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Cardiology in the Young

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Session 5: Basic Science

O5-1

Remodeling of the right ventricular infundibular region in tetralogy of Fallot involves proliferation and apoptosis

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Objectives: Tetralogy of Fallot (TOF) is a congenital heart disease due to a malaligned septum resulting in a ventricular septum defect, an overriding aorta, subpulmonary stenosis and right ventricular hypertrophy. In this study we attempt to caracterize the mechanisms which lead to subpulmonary tissue remodeling.
Methods: Tissue was harvested from eight patients with TOF (mean age: 5.4 years, range: 1.5–14.1 years) during cardiac surgery and was paraffin embedded or frozen in liquid nitrogen. Cells were caracterized using immunohistochemistry for troponin I, smooth muscle actin and von Willebrand factor. Differentiation was assessed with a skeletal muscle actin antibody. Real time quantitative RT-PCR, immunohistochemistry and Western blots were performed in order to examine the expression of markers of proliferation (PCNA) and apoptosis (caspase 3, Bcl-2, Bax, Bcl-xL and Bad), and various growth factors (IGF-I, IGF-II and TGF-β1 and -β2).
Results: The investigated samples contained high protein levels of troponin I and muscle actin, confirming that the majority of cells were cardiomyocytes. These cells stained positive for PCNA, indicating active proliferation. Furthermore, staining of endothelial cells for von Willebrand factor proved that the tissue was highly vascularized. Analysis of growth modulators by Western blotting showed the presence of high levels of TGF-β1 and -β2, mild staining of IGF-II and the IGF-I receptor, and absence of IGF-I. We were also able to identify the presence of caspase 3 positive cells, showing apoptosis in these samples in spite of a high level of Bcl-xL expression and a low level of Bad. Bcl-2 and Bax were negative.
Conclusions: These data indicate that the right infundibular region in patients with TOF is actively remodeling. This involves both proliferation and apoptosis and our results suggest that the process is modulated by TGF-β1 and -β2.

O5-2

Assessment of RV pump and contractile function using MRI directed cardiac catheterisation: in-vivo validation and clinical application in patients with RV pressure overload

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Objectives: In various clinical situations the right ventricle (RV) is subject to chronic pressure overload which may progress to the development of RV failure. MRI is widely considered as the gold standard to assess RV pump function, however, is limited in its ability to determine cardio-vascular pressures and RV contractility. Aim of this study is to propose a novel MRI technique in which RV volumes and invasive RV pressure are measured simultaneously for assessment of both, RV pump and contractile function.
Methods and Results: The proposed MRI technique was validated in preliminary in-vivo studies (6 swine) and then applied in 6 control and 6 patients with chronic RV pressure overload due to pulmonary hypertension (PHT). Indices of RV pump (CI = cardiac index), contractile function (Emax = maximal ventricular elastance) and ventricular-arterial coupling (Emax/Ea) were calculated from MRI derived pressure-volume loops. In-vivo validation was performed using conductance catheter techniques. All MRI studies were performed in an interventional 1.5 Tesla MRI unit. Cardiac cathetrisation for assessment of invasive pressures was performed with flow directional catheters guided under MRI control. Catheter guidance was performed successfully in all experiments. Bland Altman test showed good agreement between conductance catheter and MRI derived Emax (1.5 ± 0.4 versus 1.1 ± 0.6 mmHg/mL, $p = 0.62$). In patients with PHT (compared to control) RV pump function was decreased (CI = 2.1 ± 0.5 versus CI = 2.9 ± 0.4 L/m2, $p < 0.01$), contractile function enhanced

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(Emax = 2.8 ± 0.6 mmHg/mL vs. Emax = 1.0 ± 0.2, $p < 0.01$) and RV mechanical work production inefficient (Emax/Ea = 0.7 ± 0.3 vs. Emax/Ea = 1.4 ± 0.4, $p < 0.01$).
Conclusions: MRI directed cardiac catheterisation can be successfully performed in a clinical setting and allows for determination of RV pump and contractile function.

O5-3
Protective heat shock proteins are synthesized in the myocardium of infants with congenital cardiac defects via the MAP-kinase ERK1/2 pathway
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Background: Heat shock proteins (HSPs) are an important family of endogenous protective proteins the synthesis of which increases in response to a wide variety of stresses including hypoxia and stretch. Our study tests the hypothesis of whether preoperative myocardial synthesis of HSPs takes place in infants with congenital cardiac defects and whether it provides intraoperative protection.
Methods: Seventeen infants with tetralogy of Fallot (TOF) (n = 9) or with ventricular septal defects (VSDs) (n = 8) were investigated. The expression of HSP32, HSP70 and HSP90 was detected by RT-PCR and/or Western blotting in the samples of right atrial and ventricular myocardium taken before and during cardiac surgery, respectively and was related to postoperative end-point variables. The activation of Heat Shock Factor (HSF)-1 and of mitogen activated kinase (MAPK) ERK1/2 was determined by Western blotting.
Results: The expression of HSP70- and HSP90-mRNA was detected in the right atrial (RA) and ventricular (RV) myocardium of all patients. Concentrations of HSP32 and HSP70 in RA and RV were not different between patients with TOF and with VSD, but HSP90 ($p < 0.05$). Considering all patients, levels of HSP90 in RA correlated positively with arterial blood pressure 24 h po. (Spearman: 0.56, $p < 0.04$). Levels of HSP70 in RV inversely correlated with the amount of dobutrex administered 24 h po (Spearman: −0.53, $p = 0.09$). The activation of HSF-1 was not detected in the myocardium of any patients in contrast to the phosphorylation of MAPK ERK1/2.
Conclusions: Our results show for the first time that HSPs are synthesised in the myocardium of infants with congenital cardiac defects via the MAPK ERK1/2 pathway, overtly independently on the activation of HSF-1. Overexpression of HSPs before cardiac surgery may play a protective role postoperatively.

