our experience with transcatheter closure of such leaks with covered stents.
Case study: Three patients with various types of functionally univentricular hearts were diagnosed with right-to left shunts across tunnel leaks unsuitable for umbrella-type device closure. One patient was a 17 year old girl with a double inlet left ventricle and l-TGA. She received a TCPC with a fenestrated intracardial tunnel three years later. The fenestration was closed by implantation of a rashkind umbrella 4½ years after surgery. She now showed an increased cyanosis on exertion (decline from 85–90% to 70%). The second case was a 14 year old boy with double inlet left ventricle and d-TGA. At 3 years a TCPC was created with an intracardiac tunnel. He now showed signs of increasing cyanosis ($SatO_2$ 84%) and dyspnea on exertion. The third patient was a 12 year old boy with tricuspid atresia and hypoplastic right ventricle. At the age of 5 month he received a bidirectional Glenn procedure and at the age of 1½ years received a TCPC with a fenestrated intracardiac tunnel. A large tunnel leak was observed leading to severe cyanosis (Sat 88%). For implantation of the covered stents angiographic delineation of the tunnel leak was obtained. Covered CP stents were chosen with a stent length and diameter of 14 × 34 mm and 20 × 39 mm, respectively. One patient had three stents implanted sequentially with a total length of approx. 80 mm and an increasing diameter from 22 to 25 mm. Postinterventional angiographies demonstrated the correct placements of the stent in the tunnel and alleviation of R/L shunts, the oxygen saturation increased to values >95%. No rhythm disturbances were encountered. No thromboembolic complications occurred.
Conclusion: Intraatrial tunnel leaks after TCPC can be safely and effectively be treated by use of covered cp stents.

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P-142

Vascular tissue reactions to stent implantation for congenital heart disease

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Introduction: In contrast to arteriosclerotic lesions in the adult, little is known on tissue reactions to stents in neonatal or juvenile vessels.
Methods: We examined 72 specimen from animal series (n = 38) and after explanation from pediatric patients (n = 34). The stents had been implanted in the arterial duct (n = 35), in systemic arteries (n = 19), pulmonary arteries (n = 7), veins (n = 6), or modified Blalock-Taussig-Shunts (n = 5). Besides standard histology we employed scanning electron microscopy and immunohistochemistry for tissue characterisation.
Results: Within 24 hours after implantation a fibrin layer surrounding the stent struts could be demonstrated. After initial endothelial damage, recovery with formation of a endothelial layer was completed within 4 weeks. No significant cellular inflammatory reactions were detected. Intimal hyperplasia due to fibromuscular cell proliferation was seen as early as 1 week after implantation progressing with time. Immunohistochemistry demonstrated an antigen pattern of the intimal cells concordant with intimal hyperplasia in arteriosclerosis of the adult (positive stain for smooth muscle actin, vimentin, and desmin inter alia). Results were not markedly different for site of implantation or between specimen from humans and animals.
Conclusions: While endothelial damage appeared to be transient, intimal hyperplasia was progressing with time. Although different in origin, intimal hyperplasia after stent implantation in the young shows analogies to intimal hyperplasia in arteriosclerosis.

P-143

Long term outcome after balloon angioplasty of native and postoperative aortic coarctation

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Introduction: Surgery had been the traditional treatment for native coarctation, while balloon angioplasty (BAP) is an accepted treatment for recoarctation. Controversies exist about angioplasty in native coarctation especially in neonates. The intermediate and long term results are reported, focused on restenosis treated surgically or by stent placement and the relief of systemic hypertension.
Patients and Methods: From 01/99–12/05, 156 BAP were performed in 140 pts. The age ranged from 5 days to 28 y (median 110 d) with a mean body weight of 5.6 kg. 38 of them were neonates. Diagnoses: 125 interventions were performed in classical circumscript coarctation, 31 were complex coarctations (HLHS, UVH with aortic arch hypoplasia), other associated lesions were VSD, bicuspid aortic valve and mitral stenosis. Native coarctation was present in 84 pts., recoarctation after surgery or intervention in 72 pts. Balloon angioplasty was performed according standard methods with a balloon diameter of 2–18 mm (m = 7.8).
Results: There were no procedural deaths. The diameter of the stenosis was 1–14 mm (m = 4), the initial systolic pressure gradient was measured 2–110 mmHg (m = 39.4). The residual gradient was 0–40 mmHg (m = 12.2). During follow up, 1d–6y (m = 770 d), 65 pts. had surgery for definitive repair, in 16 pts. redilation was performed and in 11 pts. a stent was implanted at the site of coarctation. 72 patients were on medication for systemic hypertension with betablockers, ACE-inhibitors, AT1-receptor blockers or amlodipine.
Conclusion: BAP is a suitable tool in native and postoperative coarctation, particularly in newborns as rescue therapy. Only a minority of neonates show good long term results. Residual stenosis can be effectively treated by stent implantation.

(For P-144, please see OP-30)

P-145

Percutaneous closure of multiple ASDs

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Introduction: The fossa ovalis may contain more than one hole of different sizes.
Patients and Methods: Between January 1998 and December 2005, 96 out of 1155 patients treated for ASD had multiple (dASDs) or multifenestrated (mASD) defects. Four devices were used: Amplatzer ASD occluder (ASO), Amplatzer Cribriform occluder (ACO), Cardioseal/Starflex (CS/SF) and Helex.
Results: Mean age and weight were 38 ± 22 years and 57 ± 19 kgs, respectively. Qp/Qs ratio was 2 ± 0.5. Procedure and fluoroscopic times were 80 ± 25 and 18 ± 7 minutes, respectively.

Multifenestrated defects (mASD) were found in 22 patients. Sixty-four subjects had dASD. Finally, 10 patients had multifenestrated PFO(mPFO). mASD were closed by using one device (78%). In 13 patients an Amplatzer device was used. In 9 subjects a CS/SF was used (one device in 5 subjects; two devices in 4 patients). mPFOs were closed by using an Helex device in one case, two CS/SF in three patients, 3 ACO in one patient, one Amplatzer PFO occluder in 4 subjects. dASDs were treated in 11 pts by implanting CS/SF (one device in 4 subjetcs, two devices in 7 patients) and in 53 pts by using a device of the Amplatzer family (one device in 25 subjects, two devices in 28 patients). Complications occurred in 2 patients (2.1%). One patient experienced a significant inguinal haematoma; while the second had atrial fibrillation treated by external cardioversion. At a mean follow-up of 3 ± 1.3 years no complication occurred.
Conclusions: Percutaneous closure of multiple and multifenestrated atrial septal defects is safe and efficacious.

P-146

Extending the indications for percutaneous pulmonary valve implantation

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Introduction: Percutaneous pulmonary valve implantation (PPVI) is a safe and effective treatment for conduit dysfunction in patients with repaired congenital heart disease (CHD). Conventional indications are significant pulmonary regurgitation with increasing right ventricular size and/or exercise intolerance and/or right ventricular outflow tract obstruction with right ventricular pressure at least 2/3 systemic. PPVI is less invasive than surgery with lower morbidity. Here we report the cases from our series with formal contraindications for surgical intervention, which had PPVI.
Methods: From our total series of 113, two patients with terminal pulmonary vascular disease in the context of operated CHD and two patients in cardiogenic shock (Table) underwent PPVI. All

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patients were NYHA class IV. PPVI was performed under general anaesthesia with invasive blood pressure monitoring. Clinical and echocardiographic follow-up were documented.