O5-4
Macrophage inhibitory factor (MIF) and tumor necrosis factor α (TNFα) release in primary neonatal rat cardiac myocytes is differentially regulated by inflammatory stimuli
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Objectives: There is increasing evidence that cytokines produced by cardiac myocytes are important mediators in the pathogenesis of cardiovascular disease i.e. chronic heart failure, myocarditis and sepsis-associated cardiac dysfunction. In the present study, we investigated if defined pathological conditions result in a different response in cytokine secretion in primary neonatal rat cardiac myocytes. We therefore examined the effect of oxidative stress (by hydrogen peroxide) as it occurs in ischemia-reperfusion injury and lipopolysaccharide (LPS), which is a potent inflammatory mediator in gram-negative sepsis on the release of MIF and TNFα, two cytokines with crucial immune-modulatory functions.
Methods: Aliquots of the same probe were applied to MIF and TNFa Elisas after a 6 h incubation period with H_2O_2 (0.1 mM) and LPS (10 mg/ml) isolated from different bacterial strains (E.coli0111:B4, E.coli055:B5, S.typhimurium), respectively.
Results: Upon hydrogen peroxide stimulation, there was a significant increase of MIF in the cell supernatant with a maximum at 6 h in comparison to untreated cardiac myocytes (2,17 ± 2,0 ng/ml vs. 0,38 ± 0,48 ng/ml, $p < 0.0001$). The H_2O_2 induced increase in MIF could be completely prevented by preincubation the cardiac myocytes with Catalase (100 U/ml) and α-tocopherol (1 ng/ml), respectively. In contrast, different LPS preparations did not result in an increase in MIF synthesis. However, in these samples a significant increase in TNFα production was detectable with a maximum increase for S. typhimurium (30 ± 6 pg/ml vs. < 1 pg/ml, $p < 0.0001$). No significant increase in TNFα release was found in the cells stimulated with hydrogen peroxide after 6 h.
Conclusions: Cardiac myocytes release the cytokines MIF and TNFα differently upon stimulation with inflammatory mediators. The cytokine release in cardiac myocytes differs from the response of peripheral leukocytes to LPS and oxidative radicals. Therefore, cardiac myocytes seem to have characteristic immune-modulatory properties, which might maintain cardiac dysfunction in inflammatory cardiac disease. (Supported by DFG Br1619/5-1 to B.D.)

O5-5
Acute infections could exacerbate arterial endothelial injury in children with type 1 diabetes mellitus
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Background: Type 1 diabetes mellitus (DM-1) purports an increased cardiovascular risk partly due to accelerated atherosclerosis. Alteration in the arterial endothelial function has been suggested as a mechanistic link, even if its sole connection to the diabetic milieu appears unlikely. DM-1 increases the propensity for acute infections, which, in turn, might increase as such the cardiovascular risk. In a cross-sectional study, we investigated whether clinically manifest acute infections could exacerbate the arterial endothelial damage in diabetic children.
Methods: Endothelium-dependent (flow-mediated) dilatation (FMD) and glyceril trinitrate (GTN)-induced dilatory responses of the brachial artery were measured by ultrasonography in twenty-two children with DM-1 (mean age and diabetes duration: 14 ± 3 and 6 ± 3 years, respectively) without clinical manifestations of macroangiopathy. Of these, seven patients, of similar age, weight and diabetes profile as the remaining patients, had clinical signs of upper respiratory tract infection (body temperature >38°C and symptoms) within 6–8 weeks prior to the study day. Serum levels of LDL, HDL and total cholesterol, oxidized LDL, von Willebrand factor (vWF), C-reactive protein and orosomucoid, and HbA1c were measured on the ultrasound day. Data are mean ± SD.
Results: Overall, diabetic children had subnormal FMD (5.6 ± 2%), being inversely correlated with vWF ($r = -0.4$, $p < 0.05$), CRP

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($r = -0.6$, $p = 0.01$) and LDL cholesterol ($r = -0.6$, $p < 0.01$). The infection group had significantly lower FMD and higher levels of vWF than the remaining diabetic patients (FMD: 4.3 ± 2 versus 6.2 ± 2%, respectively; $p < 0.05$; and vWF: 1.9 ± 0.2 versus 1.3 ± 0.2 IE/ml, respectively; $p = 0.01$). In contrast, no differences in the baseline brachial artery diameter, GTN-induced dilatation, age, duration and severity of diabetes, and lipid profile were noted between these groups ($p > 0.1$).
Conclusion: This is the first study to suggest that endothelial function may be further altered in diabetic children with antecedent clinically manifest acute infections. The findings warrant large-scale prospective studies to verify the interplay between acute infections and DM-1 in the pathogenesis of atherosclerosis.

Session 7: Pulmonary Hypertension

O7-1
Beneficial effects of long-term oral sildenafil treatment in children with pulmonary hypertension on pulmonary vascular resistance and exercise capacity

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Background: Pulmonary vascular disease is a progressive and fatal disease with few treatment options. Sildenafil is a pulmonary vasodilator which prevents cGMP breakdown through type 5 phosphodiesterase (PDE) inhibition. PDE5 is localized in the pulmonary vascular bed and upregulated in pulmonary hypertension (PH). Therefore, we hypothesized that sildenafil would ameliorate symptoms in children with chronic PH.
Methods: Following baseline assessment of hemodynamics by cardiac catheterization and distance walked in 6 minutes we administered oral sildenafil at 0.5–1 mg/kg four times daily to 14 children (median age 9.8 years, range 5.3–18) with PH. Diagnoses were primary PH ($n = 4$) and secondary PH ($n = 10$) due to repaired congenital heart disease (7/10) or Eisenmenger syndrome (3/10). A 6 minute walk was repeated at 6 weeks, 3, 6 and 12 months ($n = 14$). After median follow up of 10.8 months (range 6–15.3) we repeated cardiac catheterization ($n = 9$). Data regarding subjective impressions (individual energy level and quality of life) were also collected.
Results: Following sildenafil therapy distance walked in 6 minutes increased from 278 meters (range 58–461) to 432 meters (range 300–810). Overall, 6 minute walk improved significantly at 12 months compared to baseline ($p = 0.014$). A quadratic effect was demonstrated with maximal improvement between baseline and 6 months ($p = 0.004$) and a plateau was reached, without deterioration, between 6 and 12 months ($p = 0.46$). Mean pulmonary artery pressure and pulmonary vascular resistance decreased from 69.9 ± 20.6 mmHg to 55.3 ± 15.1 ($p = 0.002$) and 20 ± 11.9 Wood units m^2 to 14.5 ± 7.8 ($p = 0.03$) respectively. There was no significant change in mean arterial, right or left atrial pressures, systemic vascular resistance, heart rate or cardiac index. There was no difference in arterial pH and CO_2 between the two assessments. Reported side effects were minor. One patient died from right heart failure after 18 months of therapy following an initial response with rescue from frank right heart failure with sildenafil treatment alone.
Conclusions: Oral sildenafil significantly improves hemodynamics, exercise capacity and quality of life for up to 12 months in children with pulmonary vascular disease. These preliminary observations suggest that oral sildenafil therapy may offer an effective alternative to current treatments of childhood PH.

O7-2
Endothelium-dependent pulmonary artery relaxation in children with idiopathic pulmonary hypertension

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Background: The management of idiopathic pulmonary arterial hypertension (iPAH) remains limited by poor understanding of its pathogenesis. Endothelial injury may be an important event in the pathophysiological course. Therefore children with iPAH children with and without response to short-term vasodilator testing were investigated for their endothelial function.
Methods: Heart-catherization in 14 awake or conscious sedated children, age 0.5–18 years. Determination of the pulmonary flow reserve (PFR, maximum blood flow velocity compared to baseline) by assessing the diameter of a pulmonary segmental artery and the change of pulmonary flow velocity as a response to graded infusions of acetylcholine with an intraluminal flow-wire (flomap®, cardiometrics). The global reactivity of the pulmonary artery system was determined by a combination of an increased inspired oxygen fraction (FiO_2), Epoprostenol infusion with increasing doses ($n = 4$) or aerolised iloprost inhalation ($n = 10$).
Results: 6 patients responded to oxygen and i.v.-epoprostenol/inhaled iloprost by a decrease of the mPAP/mSAP-ratio (mean pulmonary-arterial pressure/mean systemic arterial pressure) from 0.99 (0.65 to 1.2) to 0.37 (range 0.24 to 0.56). In this group there also was in all children a dose-dependent increase in blood flow velocity in response to acetylcholine. The mean PFR was 3.7 (range 1.6 to 6.3). In the non-responder-group with a baseline mPAP/mSAP ratio of 0.98 (range 0.69 to 1.5) and an unchanged ratio of 0.96 after combined vasodilator therapy (as above), there was no PFR (unchanged blood flow velocity) in 4 patients. 3 of these patients died during follow up despite long-term epoprostenol therapy. However, the other 4 'non-resonders' did show a PFR with a 2-fold increase in blood flow velocity (range 1.33 to 3.5). All 4 remained clinicaly stable, 3 of them improved and 2 of them even changed to responders.
Conclusions: The study is the first clinical trial to show that in pediatric iPAH endothelium-dependent pulmonary artery relaxation is preserved even in some patients who do not respond to global vasodilator testing. Even more, in some children acetylcholine induced NO generation seems to be in access. This is another indicator, that iPAH in children may not be a single entity.

O7-3
Chronic endothelin – a receptor blockade lowers pulmonary vascular resistance, augments alveolar growth, and does not alter eNOS/ET-1 signaling of lambs with increased pulmonary blood flow and pressure

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Background: Alterations in endogenous endothelial function have been implicated in the pathophysiology of advanced pulmonary hypertension. In fact, chronic endothelin receptor blockade has

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recently shown efficacy in treated advanced disease. However, the effect of endothelin-1 (ET-1) in the early pathophysiology of pulmonary hypertension secondary to increased pulmonary blood flow remains unclear. Therefore the objectives of this study were to (1) study the hemodynamic effects of chronic ET-A receptor blockade in a lamb model of increased pulmonary blood flow following in utero aortopulmonary graft placement (shunt) and (2) study the effects of ET receptor blockade on endogenous eNOS/ET-1 signaling.
Method and Results: Immediately after spontaneous birth, shunt-lambs were treated lifelong with either an ET-A receptor antagonist (PD 156707, 150 mg/kg) or placebo. At 4-weeks age PD156707-treated shunt-lambs had significantly decreased left pulmonary vascular resistance (76.5 ± 34.1 vs. 118.1 ± 32.0 mmHg/ml/min/kg, $p < 0.05$) and mean right atrial pressure (4.0 ± 1.2 vs. 5.4 ± 0.8 mmHg, $p < 0.05$) compared to placebo-treated shunt lambs ($n = 8$). All other hemodynamic variables did not differ between the two groups (mean pulmonary arterial pressure, mean left atrial pressure, mean systemic arterial pressure, ratio of pulmonary to systolic pressure, Qp, Qp : Qs, heart rate, pH, and hemoglobin). Smooth muscle thickness and distribution and number of arteries per unit area where not different by morphometric analysis. However, the number of alveoli per unit area was significantly increased in PD156707-treated shunt-lambs (190.7 ± 5.6 vs. 132.9 ± 10.0, $p < 0.05$). All measured eNOS/ET-1 signaling variables (plasma ET-1 and cGMP, tissue NOS activity and eNOS, ECE-1, ET-A and ET-B receptor protein levels) except plasma NOx did not differ between the two groups. Plasma NOx was significantly decreased in PD156707-treated shunt-lambs (6.7 ± 1.8 vs. 11.1 ± 1.5 μM, $p < 0.05$).
Conclusions: Chronic endothelin-A receptor blockade lowers pulmonary vascular resistance, augments alveolar growth, and does not alter eNOS/ET-1 signaling of shunt-lambs. The exact mechanisms need to be clarified. The decrease of NOx in the plasma may be due to an increased clearance of NOx by the kidneys. Note: This Abstract has been presented as a poster presentation at the Scientific Sessions 2003 of the AHA.

O7-4
Severe pulmonary arterial hypertension: experience of treating children with continuous intravenous Epoprostenol

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Introduction: Pulmonary arterial hypertension (PAH) is a disease characterised by elevation of pulmonary artery pressure, and pulmonary vascular resistance (PVR), leading to right ventricular failure and death. Median survival without treatment is 2.8 years (Barst RJ. 1994). Prostacyclin (Epoprostenol) is said to improve quality of life and survival (Barst RJ. 1996).
Methods: We reviewed 24 severely symptomatic children aged 3 months to 17 years, treated with Epoprostenol. Nineteen patients had Primary and 5 Secondary PAH. Mean age at initiation of treatment was 6.6y (3m–17y) [Mean age of diagnosis was 4.6 years]. All children, except two were in WHO class IV. Twelve patients were already receiving treatment, 5 with Bosentan, 3 with Sildenafil, 2 with Calcium channel blockers and 2 with both Bosentan and Sildenafil. At cardiac catheterisation mean PAP approached or exceeded SAP in all. Baseline PVR exceeded >11 Wood units $\cdot m^2$ in 90%. PVR was fixed in all children, there being no response to vasodilator testing, including nitric oxide. Epoprostenol was given for 2m–6y (mean 21 months), at a mean dose of 30 ng/Kg/min (7–60 ng). Patients were uptitrated as/when necessary.
Results: Children were monitored non-invasively. After a mean follow up of 21 months, 80% have showed clinical improvement and a beneficial shift in WHO classification, from late class III/IV to II/III (table 1). One is on active transplantation list. Three have died after a mean of 2.5 years of treatment. None had serious side effects necessitating modification of treatment. Commonly observed side effects included flushing, jaw pain and non-specific abdominal pain. Although site infection was common (70%), none had septicaemia. One Hickman line was replaced because of persisting site/tunnel infection.