Pat.	Age [years]	Diagnosis	Lesion	Clinical Status
1	12	Repaired hemi-truncus, disconnected LPA, pulmonary hypertension in solitary right lung	Pulmonary regurgitation	Saturations 78% at rest Requiring continuous O_2 Bosentan
2	34	Repaired truncus arteriosus, residual VSD, Eisenmenger's syndrome	Pulmonary regurgitation	Saturations 83% at rest
3	44	Repaired double outlet left ventricle, anterior aorta	Pulmonary regurgitation	Inotrope dependant Cardiogenic shock Ascites Renal Failure
4	9	Complex left ventricular outflow tract obstruction with Ross operation	Right ventricular outflow tract obstruction Post-surgical supravalvular aortic stenosis	Cardiogenic shock Ascites Renal Failure Acidotic

Results: PPV was successfully implanted in all patients. Mean procedure time was 126 ± 56 minutes. Patient 1 underwent an uncomplicated recovery, whereas patient 2 proved slow to wean from mechanical ventilation. Following discharge, both patients had a dramatic clinical improvement with improvement in oxygen saturations and exercise capacity. The PPVs remained competent at one year despite exposure to high valve closure pressures related to high diastolic pulmonary pressures. Patient 3 experienced a prolonged recovery phase (20 days) with requirement for inotropes and haemofiltration. He remained in class NYHA IV at one month after the procedure. Patient 4 underwent a combined procedure successfully abolishing the 140 mmHg gradient over the ascending aorta and PPVI. After an initial improvement (Central Venous Pressure dropped from 45 to 24 mmHg without blood loss) she died 24 hours later in pulmonary oedema.
Conclusion: The lower morbidity associated with PPVI permits extension of this procedure to patients with prohibitive surgical risk. Patients with pulmonary hypertension and dysfunctional conduits can benefit dramatically from restoration of a competent pulmonary valve. The procedure can be tolerated in patients "in extremis". However, rescue of haemodynamic stability will depend on pre-procedural conditions of the patients. Planning of PPVI procedures for such patients should include the availability of full intensive care support.

(For P-147, please see OP-29)

P-148
Efficacy and long-term patency of fenestrated amplatzer devices in children

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Objective: Fenestrated Amplatzer devices have been used for atrial septal defect (ASD) closure, whilst maintaining a small atrial communication in patients with pulmonary hypertension or in patients who are elderly. Experience with these devices in paediatric populations is limited. We present indications, procedural and long-term complications as well as long-term patency and efficacy in children.
Patients and Methods: Since 2003, 10 children (m = 5) between 1.5 and 15.5 years (median 4.5) had a fenestrated Amplatzer device inserted. Six patients had custom-made fenestrated devices, four received devices with local modifications of standard atrial septal occluder. Patients were assessed regularly with transthoracic echocardiography.
Results: In 9 patients, devices were implanted for severe pulmonary hypertension associated with right heart failure (n = 4) or syncope (n = 3). Two children with Down's syndrome underwent fenestrated device closure of a haemodynamically important ASD associated with pulmonary hypertension. One patient received a fenestrated device to offload the left atrium during extracorporal membrane oxygenation.

In 8 patients the device is still in situ after a mean follow-up of 24.5 months (17.5–31.0 months). Two patients underwent heart/heart-and-lung transplantation. Ventricular function improved in patients who had a fenestrated device implanted for right heart failure. Two of three patients who received a fenestrated device closure for syncope were free of symptoms. One child presented with recurrent syncopal attacks after the device was found to be no longer patent.

Device patency was confirmed in 5 patients (all custom-made) after 26 months follow up (23.6–31 months), 4 being treated with warfarin, one received aspirin.

Device patency could not be demonstrated in 4 children after a median follow-up of 10 months (2.8–14.5 months). During long-term follow-up one custom-made and all modified devices occluded. Three of those children were on aspirin, one was warfarinised.
Conclusion: Implantation of fenestrated atrial devices is clinically feasible; however patency of the fenestration during longer-term follow-up has been insufficient.

Warfarin appears to be more effective than aspirin in preventing closure of the fenestration. There is a need for the development of new fenestrated devices, which would guarantee long-term patency in patients who would benefit from an atrial communication.

P-149
Transcatheter PFO closure mitigates aura migraine headaches abolishing spontaneous right-to-left shunting

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Background: Patent foramen ovale (PFO) has been implicated in the etiology of migraine headache with aura (MHA) but the mechanisms that link right-to-left shunt to MHA are unclear and the reports on the efficacy of transcatheter PFO closure on MHA prevention are scarce.
Methods: We reviewed the clinical records of 131 consecutive patients underwent successful transcatheter PFO closure at our institution at a mean age of 45 ± 13 years because of cryptogenic stroke. 35 out of the 131 patients (27%) had a diagnosis of MHA made by the primary care physician or the referring neurologist. MHA incidence and severity were assessed by using migraine disability assessment questionnaire (MIDAS). Presence and magnitude of right-to-left shunt was assessed in all patients by means of transesopahgeal echocardiography and also by means of transcranial Doppler (TCD) in the last 50 patients (38%).
Results: Patients with MHA had a higher prevalence of thrombophilia ($p = 0.007$), a more complex atrial septal anatomy ($p = 0.001$) and they also had higher prevalence of spontaneous right-to-left shunt and of spontaneous large shunt, both at transesophageal echocardiography ($p = 0.015$, and $p = 0.028$ respectively) and at TCD ($p = 0.036$, and $p = 0.038$ respectively). After the procedure 32/35 patients (91%) had either complete resolution or significant improvement in their MHA. At a mean follow-up of

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1.7 ± 1.3 years, MHA disappeared completely in 29/35 patients (83%). Of the remaining 6 patients, 3 patients (8%) had an improvement of >2 grades in the incidence and severity of MHA, 2 patients did not show any improvement of their MHA, whereas 1 patient reported a severe relapse of MHA about 1 year after the procedure.
Conclusions: In patients with PFO, MHA is associated with spontaneous large right-to-left shunt and thrombophilic conditions. Transcatheter defect closure seems to be an effective and safe mean to treat MHA in patients with PFO.

P-150
Elective stent implantation for aortic coarctation and recoarctation
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Objective: To report early and midterm results of elective stent implantation for treatment of aortic coarctation and recoarctation in older children, adolescents and adults.
Methods: Prospective observational study. Inclusion criteria: weight above 30 kg, isolated aortic coarctation (native or recurrent) with systolic gradient >30 mmHg, informed consent. Exclusion criteria: involvement of carotid arteries, aortic arch hypoplasia, concomitant large aortic aneurysms. Endpoints: early success (immediate residual gradient <15 mmHg), early complications, midterm success (last follow-up gradient <15 mmHg at least one year after intervention), midterm complications. Patients: 47 patients (10 female, 37 male) were enrolled since 06/2001, among them 25 native and 22 re-coarctations (18 postoperative, 4 post balloon dilatation). Data at intervention are given as median (range): age 17 (8–55) years, weight 62 (30–103) kg, height 169 (138–192) cm, systolic gradient 50 (30–95) mmHg. Technique: femoral arterial access, long sheaths 11 (9–12) Charrier. Balloon expandable stents were used in 45 patients (26 Palmaz 4014, 19 CP 8zig), self expandable stents in 2 patients. Balloon diameter was 15 (12–25) mm. Follow up is 25 (1–54) months.
Results: Initial success 44/47 patients (94%). Initial failure 3/47 patients (6%), gradients 20, 20 and 30 mmHg, resp. Procedural complications were two stent dislocations to the descending aorta, a second stent was successfully implanted in both. Other early complications: one femoral artery aneurysm requiring surgery. Midterm success 33/37 patients (89%). One small aneurysm was found and treated by implantation of a covered stent into the stent. One growth-related-restenosis was successfully balloon dilated. The gradients of two of the three patients with initial failure were reduced from 20 to 15 and 30 to 20 mmHg by re-dilatation with high pressure balloons, but complete expansion was not reached. No patient developed stent dislocation during follow-up.
Conclusions: Stent implantation is an attractive alternative to balloon dilatation or surgery in isolated aortic coarctation in older children, adolescents and adults. However, postoperative recoarctation may be extremely rigid thus preventing optimal results. Postinterventional restenosis seems not to be a problem. Aneurysm formation may occur and requires attentive follow-up.