Table comparing WHO functional Class at the initiation of treatment and at present

	Class I	Class II	Class III	Class IV
Before Epoprostenol	0	0	2	22
After Epoprostenol	0	2	17	2

Conclusions: Treatment with continuous intravenous Epoprostenol improved exercise tolerance, quality of life and survival. All returned to school/nursery. Early diagnosis, prompt referral to a specialist PH centre and prompt use of specific therapies are crucial. Response to treatment needs constant review and families need extensive support.

O7-5
Role of atrial septostomy in the treatment of children with pulmonary hypertension

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Background: Atrial septostomy is used to treat patients with Primary Pulmonary Hypertension suffering from syncope and or significant right ventricular failure despite optimal chronic vasodilator therapy. Experience is limited, particularly in children. Issues requiring clarification include optimal timing of the intervention, associated morbidity and mortality, long-term efficacy.
Objectives: To assess safety and efficacy of atrial septostomy in 12 children with severe Pulmonary Arterial Hypertension (PH).
Methods: Twelve children were reviewed retrospectively. All received specific treatments for PH, 11 having Primary and 1 Secondary PH. Mean age at septostomy: 8.4 years (range: 3 months–17 years). Balloon septostomy was carried out in 9 cases (including one with a fenestrated device and one with an interatrial stent), blade septostomy in 3.
Results: Before septostomy. Syncope was present in 7; all had severe exercise intolerance and were WHO late Class III and IV. RV failure was present in 6 children. Mean systemic arterial oxygen saturation (SaO_2): 95%. ECG showed right axis deviation and RV hypertrophy in all with a strain pattern in 65%. Echocardiography showed a tricuspid regurgitant jet velocity >4 m/s in 58% and RV dysfunction in 66%. Cardiac catheterisation: Mean RAP was 7 mmHg. All had mean PAP approaching or exceeding SAP. Mean PVR was 20.7 U $\cdot m^2$ and unresponsive to vasodilators in all cases. Septostomy was performed at the initial cardiac catheterisation in 7 cases. After septostomy. The procedure was always uneventful. At mean follow-up of 2.5 years (range 1 month–5.5 years), 11/12 children are still alive. All these children have improved by at least one WHO class and none had further syncope. Interestingly, 5 out of 6 patients with RV failure improved, however only 3 of these children showed echocardiographic improvement in RV systolic

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function. Mean SaO_2 decreased by 7%. The atrial communication closed in 2 children who had had a blade septostomy, necessitating a repeat procedure.

WHO Classification before and after septostomy

	Class I	Class II	Class III	Class IV
Before septostomy	0	0	5	7
After septostomy	0	3	8	1

Conclusions: Atrial septostomy improved symptoms and quality of life in a group of children deteriorating with severe PH. It was a safe procedure. It is to be recommended in severely symptomatic children, before they become critically ill. Fenestrated devices might help ensure longer patency of the atrial communication.

O7-6
Inhaled iloprost and rolipram, a phosphodiesterase 4 inhibitor, in acute hypoxic pulmonary hypertension

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Inhaled iloprost is an effective therapy for pulmonary hypertension (PHT). However a major drawback is its short term pulmonary vasodilating effect lasting for only 60 min. Iloprost effects are mediated through the production of cAMP that is catabolized by various phosphodiesterases (PDE). Thus blocking the effect of these enzymes with PDE inhibitors may prolong and/or amplify the vasodilatory effect of iloprost. We investigated the effects of inhaled iloprost alone and in combination with rolipram, a specific PDE4 inhibitor that preferentially hydrolyze cAMP, in a pig model of hypoxic PHT.
Material and Methods: Stable hypoxic pulmonary hypertension was established by keeping F_iO_2 between 0.1–0.14. Ventilatory parameters and pH were kept constant to avoid confounding factors. The animals were kept sedated and paralyzed for the entire study. Two groups of 4 pigs were studied. After stable hypoxia a first group (Ilo) received a 15 microgr iloprost inhalation over 15 min and was hemodynamically monitored. A second group (Ilo + Rol) received the same dose of iloprost but with simultaneous administration of 15 microgr/Kg IV rolipram over 15 min and was monitored. Complete hemodynamics were measured at baseline (B), on stable hypoxia (H), at the end of drug delivery, and after 15 min, 30 min, 1, 2 and 3 hours.
Results: Results are presented as pulmonary to systemic vascular resistance ratio (PVR/SVR) changes.

	B	H	End of ttt	15 min	30 min	1 h	2 h
Ilo	0.13 ± 0.04	0.47 ± 0.01⋆	0.22 ± 0.03#	0.36 ± 0.14	0.32 ± 0.14	0.32 ± 0.03	0.34 ± 0.05
Ilo + rol	0.13 ± 0.03	0.35 ± 0.07⋆	0.21 ± 0.06#	0.27 ± 0.06	0.32 ± 0.05	0.32 ± 0.05	0.29 ± 0.07

⋆$p < 0.001$ versus baseline, #$p < 0.05$ versus hypoxia (Repeated measures ANOVA).

Conclusions: As expected inhaled iloprost alone significantly decreased PVR/SVR ratio. A significant effect was observed only at the end of inhalation. The addition of rolipram did not prolong or increase significantly the effect of inhaled iloprost in this animal model of PHT. Further investigations with PDE inhibitors allowing for prolonged efficacy of inhaled iloprost in acute hypoxic PHT should focus on other PDE inhibitors, dose effective studies, and/or different modes of drug delivery.

Session 8: Arrhythmia

O8-1
Cardiac resynchronization as therapy for congestive heart failure in children with congenital heart disease

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Cardiac resynchronization improves short-term and long-term hemodynamics in adult patients with congestive heart failure and left bundle branch block. In children, only two cases of permanent biventricular (BV) pacing have been reported.
Objectives: To evaluate clinical and echocardiographic outcome of biventricular (BV) pacing as therapy for congestive heart failure in children with congenital heart disease.
Patients and Methods: 6 consecutive children underwent BV pacing at a median age of 18 months, range 3 months to 13 yrs. Five pts had conventional pacemaker therapy for atrioventricular (AV) block and one pt had left bundle branch block. Five pts had underwent biventricular repair for complex congenital heart disease, 1 pt had congenital complete AV-block and dilated cardiomyopathy. A transvenous BV system was implanted in 1 pt (6 yrs) and epicardial systems were used in 5 pts. The optimal LV pacing site was assessed by TEE during the procedure. Clinical parameters, ECG and echocardiographic data including Tissue Doppler Imaging (TDI) were monitored during follow-up.
Results: No complication occurred during the procedures. In all 6 patients the ECG showed reduction of the QRS width and M-Mode registration or TDI imaging (septal-to-lateral delay) showed improved resynchronization. Median follow-up was 16 months, range 1 to 26 months. No patient died during follow-up. Significant acute clinical improvement was observed in 5 pts with increased exercise tolerance and catch-up growth during follow-up. Diuretic therapy was reduced or stopped in 5 pts. The mean EF improved from mean 25% (SD ± 4) to 37% (SD ± 5) ($p < 0.05$). LV enddiastolic diameter decreased in 4 pts.
Conclusions: Cardiac resychronization is effective as therapy for congestive heart failure in children with congenital heart disease. BV-pacing should be considered as alternative to conventional RV-pacing in postoperative children with left ventricular failure and AV-block.

O8-2
Catheter cryoablation of tachyarrhythmias in children and adolescents

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Catheter ablation using cryoenergy has emerged as a novel technology for ablation of various arrhythmias in adults and has been considered safe and effective, especially for AVNRT. However, data in pediatric patients with smaller and immature hearts are lacking. We describe our initial single center experience with catheter ablation in children and adolescents.
Methods: After confirming the arrhythmia mechanism by a standard EP study, a two step ablation procedure was performed using a 4-mm tip 7 French cryocatheter (CryoCath Technologies Inc, Montreal, Canada). The first step, called cryomapping, consists in cooling the catheter to −30°C. At that temperature, reversible loss

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of the electrical activity of the anatomic substrate allows us to assess the efficacy and safety of the potential ablation site. If the site is inadequate, passive rewarming of the catheter results in full recovery of the electrical activity. If the site is adequate, the second step, called cryoablation, is performed and consists in cooling the catheter to −70°C for 4 minutes to create a permanent lesion.
Results: Thirty two cryoablation procedures were done in a pediatric population aged 17 ± 4 years (range; 6–21). Mean weight and height were 60 ± 16 kg and 166 ± 17 cm respectively. There were 20 AVNRT, 10 AVRT and 2 VT. The mean procedure time and fluoroscopic time were 140 ± 72 min and 35 ± 47 min respectively. Median of cryomapping and cryoablation was 4 (range; 1–24) and 3 (range; 1–19) respectively. Acute success was achieved in 81% of all procedures and 85% in AVNRT procedures. There were no significant adverse effects and particularly no permanent AV nodal block.
Conclusion: Catheter cryoablation of tachyarrhythmias in children and adolescents appears to be safe and effective.

O8-3
Left ventricular apex pacing results in excellent hemodynamical performance in children with narrow QRS-complex
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Introduction: The right ventricular apex (RVA) is the conventional ventricular pacing site, despite the fact that RVA pacing acutely and chronically depresses left ventricular (LV) contractile function in children and adults. The present study investigates whether pacing the left ventricle results in better hemodynamic performance than RVA pacing in paediatric patients.
Methods: In 10 children (median age 2 years and 6 months, range from 2 months to 17 years) that underwent cardiac surgery for congenital heart disease with normal systemic LV anatomy and conduction (aortic stenosis, tetralogy of Fallot, atrial and ventricular septum defects), high-fidelity electrocardiographic (ECG), LV and arterial pressure measurements were performed during sinus rhythm and epicardial RVA, LV apex (LVA) and LV lateral free wall (LVFW) pacing. Pacing was performed after complete recovery from cardiopulmonary bypass. In order to avoid fusion beats caused by intrinsic activation, all children were paced in the DDD-mode with a short atrioventricular (AV) interval (50% of the intrinsic AV-delay).
Results: LVdPdtmax and Pulse Pressure decreased significantly during RVA (−10.4 ± 6.4% and −9.8 ± 8.5%, respectively) and LVFW pacing (−9.6 ± 6.0% and −8.1 ± 7.0%, respectively) as compared with sinus rhythm, but remained at sinus rhythm level during LVA pacing (−1.7 ± 6.8% and −1.6 ± 9.5%, respectively). LVdPdtmax was significantly higher during LVA than during RVA and LVFW pacing. Hemodynamic differences between pacing sites occurred without significant differences in LV end-diastolic pressure. QRS-duration was significantly longer during LVFW pacing (130 ± 18 ms) as compared with sinus rhythm (96 ± 9 ms), whereas RVA pacing (120 ± 26 ms) and LVA pacing (120 ± 29 ms) did not change QRS-duration significantly.
Conclusion: In children, LV apex pacing is superior to RVA and LVFW pacing and maintains cardiac pump function at sinus rhythm level. Therefore, the LVA appears to be an optimal pacing site. QRS-duration is a poor predictor of the optimal ventricular pacing site. Further studies are indicated to evaluate whether the beneficial effect over RVA pacing is maintained in chronically LVA-paced children.

O8-4
Detection of ventricular asynchrony using the novel tissue Doppler imaging in TOF patients following corrective surgery
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Background: In long term follow up TOF patients may have reduced right and/or left ventricular function. Recent studies showed that reduced ventricular function may be related to intra- or inter-ventricular asynchrony. The aim of this study was to investigate weather postoperative TOF patient have intra and or inter-ventricular asynchrony using the novel tissue Doppler derived strain.
Material and Methods: Twenty five postoperative asymptomatic TOF patients with a mean age of 27 ± 19 years were examined with tissue Doppler imaging (TDI) after mean postoperative period of 18 years. The data were compared to that from 25 aged matched control subjects. The myocardial TDI derived strain curve at the basal and middle segments in RV free wall, interventricular septum (IVS) and LV lateral wall were obtained through apical four chamber view. The time interval from the Q wave of associated ECG to the start of the strain curve (Ts) was measured in 3 consecutive cardiac cycles. To assess the LV systolic asynchrony, the Ts standard deviation (SDLV) from the examined four segments in LV lateral wall and IVS in each patient was calculated. The greater the value SDLV, the more severe the intra-LV systolic asynchrony. By a similar way the intra-RV (SDRV) and the inter-ventricular (SDRV-LV) standard deviation were quantified.
Results: There was a significant difference between the TOF patients compared to the control group regarding the SDRV ($p = 0.001$) and SDRV-LV ($p = 0.035$). There was no significant difference between both groups regarding the SDLV. Neither the SDRV nor the SDRV-LV correlates with the QRS duration.
Conclusions: In asymptomatic TOF patients, intra-RV and interventricular asynchrony can be detected using tissue Doppler imaging derived strain rate. The observed asynchrony seems not to be related to the right bundle branch block induced electrical delay.