(For P-151, please see OP-27)

P-152
Transcatheter device closure of fenestration after extracardiac Fontan procedure
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Objective: To review the results of routine interventional fenestration closure in a single center regarding haemodynamic data before and after closure.
Methods and Patients: Between 5/2001 and 8/2006 40 consecutive patients received interventional closure of a 4 mm fenestration in an extracardiac GoreTex-Konduit three months after surgery. Diagnoses were HLHS in 16, classic single-ventricle in 7, imbalanced CAVC in 3, TA in 5, DORV in 6, PA/IVS in 2 and CCTGA in 1 patient. Mean age was 53 months, median-weight 16 kg (12–62 kg), median length 102 cm (82–175 cm), 30 patients were in constant sinus-rhythm, 10 in atrial-rhythm. CVP ranged from 9–21 mmHg (median 13 mmHg) with a transpulmonary gradient from 3–11 mmHg (median 5 mmHg), mixed-venous-saturation was at a median of 65% (39–72%) whereas aortic saturations were at a median of 88% (75–94%).
Results: All but one fenestrations were closed successfully with Amplatzer-ASO in 34 and Helex-occluder in 6 cases with one significant residual shunt in the Helex-group. CVP stayed at same level (9–21 mmHg, median 13 mmHg), mixed venous saturation dropped slightly to a median of 62.5% (38–72%). As aortic saturation rose to a median of 95% (87–98%) there was bigger AV-saturation difference of 32.5% in median (22–50). One patient with a MVO of 38% had congestive heart failure and the fenestration was left open. All patients again were put on cumarin. In a median follow up of 25 months (3–48 months) thrombus formation in the fontan-baffle without clinical symptoms was seen in one case. One patient developed severe plastic bronchitis and the fenestration had to be reopened surgically. Until now no cases of congestive heart failure or protein-loosing enteropathy were observed. No further loss of sinus-rhythm could be seen.
Conclusion: Transcatheter closure of fenestration in extracardiac Fontan-baffles is a safe and uncomplicated procedure. We could not find significant changes in central venous pressures and the partially remarkable increase in arterio-venous saturation difference had no clinical impact up to the observed level. So until yet in our experience no clear cut-off point whether a fenestration can be closed or not can be defined.

(For P-153, please see OP-19)

P-154
First peripheric stem cell transplantation in children with dilated cardiomyopathy
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Introduction or Basis or Objectives: Recent experimental studies suggested that bone marrow-derived progenitor cells or circulating progenitor cells might contribute to the regeneration of infarcted myocardium and enhance neovascularization of ischemic myocardium. There is only one report in the literature on patients with dilated cardiomyopathy who received such therapy.
Methods: Two patient (Pt) were added to our trial clinical study (9/F and 6/M). The patient's permissions were taken and the conditioning for peripheric blood stem cell transplantation (PBSCT) began with intravenous GCS-F (5 μg/kg) for 4 days. The peripheric stem cells were collected by Fresenius Aphesis System (AS-TEC204, Germany). At the fourth day of the GCS-F therapy cardiac catheterization was performed and the collected stem cells were infused to coronary arteries with slow infusion.

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The echocardiographic examinations were performed every week after the PBSCT.
Results: After 5 weeks of PBSCT the patient's clinical status became good in parallel to their echocardiographic findings. Ejection fraction and fractional shortening were increased (Pt1 16 vs 36, Pt2 37 vs 47). Also, their congestive heart failure was regressed and the inotropic support was ceased.
Conclusion: Intracoronary infusion of progenitor cells is declared as safe and feasible in patients with myocardial injury. Also, we observed no complication in our patients both during and after the PBSCT. The influence of treatments other than PBSCT on changes in the cardiac function is unlikely, because of there were no changes in medical therapies during the observation period. Based on the observations of the present trial, this novel form of the regeneration enhancement therapy holds the promise to improvement of the patients' clinical status while they are waiting for donor.

P-155
Stent fracture in percutaneous pulmonary valve implantation: a mathematical modelling study
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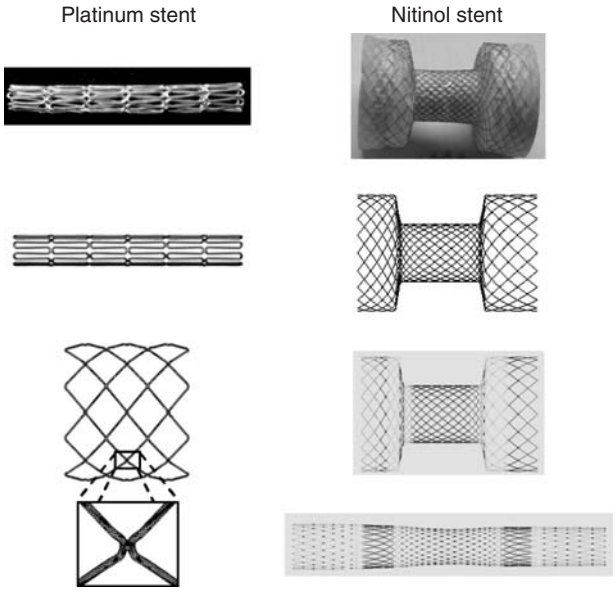
Introduction: Stent fracture is a recognized complication following percutaneous pulmonary valve implantation (PPVI). In our series (n = 101) we have seen stent fractures occur in 15 patients (53% male, age 9–38 years): spontaneously in 10 patients, following balloon dilatation in 3, and following implantation of a second percutaneous valve in 2. Many different factors play a role in the generation of device fracture including: stent material and design, the nature of the implantation site and stress induced by the functioning valve. The aim of this work is to study some of the factors responsible for stent fracture by means of mathematical modeling in order to develop new device design.
Methods: Finite element method is the technique used to perform the numerical analyses. Three different stent models were created: the first from platinum-10% iridium alloy; the second with the same geometry but with the addition of gold welds (this is the device currently used for PPVI); and the third from nitinol, a shape memory alloy, with a different design. Mathematical simulation of the deployment of these devices was performed.
Results: The models suggest that the platinum stent tends to fracture at the strut intersections, where there is evidence of highest stresses. The stent with gold welds show as the gold reinforces the crossing sections of the platinum device protecting them from fracture. However, the platinum parts of this stent present higher stress then the device without gold welds, because of the reinforcement itself. This suggests that the fracture occurs just distal to the welds. The nitinol stent has good elastic property but not enough force to open a calcified outflow tract. Furthermore, the highest stress occurs where the device tapers, which may affect patency should fracture occur (figure).
Conclusions: Mathematical modeling is an important technique to understand stent behaviour and integrity and may aid device design and material before many prototypes are actually manufactured.



P-156
Natriuretic peptides as markers of cardiac function in children with mechanical circulatory support
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Objective: Mechanical circulatory support (MCS) systems are used in children to sustain failing cardiac circulation. Treatment aims are either recovery or cardiac transplantation. Atrial and brain natriuretic peptides (NP) are expressed in the myocardium due to mechanical stretching of the myocardial walls. We measured plasma concentrations of NP in children under mechanical support to determine steady-state levels under support and the time of down-regulation as markers of myocardial recovery.
Methods: Between 12/2001 and 12/2005 blood samples were collected from 13 children (median age 12, range 0.2–17.5 years, median body weight 25, range 4–58 kg), all supported with a pneumatic pulsatile extracorporeal ventricular assist device (EXCOR, BerlinHeart). The underlying disease was cardiomyopathy (DCM) in 12 children and complex congenital heart disease in 1. One had a biventricular and 12 a left ventricular device. The median supporting time was 55 (range 6–220) days. Levels of midregion pro-ANP (mpANP), BNP and NT-pro-BNP were analysed using sandwich immunoassays.
Results: The actuarial survival rate was 84.6%. Nine children underwent heart transplantation (median follow-up 284, range 51–1469 days), 2 died und 1 patient was weaned from the device. Serial analyses of the biomarkers are listed in the table.

	Before MCS median (range)	After 1 week of MCS median (range)	After 4 weeks of MCS median (range)
mpANP (pmol/l)	1120 (255–2350)	282 (216–575)	261 (78–1210)
BNP (pg/ml)	2216 (165–13426)	329 (202–1776)	185 (11–785)
NT-pro-BNP (pg/ml)	49429 (7528–112057)	4733 (1022–15420)	2126 (716–8275)

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All children showed down-regulation of mpANP, BNP and NT-pro-BNP during support, although none reached normal plasma values. The lowest NT-pro-BNP level (250 pg/ml) was found in the later weaned child, a DCM patient, after 4 months on support and continuous decline. After 12 weeks of successful weaning he reached a normal NT-pro-BNP level (71 pg/ml).
Conclusions: Extremely high levels of all 3 NP reflect the severity of myocardial failure before device implantation. The time courses of mpANP, BNP and NT-pro-BNP differ. NT-pro-BNP appears to be a promising tool for the estimation of cardiac function in children and may be important when considering children for weaning from the device.

P-157

Perioperative changes of N-terminal brain natriuretic peptide,troponin I and C-reactive protein in neonates and children operated for congenital heart diseases

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Objective: To investigate perioperative secretion pattern of N-terminal BNP (Nt-BNP), high sensitivity C-reactive protein (hsCRP) and troponin I (TnI) and their relations with outcomes in children undergoing cardiac surgery.
Methods: Nt-BNP, hsCRP, TnI were measured in 43 children (25 male, 2 days–10 years) undergoing cardiac surgery at our institution fron April to December 2005 including 19 pts with LV overload and 24 pts with RV overload. Nt-BNP and TnI were determined by ECLIA assays and hsCRP by BN II Nephelometer. Blood samples were obtained within 48 hours before (Preop) and 3 (Postop1) and 8 days (Postop2) after operation. Postoperative clinical data were collected.
Results: Nt-BNP significantly increased after surgery (Mean(SD)) (Preop 1555 (3203) vs Postop1 4359 (4779) and Postop2 4146 (7296) pg/ml, $p < 0.05$) but not significantly decreased in Postop2 vs Postop1. TnI and hsCRP significantly increased after surgery and significantly decreased in Postop2 vs Postop1 (TnI Preop vs Postop1: 0.0344 (0.0545) vs 0.3991 (0.4812) ng/ml, $p < 0.001$; Postop1 vs Postop2: 0.3991 (0.4812) vs 0.1103 (0.1439) ng/ml, $p < 0.001$; hsCRP Preop vs Postop1: 0.5163 (0.6564) vs 5.4289 (5.3384) mg/dL, $p < 0.001$; hsCRP Postop 1: vs Postop2: 5.4289 (5.3384) vs 2.7229 (4.8872) mg/dL, $p < 0.05$). Preoperative Nt-proBNP and TnI levels positively correlated with complications (numbers) ($r = 0.45$; $p < 0.05$ and $r = 0.40$; $p < 0.05$) and ICU time stay (days) ($r = 0.55$; $p < 0.001$ and $r = 0.53$; $p < 0.001$). Pre and postoperative Nt-BNP were positively correlated with pre and postoperative TnI ($r = 0.83$; $p < 0.01$ and $r = 0.56$; $p < 0.001$). Only postoperative hsCRP were correlated with postoperative NT-BNP and TnI ($r = 0.43$; $p < 0.05$ and $r = 0.36$; $p < 0.05$).
Conclusions: Nt-BNP, TnI, hs-CRP levels increase after surgery in children with congenital heart diseases suggesting important role of neuroendocrinal activation, inflammatory response and cardiac injury operation. Pre operative Nt-BNP and TnI can predict post-surgical outcomes. Nt-BNP, TnI and hsCRP release are strongly correlated.

P-158

Interfacility transport of infants and children with cardiac disease: a multi-centre analysis

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Introduction: Infants and children with cardiac disease (CD) are often transferred to paediatric tertiary care centres for diagnostic, surgical, and/or critical care interventions. We hypothesized that these children are more severely ill, require more interventions, and are at a greater risk of dying compared to those from other diagnostic categories.
Methods: Multi-centre paediatric specialty care transport team cohort study (N = 4905) evaluating all patients with and without CD at the referring hospital and during transport. Differences between groups were determined by χ^2 analyses expressed as (n, %) for categorical data and Mann-Whitney Rank Sum Test for continuous variables presented as median [interquartile range]. Hospital morbidity and mortality rates were examined.
Results: Four-hundred and fifty (9.2%) children transported had a diagnosis of CD. They were younger in age, had higher pre-ICU PRISM scores, received more interventions, required longer scene times, and were admitted to the ICU more frequently. Mortality rates were significantly higher for CD patients, and this relationship remained after adjusting for pre-ICU PRISM scores and receiving tertiary centre (OR 2.70 [1.91–3.81]; $p < 0.001$).
Conclusions: Children who are transported with a diagnosis of CD require more interventions, have a higher severity of illness, suffer more unplanned events, and are at an increased risk of mortality. The evidence from this large multi-centre study demonstrates that children with CD may benefit from the expertise of a paediatric speciality care team. Supported by 1-H34-MC-00040-01 (EMSC).

Variable	Cardiac disease (N = 450)	All other diagnosis (N = 4455)	*p* value
Age (Months)	0.50 [0.25–15]	23 [4–84]	<0.001
Male gender (n, %)	266 (59.2%)	2547 (57.2%)	=0.431
Pre-ICU PRISM score	6 [4–11]	5 [0–9]	<0.001
TISS score	8 [5–16]	11.5 [6–23]	<0.001
Intervention (n, %)	263 (58.4%)	1973 (44.3%)	<0.001
Scene time (Minutes)	42.5 [26–60]	30 [20–47]	<0.001
Transport unplanned event (n, %)	43 (9.6%)	227 (5.1%)	<0.001
Admitted to ICU	408 (90.7%)	3117 (70%)	<0.001
Hospital mortality (after transport)	66 (14.7%)	226 (5.1%)	<0.001

P-159

Health-related quality of life and behavioral adjustment in children and adolescents after open-heart surgery

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Objectives: To assess health-related quality of life (HrQoL) and behavioral adjustment in school-aged children with congenital heart disease (CHD) who have undergone open-heart surgery and to identify medical, individual and familial predictors of outcome.
Methods: Between 1995 and 1998 155 children with CHD (40% cyanotic) underwent open-heart surgery in our hospital. 122

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patients (response rate 79%) were investigated at a mean age of 10.4 years (range 6.5–16.8). Child and parent rated HrQoL was evaluated by the TNO-AZL Child Quality of life Questionnaire. Psychosocial adjustment was assessed by the Strengths and Difficulties Questionnaire. Scores were compared with healthy reference groups. Predictive values of sociodemographic variables, medical variables, and family-related variables for HrQoL and behavioral adjustment were assessed.
Results: Mean age at operation was 2.3 years (range 0–8.7 years), mean ECC time was 95 min (range 5–206) and circulatory arrest was performed in 9% of the children. Most dimensions of HrQoL in children and adolescents with CHD were impaired compared to reference values. Deviations were an impairment of autonomy ($p = 0.02$), of motor ($p = 0.02$), social ($p < 0.001$), and emotional ($p < 0.001$) functioning. In addition, parents rated cognitive functioning of their children as diminished ($p < 0.001$). Also, behavioral adjustment in patients with CHD was significantly impaired compared to norms. Specifically, the prevalence of emotional symptoms (17.1%) and hyperactivity (13.1%) was increased. Severity of CHD and of medical therapy (e.g., ECC time and number of operations), deteriorated family relationships, and posttraumatic stress symptoms in parents had a negative impact on most dimensions of HrQoL and behavioral adjustment.
Conclusions: HrQoL and behavioral adjustment are impaired in children with CHD after open-heart surgery. Disease- and treatment-related variables, the quality of family relationships and parental adjustment are important predictors. Results show the importance of assessing HrQoL and behavioral adjustment in children and adolescents with CHD.

P-160
Quality of life in the pediatric ICD population
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The implantable cardioverter-defibrillator (ICD) can present significant psychological problems for children, because of increased lifestyle disruption and higher incidence of shocks.

The objective of this study was to evaluate the quality of life (QoL) in pediatric patients with ICDs in The Netherlands.
Methods: Between January and April 2005 children who underwent ICD implantation were studied. The study group consisted of 12 males and 10 females, mean age 15 yrs (SD 3 yrs, range 11–19 yrs). QoL was assessed with: 1) a disease specific QoL questionnaire for children: the Worries About ICD Scale (WAICD) 2) the SCL-90 questionnaire for anxiety, depression and sleeping disorders and 3) the TNO-AZL children's QoL questionnaire (TACQOL)

The mean age at ICD implantation was 13 yrs (SD 3 yrs), mean duration of ICD therapy was 3 yrs (SD 2 yrs). The indications for ICD therapy were aborted sudden cardiac death (31%), syncope (37%) or ventricular fibrillation(32%). Underlying cardiac disorders were congenital heart disease (26%), cardiomyopathy (26%) and primary electrical disease (48%).
Results: The internal consistency of the WAICD (26 items) was adequate (Cronbach's alpha (CA) 0.77). However 15 items showed a negative item-total correlation or lack of variance. Therefore a short form 11-item WAICD version was used (CA 0.86).