O8-5
Detrimental effect of long-term apical right ventricular pacing, on left ventricular dyssynchrony, morphology and function, in patients with congenital heart block
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Although, dual-chamber pacing improves cardiac function in patients with congenital heart block (CHB) by restoring physiological heart rate and atrio-ventricular synchronization, the long-term detrimental effect of asynchronous electromechanical activation induced by apical right ventricular pacing (RVP) has not been well clarified. We assessed in such patients consequences of long-term permanent RVP on exercise performance and left ventricular (LV) function, morphology and dyssynchrony.
Methods: Twenty-one CHB adults (23 ± 5 years, 62% male), with a DDD transvenous pacemaker, underwent 1] before implant, a

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conventional echocardiography 2] after at least 5 years of RVP, an exercise testing and an echocardiography coupled with Doppler tissue imaging (DTI) and tissue tracking. They were compared with 30 age sex and weight-matched healthy controls.
Results: After 8 ± 3 years of RVP, CHB adults had significantly higher values vs controls in terms of intra-LV asynchrony (respectively 59 ± 18 vs 19 ± 11 ms; $p < 0.001$), extent of LV myocardium displaying delayed longitudinal contraction (39 ± 15 vs 10 ± 7%; $p < 0.01$) and septal-to-posterior-wall-motion-delay (87 ± 23 vs 18 ± 9 ms; $p < 0.01$). The ratio of early-activated septal to late-activated posterior wall thickness was lower before vs after implant (1.1 ± 0.2 vs 1.3 ± 0.2; $p = 0.05$) and vs controls (1 ± 0.1; $p < 0.05$). The percentage of patients with increased LV end-diastolic diameter was higher after long-term RVP than before implant and controls (62 vs 15%; $p < 0.05$; vs 0% $p < 0.01$). CHB patients with long-term RVP had a lower cardiac output vs controls (3.8 ± 0.6 vs 4.9 ± 0.8 l/mn; $p < 0.05$) and exercise performance (123 ± 24 vs 185 ± 39 watts; $p < 0.001$).
Conclusions: In CHB patients with long-term endovenous RVP, asynchronous LV activation induced asymmetrical hypertrophy associated with an increased LV dilatation, an LV function impairement and exercise performance lowering. Alternative sites of ventricular pacing should be investigated to preserve ventricular synchrony and LV function in patients requiring long-term permanent pacing.

O8-6
Cryoablation of accessory atrioventricular pathways in children and adolescents: usefulness of cryomapping
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Introduction: Recently, a new energy source using cryoenergy has evolved as a safe and effective alternative for ablation. Theoretically, two specific features of this technique make it very attractive for pediatric patients: prediction of the effect of energy application by creating reversible block of conduction (cryomapping) and avoidance of dislodgment of the catheter by freezing of the catheter tip to the endocardium therefore minimizing the risk of complications and the time of fluoroscopy. The aim of the study was to analyse the predictive value of cryomapping in pediatric patients.
Patients and Methods: Since July 2003 11 patients (mean age 9.1 years) underwent electrophysiological study due to symptomatic supraventricular tachycardia based on an accessory pathway (AP) under the guidance of the Loca Lisa® system. After identification of the arrhythmia substrate cryomapping was performed using the Freezor® 4 mm or Freezor Xtra® 6 mm tip catheter with a target temperature of #30°C for a maximum of 60 seconds. If an acute interruption of the AP was achieved cryoenergy was delivered at the same spot at #75°C for a total of four minutes (cryoablation).
Results: In all patients studied cryoablation of the AP was finally successful. However, only in 9 patients interruption of the AP was achieved during cryomapping. In the remaining 2 patients cryomapping was unsuccessful. However, cryoablation resulted in AP interruption within 30 seconds at the identical location verified by the Loca Lisa® system. A median of 3 (1–10) cryoablations were delivered. No complications occurred in all patients studied.
Conclusions: According to our preliminary results cryomapping was predictive in only 9 out of 11 patients studied. The results of the remaining two patients may in part be explained by a smaller and therefore insufficient lesion size during cryomapping and/or a deeper location of the accessory pathway.

Session 9: Cardiac Surgery

O9-1
Paralysis of the phrenic nerve as a significant risk factor for failing Fontan hemodynamics
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Objectives: The introduction of the Fontan operation for single ventricle physiology was based on the dual principle of the pulmonary blood flow. It is postulated that normal breathing movements are necessary for passive blood flow into the lungs. We compared patients with and without diaphragmatic paralysis regarding the total cavopulmonary connection (TCPC) hemodynamics.
Methods: We analyzed 85 consecutive patients, who were available for follow up after completion of their TCPC circulation between 2/1992 and 2/2003. The median age at TCPC completion was 4.3 (range, 1.3–37) years. Sixty were operated on with an extracardiac conduit and 25 with lateral tunnel. The diagnosis of diaphragm paralysis was made by ultrasound, heart catheterization (n = 50, all patients with diaphragm paralysis) and X-ray examination. Surgical diaphragm plication was performed in 12 patients (2 before and 10 after TCPC) at a median of 2.2 years after the diagnosis.
Results: Twenty-one patients developed fixed palsy of the phrenic nerve during a total of 225 operations. There were no differences in the incidence of phrenic nerve paralysis between children under 3 years of age (n = 22) and older patients or between the extracardiac and intracardiac Fontan procedures. There were no significant differences in the early postoperative course regarding the duration of mechanical ventilation; however, prolonged pleural effusions and a hospital stay longer than 2 weeks were noted more frequently in patients with palsy ($p < 0.05$). During the median follow up of 4.6 (range 0.7–11.4) years significantly more patients with phrenic nerve palsy developed chronic ascites compared to those without palsy (8 of 21 vs. 2 of 65; $p < 0.001$). The median mean pulmonary artery pressure was higher in patients with paresis compared with those without (RPA: 10 vs. 12.5 mmHg and LPA: 10 vs. 15 mmHg; $p < 0.05$). Angiographically slower flow through the conduit or tunnel and flow redistribution away from the affected side were observed in patients with phrenic nerve palsy.
Conclusions: Phrenic nerve palsy was recognized as a significant risk factor for failing Fontan hemodynamics. It remains to be shown whether early diaphragm plication is favorable to optimize Fontan hemodynamics. In patients with preexisting diaphragm paralysis the completion of TCPC should be discussed critically.