A high correlation was shown between the 11-item WAICD and the duration of ICD therapy (Pearson Correlation (PC) 0.72), which indicates that longer duration of ICD therapy in children is associated with more worries. Children with ICD therapy longer than 2 yrs ($n = 12$) showed significantly more worries (Student T test $p = 0.02$). The most important factor for the worries was the larger number of mostly inappropriate shocks (PC 0.67).

The SCL-90 and TACQOL scores were, compared with the group means of normative populations, within the normal range.
Conclusions: The number of shocks has an important impact on worry and anxiety in the pediatric ICD population. This result emphasizes the need to avoid or reduce inappropriate shocks to improve QoL in this group of patients and is relevant to improve the psychological support.

P-161
Effects of bosentan on clinical and exercise capacity in patients with Eisenmenger physiology
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Objective: To study the clinical and exercise capacity of chronic oral administration of bosentan in a cohort of patients with Eisenmenger physiology.
Methods: Eleven patients were included in the study from July 2003, to September 2005, including 7 women and 4 males with a mean age of 34 years (17–51 years). Patients were in NYHA functional class III to IV with a resting O_2 saturation 77 ± 9%. One of them had previously received intravenous prostanoid treatment. Bosentan was administered at an initial dose of 62.5 mg BID and increased 4 weeks later to a dose of 125 mg BID. Patients underwent clinical, exercise evaluation at baseline and 1, 2, 3, 6 and 12 months of treatment and treatment-related changes were compared using GEE method.
Results: Bosentan improved significantly NYHA class after 3 months of treatment, Borg dyspnoea index after 2 months of treatment and 6-min walk test after 6 months. At one year of treatment, these modifications were still present showing significant improvement from baseline : NYHA class 2.54 ± 0.7 versus 3.82 ± 0.7, Borg dyspnoea index 3.81 ± 1.32 versus 6.54 ± 2.29 and distance travelled in 6-min walk test 322 ± 62 m versus 216 ± 111 m. No patient died during the study and no significant changes of liver enzyme, haemoglobin level and resting O_2 saturation were observed.
Conclusion: Bosentan was safe and well tolerated in patients with Eisenmenger physiology both at introduction and after 1 year of treatment. Larger studies are necessary to establish the safety and efficacy of bosentan in the long-term follow-up in this population.

P-162
Creation of interatrial communications in iPAH: Fenestrated Amplatzer occluder or atrial septostomy and stent implantation
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Introduction: Atrial septostomy (AS) emerges as a potential therapeutic option in iPAH. Fenestrated Amplatzer occluders were used in iPAH with a hemodynamically relevant ASD II.

We describe five patients with iPAH. In two children (7 and 9 yy) a fenestrated ASD occluder was implanted, and in three others an AS was performed because of severe right ventricular failure. In one of them, – a 3-year old boy – not responding to prostacyclin, bosentan, sildenafil and imatinib an interatrial stent was placed as a definitive fenestration for rescue treatment of acute RV-failure.

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Methods: One child received an 18 mm Amplatzer-ASD-occluder (ASO) with a customized fenestration of 6 mm. In a 9-year old boy a self-fabricated fenestrated ASO was created to achieve an interatrial pop-off for right-to-left shunt, if necessary. The definitive fenestration was performed by utilizing a premounted balloon-expandable Genesis stent (20 × 10 mm), which was delivered through a long 13-F sheath guided by a 0.035 inch guidewire and pushed through a prepared fenestration within the fully deployed ASO. After exact placement the stent was expanded to an hourglass shape with a minimal central waist of about 6–7 mm.

In further three pts. an atrial balloon septostomy was performed after transseptal puncture using a Brockenbrough needle. A guide wire was passed into the left atrium and lodged in a pulmonary vein followed by serial dilation of the interatrial septum. Stenting of the interatrial septum in the 3-year-old boy was performed in a technique as reported previously in newborns.

Results: After implantation of the self-fabricated fenestrated ASO the ratio of the pulmonary-arterial to systemic arterial pressure decreased to less tan 0.4 by our test protocol, which allowed long-term treatment with amlodipine. In the manufactured fenestrated ASO, patency of the interatrial communication could be demonstrated for at least three years. The patient with RV-failure survived due to the effective right-to-left shunting in situations of acute pulmonary arterial crises. One pt. with septostomy died 4 months later, awaiting heart-lung transplantation.

Conclusion: Variable techniques can be used for establishment of interatrial pop-off ventil; such a communication seems to be a potential therapeutic option in some patients with iPAH.

P-163

Brain natriuretic peptide as a predictive marker of right ventricular dysfunction and clinical worsening in pediatric patients with pulmonary arterial hypertension

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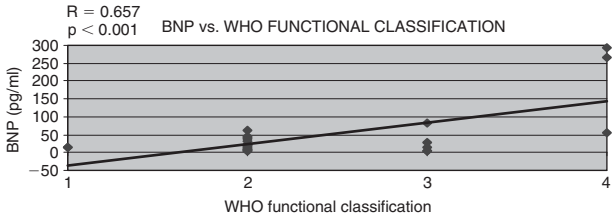
Introduction: Brain Natriuretic Peptide (BNP) is an endogenous peptide hormone secreted mainly by the cardiac ventricles. BNP secretion is released by myocardial ventricular stretch, which makes plasma BNP a suitable biomarker for the evaluation of right ventricular dysfunction. Recently, the role of BNP in adult patients with pulmonary arterial hypertension (PAH) and right ventricular failure has been examined. However, little is known about the diagnostic role of BNP in right ventricular dysfunction and pulmonary arterial hypertension in infants and children.

Methods: Serum BNP levels from January until December of 2005 were retrospectively evaluated in children with idiopathic PAH (IPAH) and PAH associated with congenital heart defects (CHD) from the Pulmonary Hypertension Program at The Children's Hospital in Denver. BNP was correlated to hemodynamic data measured invasively in the cardiac catheterization laboratory. 25/37 patients met the inclusion criteria as they had BNP levels drawn within four months of cardiac catheterization. BNP levels were drawn within ten days of catheterization in 20/25 patients; whereas 5/25 patients had BNP levels drawn within four months of catheterization without undergoing change in therapy during that time.

Results: Patient demographics were as follows: A total of 25 pediatric patients were identified, 48% with IPAH and 52% with PAH secondary to CHD. 14/25 were female (56%) and 11/25 were male (44%). The mean age was 10 years (range: 2 years–18 years). BNP levels correlated with: (1) WHO functional classification ($R = 0.657, p < 0.001$) (see graph), (2) mean right atrial pressure ($R = 0.69, p < 0.0001$) and (3) pulmonary vascular resistance index ($R = 0.78, p < 0.0001$). A trend was noted between elevated BNP levels and mean pulmonary artery pressure ($R = 0.37, p = 0.05$). As expected, no correlation was observed between elevated BNP levels and pulmonary capillary wedge pressure ($R = 0.009, p = 0.96$).



Conclusions: Serum BNP levels correlate with the severity of right ventricular dysfunction as indicated by elevated right atrial pressures and WHO functional classification. Therefore, serum BNP maybe a useful marker of clinical worsening in pediatric patients with IPAH or PAH secondary to CHD.

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P-164

Epoprostenol treatment improves durvival and quality of life in children with severe pulmonary hypertension

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Introduction: Pulmonary hypertension (PHT) is a progressive disease with poor prognosis. Epoprostenol has been shown to improve exercise tolerance, haemodynamics and survival. Limited data are available in paediatric cohorts. We report our long-term experience of epoprostenol treatment in children with PHT, focusing on outcome, feasibility and associated morbidity.