O9-2
Age dependence of the innate immune response in children in relation to cardiovascular surgery with and without Cardiopulmonary Bypass (CPB)
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Objective: Children with congenital heart disease frequently undergo corrective cardiac surgery with CPB. CPB induces a systemic inflammatory response that relates to postoperative complications. Postoperative morbidity and mortality is increased in young children an neonates. The aim of the study was to characterise age dependence of the innate cellular immune response to CPB.

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Methods: Children (age groups: 0–28 d; 29–365 d, 1–5yrs; 6–10yrs; 11–16 yrs) undergoing surgery with (n = 72) or without CPB (n = 37) were enrolled. Peripheral blood was drawn 1 d preoperatively, after anaesthesia, at CPB onset, at reperfusion, 4 h, 1 d, 2 d after surgery, at discharge and 3 months postoperatively. Phenotype, cell count and degree of activation was determined by flow cytometry. Leukocytes were stained with monoclonal antibodies for neutrophils, monocytes, T-, B- and natural killer (NK) cells. In addition CD11a, CD11b, CD16, CD18 and HLA-DR expression were determined as measures for the degree of neutrophil and monocyte activation.
Results: Surgery with and without CPB induced except in neonates neutrophil and monocyte count increase starting at reperfusion. This increase was age dependent ($p < 0.001$). With CPB neutrophil and monocyte count elevation was more sustained than without CPB ($p < 0.05$). Activation marker expression decreased similarly and age independently with and without CPB and reached baseline at discharge. T- and B-lymphocyte count decreased during surgery both with and without CPB and reached baseline at discharge. In contrast, NK cell count increased 2 to 3-fold during reperfusion in the CPB group but remained unchanged without CPB ($p < 0.01$). Baseline was reached immediately after surgery. In contrast to the monocytes and neutrophils these reactions were age independent.
Conclusions: Cardiovascular surgery induces an innate immune response that becomes more pronounced with age. In neonates this response is marginal, probably due to pool limited for neutrophils an monocytes that can be mobilised. This reduced responsiveness could contribute to increased risk for infections and post-operative complications in small children. Because NK-cell mobilisation is the only specific response to CPB it is therefore indicative for or may even be involved in the reactions to the foreign surfaces.
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O9-3
Location of tracheobronchial compression in 108 children with congenital or acquired heart disease. A prospective blind controlled study
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Aim of Study: To evaluate the impact of heart disease on the tracheobronchial tree in children undergoing surgical correction of congenital or acquired cardiac anomalies.
Method: A flexible bronchoscopy was performed through the endotracheal tube during anesthesia for the surgical treatment (n = 108). The examiner was kept unaware of the child's diagnosis and the planned surgery. All examinations were recorded on videotapes for a secondary check. The anomalies encountered from the tip of the endotracheal tube to the lobar bronchi were described. Location and severity were evaluated with a specially designed analog visual scale to improve accuracy of evaluation. A matched control group was obtained from non-cardiac patients (n = 42) undergoing non thoracic procedures.
Results: In the control group, 18 patients had slight airway compressions: 18 had a 10% compression (14 on the left main bronchus and 4 on the lower trachea) and one patient had a 20% sus carinal compression. Therefore, compressions in the study group were considered abnormal if greater than 10% in the left main bronchus, greater than 20% in the trachea and any compression in the right main bronchus. In the study group, airway structural anomalies were found in 8 children (7%): 1 situs inversus, 2 left and 3 right isomerisms and 2 tracheal right superior lobe bronchus. External airway compressions were found in a surprisingly high percentage of patients. The most frequent obstructions were located in the left main bronchus: 71 patients (66%) had a compression ranging from 20% of the lumen to complete obstruction. Different pattern have been recognized and are described. Obstruction of the right main bronchus was found in 16 patients (15%) ranging from 10 to 80 percent of the lumen. Obstruction of the lower trachea was found in 4 patients but the trachea investigation was limited to its distal portion by the endotracheal tube position.
Conclusions: We disclosed a high incidence of left main bronchus external compressions in children with cardiopathies with a full spectrum of severity, from normal to complete obstruction. These anomalies may contribute to pulmonary complications in the pre- and post-operative period, such as atelectasis or lung infections. Airway external compression should be considered and searched actively when pulmonary complications occur in patients with cardiac anomalies.

O9-4
Closed or restrictive foramen ovale in hypoplastic left heart syndrome: a significant risk factor?
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Although surgical results in children with hypoplastic left heart syndrome (HLHS) have substantially improved, there is still a significant mortality and morbidity. A restrictive or absent foramen ovale (FO) has been shown to cause significant impairment of pulmonary vessels. The purpose of this study was to compare outcome in patients with HLHS with restrictive or closed FO ovale to those with unrestrictive FO to test the hypothesis, that a restrictive FO is a significant risk factor.
Patients and Methods: Between 1997 and 2004, 60 consecutive newborns with HLHS and intention to treat were admitted to our institution. In 14 patients (23%) a restrictive FO (<3 mm with a mean gradient of 10 mmHg or more by Doppler) was found (Group A). FO was completely closed in 3 of them. Group B consisted of 46 newborns with typical HLHS and non-restricitive FO. Both groups were comparable in terms of gestational age at birth, birthweight, sex, type of Norwood operation (RV/PA conduit, modified BT shunt) and length of follow-up.
Results: are shown in Table I. Despite prenatal diagnosis and postnatal emergency treatment all 3 patients with closed FO died immediatly after birth prior to surgery due to severe hypoxemia and pulmonary lymphangiectasia. There was no significant difference in survival between both groups, however patients of group A required significantly more and longer intensive care therapy after the Norwood operation and also after the modified Glenn procedure. Five patients with restrictive FO had procedures performed prior to stage I Norwood (surgical atrioseptectomy: 1, ballon atrioseptostomy: 4) to relieve pulmonary venous congestion. Patients of group A also had significantly more days of ventilation and days of NO therapy.