Patients and Methods: All children treated with continuous intravenous epoprostenol in our centre since November 1997 were included in this retrospective study. Thirty-nine patients (18 male) between 4 months and 17 years of age (median 5.4 years) were included. Eleven patients were treated with epoprostenol alone, while 28 have been on concomitant therapy with bosentan or sildenafil. Twenty-five patients had idiopathic PHT and 14 patients had PHT associated with congenital heart disease (n = 9), connective tissue disease (n = 2), chronic lung disease (n = 2) or HIV (n = 1). In 26 patients pulmonary artery pressure equalled or exceeded systemic pressures. 13 patients had half to systemic pulmonary artery pressures. Patients were regularly assessed by physical examination, functional class, electro- and transthoracic echocardiography and six-minute walk test.

Results: Mean follow-up was 27 ± 21 months (range: 1–90 months). Seven patients died and 8 patients underwent transplantation during follow-up. Cumulative survival at 1, 2, and 3 years, was 94%, 90% and 84%. Survival of patients with idiopathic PHT at 1, 2, and 3 years was 96%, 91% and 83%, respectively. Six-minute-walking distance improved by a mean of 103 ± 62 m ($p < 0.003$) on treatment. Similarly, functional class improved significantly during the first year ($p < 0.001$) and this effect was maintained up to 3 years of treatment ($p < 0.002$). In addition, the children's weight improved significantly (z-score at baseline: −1.55 ± 1.74, on treatment −1.16 ± 1.8; $p < 0.03$).

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On 43 occasions antibiotic treatment was given for line/insertion-site infections. Line sepsis occurred in four patients. Overall, 29 Hickman-catheter changes were necessary in 15 patients during follow-up (=0.33 line changes/patient year) due to infection (n = 20 occasions), leaking of the line (n = 5) and catheter dislocation (n = 4).
Conclusions: Epoprostenol therapy improved survival, WHO functional class, exercise tolerance and ability to thrive in children with severe PHT. Though associated with considerable morbidity epoprostenol represents a feasible therapy even for young children.

(For P-165, please see OP-12)

P-166
COArctation Long-term Assessment (COALA-Study): perioperative and long-term mortality of surgical coarctation repair

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Aim of the Study: To assess survival and the cause of death after surgical coarctation repair.
Patients and Methods: From 1974 to now 404 patients born before 1.1.1965 underwent surgery for isolated aortic coarctation in our hospital. In the COALA study their follow-up was investigated. In deceased patients the cause of death was extracted from hospital charts, from personal reports of relatives, or from the German death register.
Results: In total, 22 patients (5.4%) died at surgery and during the follow-up of up to 30 years. One 10 days old infant died at coarctation repair (year 1981). Six infants, all under the age of 2 months, died postoperatively (year 1976–1983). There were no other perioperative deaths. At logistic regression analysis only young age at surgery was an independent risk factor.

At long-term follow-up 2 patients died from an ascending aorta aneurysm, one of them with a genetic mosaic of Turner syndrome at the 35th week of her second pregnancy. Three patients died from "heart failure" (16, 19, 49 years old) and in 8 patients (24, 34, 36, 38, 40, 41, 44, 60 years) cause of death could not be assessed. In another 2 patients a cardiac cause could be excluded: one died at a car accident (18 years) and one of cancer (44 years).

COX regression analysis revealed an older age at surgery ($p = 0.0005$) as single risk factor for late mortality with patients older than 20 years being at higher risk ($p < 0.0005$). Surgical technique and sex were not independently significant.
Conclusion: Perioperative mortality at coarctation repair was confined to infants who could not be stabilized preoperatively. Late mortality after repair is primarily related to a higher age at surgery.

P-167
Biological solutions in children for redo-operations in mid and distal aortic arch stenosis

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Objectives: Reconstruction of long segment coarctations and interrupted arch pathology can lead to re-stenosis. In some of these malformations children have been operated with the use of interposition grafts. The lack of growth potential and the inability to resolve arising problems with catheter interventions remain a major concern, especially in the infant group. This study describes our experience with this surgical challenging group of patients with a trend to a more definite solution.
Methods: Between June 2003 throughout August 2005, 12 patients underwent surgical correction for mid and distal aortic arch problems. Five out of the 12 patients had interposition grafts (Goretex tubes) ranging from 3 to 7 centimeters for several diagnoses like coarctation aortae and interrupted arch. Other patients had non biological patches, not successfully amenable to catheter interventions. All patients were re-operated through a midline sternal approach with moderate hypothermic circulatory arrest and antegrade cerebral perfusion. The primary goal in all patients was to establish some kind of connection between native tissues, whereas the other side of the arch was enlarged using bovine pericardial patch. In 2 patients a hybrid approach was administered to ease the procedure, whereas one patient received an aortic autotransplantation with the use of its own ascending aortic wall.
Results: In all patients a native re-connection could be achieved with extensive mobilization and dissection. In the patient with a 7 centimeters long interposition graft of Goretex an aortic autotransplantation was necessary. There was no operative mortality and no persisting morbidity. All patients are doing well after their intervention. The follow-up ranges from 7 to 34 months with up until now no signs or symptoms of re-stenosis. Complications like bronchial obstruction or neurological sequelae were not encountered.
Conclusions: Although interposition grafts and non biological patches are rarely used nowadays, they still exist in earlier operated patients. In stead of continuously replacing these grafts and patches, it is possible to exchange those for a more permanent solution, with no mortality and low morbidity. Especially in the growing infant and child this takes care of a more prosperous outlook for the future.

P-168
Is intermittent warm cardioplegia hazardous in pediatric cardiac surgery?

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Objective: To analyse our initial experience with warm blood cardioplegia in pediatric surgery.
Methods: Since 2001, we have employed this technique in 1400 consecutive patients, with the exception of infra-diaphragmatic total anomalous pulmonary venous drainage. During aortic cross clamping, cardioplegic solution mixed with warm blood was injected every 15 minutes in the aortic root or in each coronary artery.

Efficacy and safety were evaluated from several parameters:

- Spontaneous rythm following aortic unclamping
- Troponin I rise 12 hours from aortic cross clamping
- Postoperative ventilation in 4 selected groups (148 ventricular septal defect under 6 months, 109 Fallot's Tetralogy under 1 year, 52 complete atrio ventricular septal defect under 1 year and 54 neonatal switch)
- Length of Intensive Care stay for the whole group.

The results are retrospectively compared with those of 950 patients operated on with cold blood cardioplegia, including 71 ventricular septal defect, 76 Fallot's Tetralogy, 66 complete atrio ventricular septal defect and 52 neonatal switch.
Results: The results are shown in comparison with those of cold blood cardioplegia. All parameters differed in a statistically significant fashion (t-test: $p < 0.05$).

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- Resumption of sinus rhythm after release of aortic clamp: 99% vs 77%;
- Troponin I rise <10 nanograms: 44% vs 37%;
- Ventilation time: Ventricular septal defect 9 ± 8 hours vs 13 ± 10, Fallot's Tetralogy 8 ± 6 hours vs 14 ± 17, atrio ventricular septal defect 44 ± 82 hours vs 76 ± 101, Transposition of the great arteries 41 ± 29 hours vs 83 ± 105 (the 4 groups of patients operated on with warm or cold cardioplegia being equivalent with respect to mean age, mean weight, percentage of Down syndrom in the atrioventricular septal defect group).
- Intensive care length of stay: less than 2 days in 86% of patients vs 75%.

Conclusions: Although improved clinical outcomes may not be due exclusively to warm blood cardioplegia, its use was demonstrated to be safe and efficacious. This technique deserves to be considered a valid alternative to "classic" cold blood cardioplegia methods.

(For P-169, please see OP-15)

P-170

Is cavo-bipulmonary anastomosis obligatory in the surgical correction of type C and D Ebstein anomaly?

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Objective: The correction of type C and D Ebstein anomaly is difficult and may require the association of cavo-bipulmonary anastomosis (CBPA). In order to reduce the incidence of CBPA, we opted for a new technical approach, consisting of partial plication of the atrialized chamber, hence transforming this dyskinetic chamber into an akinetic one. By including the partially plicated atrialized chamber into the right ventricle (RV) during correction, the volume of the RV is thereby increased, diminishing the necessity for CBPA.
Methods: From 1998 to 2005, 22 cases of Ebstein's anomaly, 18 of type C and 4 of type D, underwent surgical correction with the technique described above. Mean age was 16 ± 2 years (11 months–58 years). Associated lesions included 21/22 cases with patent foramen ovale (PFO) or atrial septal defect (ASD), 2/22 cases with pulmonary stenosis, 1/22 cases with ventricular septal defect (VSD), and 1/22 cases with moderate left ventricular (LV) hypoplasia. In all 22 cases, tricuspid insufficiency was grade III or IV on preoperative echocardiography.
Results: There were no peri- or postoperative deaths at midterm. During initial surgery, 3/4 type D patients required concomitant CBPA; however, not one type C patient required CBPA. The patient with LV hypoplasia (type D) required RV assistance, which could be weaned by the 9th day. One case (type C) needed concomitant ablation of a Wolf-Parkinson-White aberration, and another (type C), isthmic ablation for atrial flutter. Mean follow-up was 26 ± 8 months. All patients are currently in sinus rhythm, and at the last echocardiographic control, 1/18 type C cases has moderate residual tricuspid insufficiency, and 1/18 discrete to moderate insufficiency. The mean trans-tricuspid gradient for all 22 cases at short and midterm is 3 ± 2 mmHg. During follow-up, one patient with type C presenting with annular pulmonary stenosis has required CBPA 3 months after initial surgery.
Conclusion: With this modified technique of partial plication of the atrialized chamber, the need for CBPA in type C Ebstein anomaly decreases, but CBPA remains obligatory in the majority of type D anomalies.

P-171

Initial experience with hybrid surgery in congenital heart disease in a single center

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Introduction: A hybrid intervention is a joint procedure involving the interventional cardiologist and the cardiac surgeon. At our institution we have opted for this type of approach in congenital heart disease since 2005. We report here our initial experience.
Cases:

1. A 3 year old boy with double aortic arch and multiple muscular ventricular septal defects (VSD), was readdressed for pulmonary band (PAB) removal and residual VSD closure after previous palliation. After surgical removal of the PAB, the surgeon provided a minimal transventricular access for placement of a 6 mm Amplatzer® muscular VSD occluder by the cardiologist under transoesophageal guidance. The patient was extubated the same day and discharged after 5 days.
2. An 8 year old girl with Williams syndrome was followed for two large VSDs and severe peripheral pulmonary arteries (PA) stenosis. The membranous VSD was closed surgically, the muscular VSD during the same operation by direct placement of a 12 mm Amplatzer® muscular VSD occluder. During rewarming, balloon angioplasty of peripheral PA stenosis was achieved under fluoroscopy. Patient was extubated the following day and discharged after 8 days.
3. A 9 year old boy post tetralogy of Fallot repair had severe distal stenosis of the right ventricular to PA conduit. With patient on partial cardiopulmonary bypass, an incision was made on the conduit and a CP 8 Zig 16 stent placed on the stenosis. The child passed on full bypass and the definitive placement of the stent achieved. The child was extubated at the end of the intervention and discharged after 6 days.
4. A newborn presented at 2 days life with complex aortic arch anatomy: left aortic arch and right descending thoracic aorta perfused directly from a right arterial duct and left PA atresia. The arterial duct was stented with a Genesis XD stent dilated at 7 mm. Two days later the cardiac surgeon made banded the right PA. The child was extubated after the operation and discharged a week later.

Conclusion: Hybrid approach opens new ways of correction or palliation in congenital heart disease with encouraging results and less morbidity.

(For P-172, please see OP-13)

P-173

Surgery for congenital heart disease in adult age: Italian multicentric study

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Objective: A multicentric study involving 7 major italian centers, so as to evaluate the impact of cardiac surgery in adults with congenital heart disease in our country.
Methods: We collected data of 856 patients who underwent 1179 procedures from January, 1st 2000 to December 31st 2004. Patients were divided into three groups: *Group I – Palliation (3.1%):* any operation performed to improve patient's clinical status without restoring normal anatomy or physiology. *Group II – Repair (69.7%):* First operation performed in the patient, to achieve an anatomic or physiologic repair by separation of the pulmonary from systemic circulation (including also Fontan-types, and 1 and ½ ventricle repairs). Most frequent procedures were: atrial septal defect closure (35.8%), partial anomalous pulmonary venous connection repair (7.2%), ventricular septal defect closure (5.3%). *Group III – Reoperation (27.4%):* All procedures performed after repair either anatomic or physiologic. The most frequent procedures were conduit replacement (9.8%), aortic (8.6%) or pulmonary valve replacement (7.7%).
Results: Preoperatively 34.6% of patients were in NYHA class I, 48.4% in class II, 14.2% in class III and 2.8% in class IV. Synus rhythm was present in 83%.

There were 1179 procedure performed in 856 patients (1.37 procedure/patient), with a hospital mortality of 3.1%. Overall mean intensive care unit stay was 2.3 days (range:1–102 days). Major complications were reported in 247 pts (28.8%), with postoperative arrhythmias being the most frequent (26%).

At mean follow-up of 22 months (range 1 month–5.5 years), 86% of data were available. Late death occurred in 5 patients (0.5%). Patients were in NYHA class I in 79.3%, II in 17.6%, III in 2.9%, and only one patient in class IV (0.11%).

Overall survival estimates is 89%, 95% and 89% at 5 years for groups I, II, III respectively. Freedom from adverse events at 5 years is 97.8% for acyanotic vs 88.2% for preoperative cyanotic patients ($p < 0.05$).
Conclusions: Surgery for congenital heart disease in adult age is a safe and a low risk treatment. However patients with preoperative cyanosis show a higher incidence of late non-fatal complications.

P-174
Off label use of an adjustable gastric banding system for pulmonary artery banding: an animal study

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Objective: Pulmonary artery banding (PAB) is proposed as a first palliation in infants with complex CHD, and high pulmonary blood flow. It may also be used to retrain the left ventricle (LV). Optimal tightening may be difficult to obtain leading to reoperation. An implantable device for PAB with telemetric control was recently developed allowing for repeated adjustment but it is presently limited to patients less than 20-kg. We aimed to test, in large animals, an adjustable gastric banding system off label for PAB.
Methods: Twelve ewes weighing 50–75 kg underwent Lap-Band implantation around the main PA through a left thoracotomy. All had functional evaluation with progressive occlusion and opening of the device at implantation, and every two weeks until sacrifice that was done immediately after implantation (group 1, $n = 6$), at 1 (group 2, $n = 3$) and 3 months (group 3, $n = 3$). Invasive pressure measurements in the right ventricle (RV) and aorta (Ao) were carried out each time.
Results: Devices could be implanted easily in all animals. Progressive occlusion and re-opening were possible in all animals during each time points. Tightening correlated with elevation of RV pressure and RV to Ao ratio. Two animals from group 3 died after 10 weeks because of severe cardiac dysfunction following inappropriate and excessive tightening of the band.
Conclusion: Using this implantable device, we were able to adjust the PA diameter in animals. Its use in humans would open the present indication in patients requiring LV retraining and weighing more than 30 kg.

(For P-175, please see OP-16)

P-176
Long-term results of truncus arteriosus repair: analysis of 138 patients

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Objective: Analysis of 138 patients of truncus arteriosus operated in our institution from 1985 to 2005.
Methods: In this period, the majority of patients (92%) with diagnosis of truncus arteriosus were repaired with two distinct techniques: conduit and directed anastomosis through the right ventricle-to-pulmonary arteries (Barbero Marcial's technique). Selected patients with truncus types I or II (Collett-Edwards classification), age less than 6 mos and, without associated anomalies or anomalous coronary artery crossing the right outflow tract were submitted to Barbero-Marcial's repair. We analyzed pre, intra and postoperative variables and compared survival and reoperations data from these two techniques.
Results: Age at operation ranged from 2 days to 4.65 years (median of 3.63 months) and weight from 2.38 kg to 12.3 kg (median of 3.85 kg) for entire group. The mean follow-up was 5.55 ± 5.64 years (range from 30 d to 18 y). Conduit repair was done in 69 patients, Barbero- Marcial's in 57, and palliative procedures in 12. There was statistically difference between age ($p = 0.001$), weight ($p = 0.007$) and cardiopulmonary bypass time ($p = 0.004$) between Barbero-Marcial and conduit group. Overall mortality rate was 31.9% ($n = 44$). Log-rank test to survival analysis showed high mortality in the palliative group ($p = 0.001$) and estimated survival analysis was 67% in the conduit group and 72% in Barbero-Marcial's technique. Cox risk analysis model revealed year of operation ($p = 0.010$), presence of interrupted aortic arch ($p = 0.041$) and ventricular fibrillation during induction anesthesia ($p = 0.002$) as risk factors for death. The estimated relative risk was 0.914 for each year, being higher in the beginning of our experience. There was no difference between age, weight, cardiopulmonary bypass time and technique regarding survival. Fifty-eight reoperations were performed in 49 patients (35.5%), 30 in the conduit group and 27 in Barbero-Marcial's group. Freedom from reoperation was 77% for conduit group and 70% for MBM group (NS).
Conclusions: Truncus arteriosus repair without extracardiac conduits can be performed in selected patients with satisfactory long-term results. Interrupted aortic arch, ventricular fibrillation after anesthesia, and year of operation were significant risk factors for death.

P-177
Right axillary incision: a cosmetically superior approach to repair a wide range of congenital cardiac defects

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Objectives: We sought to evaluate the safety of a right axillary incision, a cosmetically superior approach than anterolateral thoracotomy, to repair various congenital heart defects.
Methods: All the patients who were approached with this incision between March 2001 to December 2005 were included in the study. There were 110 patients (median age: 4.5 years) with atrial septal defect closure (52 patients), repair of partial abnormal pulmonary venous return (18 patients), partial atrioventricular canal (17 patients), and perimembranous ventricular septal defect (18 patients). The surgical technique involved peripheral and central cannulation for institution of cardiopulmonary bypass. Electrically induced ventricular fibrillation was used for defects located in front of the atrioventricular valves, and cardioplegic arrest was used for those located at the level or behind these valves.
Results: The repair was possible without need for conversion to another approach. One patient sustained a transient neurologic deficit. The patients were all in excellent condition after a mean follow-up of 18 months. The cardiac defect was repaired with no residual defect in 75 patients and with trivial residual defect in 5 patients (3 with mitral valve regurgitation, 1 with atrial septal defect, and 1 with ventricular septal defect). The incision healed properly in all, and the thorax showed no deformity.
Conclusion: The right axillary incision provides a quality of repair for various congenital defects similar to that obtained by using standard surgical approaches.Because it lies more laterally and is hidden by the resting arm, it provides superior cosmetic results compared with conventional incisions, including the anterolateral thoracotomy. Finally, the incision is unlikely to interfere with subsequent development of the breast.

P-178
The Ross-Konno procedure – an adequate approach to severe LVOT-obstruction in infants and young children?
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Background: The Ross-Konno procedure may present an attractive surgical option for the management of severe left ventricular outflow tract (LVOT) obstruction in children. However, experiences are limited. The aim of this study was to analyse the long-term results of the Ross-Konno procedure in infants and young children.
Material and Methods: Between 6/1996–1/2004, 13 consecutive patients up to 8 years of age (range: 4 days to 92 months, median: 24 months) underwent a Ross/Konno procedure at Hopital Necker Enfants-Malades. All patients had severe aortic stenosis and LVOT obstruction. 7 patients had undergone previous interventions (AVR (1), balloon valvolotomy (3), aortic arch reconstruction (1), VSD closure (2), coarctation resection (4)).
Results: Median cross-clamp time was 128 min, CPB time 190 min. 3 patients underwent additional procedures (aortic arch reconstruction (3), VSD closure (1)). Two early deaths (4 and 17 days old) at postop day 0 and 3 counting for the overall mortality of overall mortality is 15% (95% CI:0.02–0.46). Both patients showed severe annular hypoplasia and LV-dysfunction. During follow-up (median 4.3 years, range: 1–107 months) none of the patients showed recurrent LVOT-obstruction or aortic annular dilatation. No patient required additional procedures for aortic valve disease. The left ventricular function recovered within 3–6 months in all patients and no more than trivial aortic regurgitation was observed during follow-up. The autograft demonstrated somatic growth. 3 patients required a pulmonary allograft replacement after 8 years postoperatively. All patients are in sinus rhythm.
Conclusions: The Ross-Konno procedure presents an excellent approach to the aortic valve with LVOTobstruction in young children and can be performed with a low rate of morbidity, mortality, and re-operation. However, given the high early mortality rate, this procedure might not be a good option for the neonate patient in the setting of critical preoperative conditions.

P-179
The course of pulmonary arteriovenous malformation after Fontan completion
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Introduction: Conversion from cavopulmonary shunt to the Fontan circulation can improve cyanosis in patients with potential risk of development of pulmonary arteriovenous malformation (PAVM).
Methods: We selected 32 patients who have developed PAVM after cavopulmonary anastomosis. Among them, 15 patients had left isomerism with interrupted inferior vena cava (IVC). Eleven patients (34.4%) had the clinical PAVM ($SpO_2 \leqq 80\%$, group A) and 21 (65.6%) had the subclinical PAVM ($SpO_2 > 80\%$, group B) before Fontan operation. Seven patients (63.6%) among 11 with clinical PAVM had interrupted IVC. All patients have taken completion Fontan operation.
Results: One patient in group A died due to severe hypoxia immediately after Fontan operation. Follow-up catheterization was performed in all survivors at a median duration of 33 months (7–62 months). In group A, all survivors improved, and the mean oxygen saturation increased from 73.9% to 95.3%. In group B, right to left shunt through PAVM decreased or disappeared in 17 patients. Subclinical PAVM in the remaining 4 aggravated to clinical PAVM at follow up catheterization from 24 to 58 months after hepatic vein inclusion, and then 3 with severe hypoxia who underwent Fontan revision with rerouting of hepatic venous flow through a azygous vein, reversed the progression of severe PAVM.
Conclusions: PAVM including subclinical type resolved after inclusion of hepatic venous flow in the majority of our patients with or without interrupted IVC after Fontan operation. However, some patients may have persistent hypoxemia if they had streaming of hepatic vein to pulmonary artery flow to one lung.

(For P-180, please see OP-14)

(For P-181, please see OP-11)

P-182
The Contegra bovine jugular vein conduit for pediatric RVOT reconstruction: 6 year-results from the European Multicenter Trial
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Background: The Medtronic Contegra® is a glutaraldehyde fixed bovine jugular vein segment for RVOT reconstruction in congenital heart disease. We report the Contegra® European Multicenter Study (prospective, controlled) 6 year results.
Methods: Between May 1999 and January 2004, 8 institutions implanted 165 Contegras. Patient age: 2 days to 18.5 years, median 2 y, 35% <1 y. Indications: Fallot (39%), Transposition (6%), Truncus (30%), DORV (10%), Pulmonary atresia (6%), Ross (7%). 22% (n = 36) had previous conduits. Implanted sizes (mm, number, frequency): 12 mm/52/32%; 14 mm/31/19%; 16 mm/26/16%; 18 mm/10/12%; 20 mm/15/9%; 22 mm/21/13%. Standardized follow-up examinations (at 1,3,6 months, then every 6 months) included 1257 core lab evaluated echos.
Results: Development over time was not critical for: dilatation (no general trend despite 2 related explants), regurgitation (at least 85% mild or less at each follow-up interval) and gradient (mean constantly <12 mmHg). Early mortality: 10.9%. 6 year freedom-from-rates (number of patients with events): Conduit related death: 100%, catheter intervention 72% (n = 34), explantation 76% (21), any postoperative procedure (43): 60% Once a postoperative procedure was needed, 59% needed another within 1 year.

Explantation reasons: Distal pannus (7), thrombus (4), pulmonary artery branch stenoses (3), valve degeneration (3), dilatation (1), endocarditis (2), outgrowth (1). Histology (n = 17) showed calcification: absent (4), trace (5), moderate (3), extensive (5), local (1); pannus in all cases. Mild calcifications were seen in 1% of the 646 echos where this item was assessed, 99% showed no calcification.
Conclusions: The 6-year results of the European Contegra® Multicenter Study make this conduit an interesting alternative for RVOT reconstruction of congenital malformations. The results compare well with alternative devices.

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