Table I. Group A: patients with HLHS and restrictive/closed FO; Group B: patients with HLHS without restrictive FO; values given as median (range).

	Group A (N = 14)	Group B (N = 46)	p-value
Pre-operative death	3	3	
Death after Norwood I	1/11	16/43	
Death between Stage I and II	0/10	4/27	
Death after Glenn	2/8	1/22	
Death after Fontan	0/2	0/8	
Median days in ICU (Norwood)	22,0 (9–31)	12,0 (6–35)	0,008
Median days to discharge (Norwood)	38,5 (21–63)	30,5 (15–53)	0,02
Median days in ICU (Glenn)	3,0 (1–65)	2,0 (1–7)	0,008
Now alive	8/14	22/46	

Conclusions: A closed FO in patients with HLHS was associated with preoperative death despite emergency treatment. Patients with restrictive FO had more postoperative (respiratory) problems, which could be handled with more and longer intensive care therapy. Despite there was no difference in short time survival, further studies must be performed to assess long term outcome. Speculations: fetuses with HLHS and closed FO could be considered as possible candidates for in-utero atrioseptostomy to relieve pulmonary venous congestion and to prevent pulmonary lymphangiectasia.

O9-5

The International Nomenclature Project for Paediatric and Congenital Heart Disease: goals achieved for global comparison in paediatric cardiology and cardiac surgery

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Outcome analysis of paediatric cardiac interventions among physicians, centres and countries is of major importance. We wish to report progress towards this goal relating to 4 domains: **1. common nomenclature, 2. standardized complexity score, 3. international data base, 4. data control:**

1. At the 1st. International Summit on Nomenclature for Congenital Heart Disease in Toronto 2001 it was decided to cross-map the Short and Long lists of the EPCC and the EACTS/STS-Nomenclature. Since then the short list x-map has been published and 4 meetings have been held by the International Nomenclature Working Group to map the Long Lists and develop a single "super-tree", the **International Paediatric and Congenital Cardiac Code (IPCCC)**, that will be presented at the next World Congress 2005 in Argentina.
2. A new method has been developed to stratify complexity of the diverse congenital heart surgery procedures: the **Aristotle score**. This approach is based on the opinions of a group of internationally recognised surgeons and centres. It allows stratification of procedures according to mortality, morbidity, and surgical difficulty into six categories of complexity.
3. The **EACTS Congenital Heart Defects Database** was initiated in 2000 to compare data on outcome of patients throughout the world utilizing the identical nomenclature and database used by the STS. To date, 114 centres from 47 countries and four continents have registered in the EACTS congenital database and more than 19,313 procedures in 18,546 patients were collected. During the same time period the **STS congenital database** has analyzed 16,920 procedures in 15,207 patients.
4. Development of data validation protocols is work in progress, but represents the most challenging hurdle. In the UK, nationwide validated data collection is routine, but current coding in this system is insensitive.

Conclusions: Significant progress has been made towards an international outcomes analysis system, particularly in three of the four areas above. However, much work remains to be done, especially involving data and complexity factor validation.

O9-6

Acquired coronary fistulae in the postoperative period following correction of Tetralogy of Fallot

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Background: Acquired coronary artery fistulae are known to occur after the repair of Tetralogy of Fallot (TOF). The incidence of these lesions has not been prospectively evaluated and its implications are ill defined. The objectives of our study were to detect coronary fistulae in the postoperative period and to study its effects.
Patients and Methods: All consecutive Children undergoing surgical repair of TOF between October 2001 and November 2003 were enrolled in to this study. Pre operative Echocardiograms were performed in all to delineate morphology of the defect and to define the coronary anatomy. All patients had serial echocardiograms in the immediate postoperative period, specifically to look for coronary artery fistulae. The first scan was done soon after the patient returning from theatre. Subsequent scans were done when required, but not beyond 48 hours of the preceding scan. Serial daily ECGs were performed in all. Details regarding period of ventilation, ITU stay, complications, inotropic support, interventions and other complications were documented.
Results: Seventeen patients underwent complete repair of TOF during the study period. None were palliated with BT shunt. Eight were females (47%). The mean age at operation was 5.1 months (1–12). Eight patients required transannular patch. The average bypass time was 92.4 min (55–180) and cross clamp time 64.5 min (range 30–104). 64% of the children had right ventriculotomies and 11 (82%) had right ventricular myomectomy. There was no immediate hospital mortality. Continuous flow from coronary artery to the right ventricular outflow tract were demonstrable on colour flow mapping in 4 children (23.5%) All had transannular patch, right ventriculotomy and RVOT resection. There was no haemodynamic compromise to warrant intervention. In patients with fistulae, there was significant difference in the duration of ventilation 6 days (4–8) Vs 2.07 days (1–5), PICU stay 7 days (5–9) Vs 2.9 days (1–7) and duration of inotropic support 6.5 days (5–8) Vs 2.38 days, (0–7) when compared to those without fistula.
Conclusions: Our study has shown a high incidence of coronary artery to RV fistula following complete repair of TOF. Hypertrophied right ventricle with prominent coronary arteries exposed after RVOT resection may account for this. It has deleterious effect on the immediate postoperative management.

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Session 10: Interventional Cardiology I

O10-1

Determinants of cardiopulmonary functional improvement after transcatheter atrial septal defect closure in asymptomatic adults

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Objectives: To evaluate the course of cardiopulmonary function after transcatheter atrial septal defect (ASD) closure and to identify the physiopathologic mechanisms leading to this change.
Background: Conflicting reports exist on cardiopulmonary functional improvement in asymptomatic adults after transcatheter closure of secundum ASD.
Methods: 32 consecutive adults (13 males; age 42.6 ± 16.7 years) underwent maximal cardiopulmonary exercise testing and transthoracic echocardiography both the day before and 6 months after transcatheter ASD closure. Mean pulmonary artery pressure, pulmonary-to-systemic flow ratio (Qp:Qs) and ASD diameter were measured before closure.
Results: Peak oxygen uptake (VO_2; $p < 0.001$), peak O_2 pulse ($p = 0.0027$) and vital capacity ($p = 0.0086$) improved after ASD closure, but not peak heart rate. A significant correlation was found between peak VO_2 improvements and Qp:Qs ($p = 0.0013$). Left ventricular ejection fraction (LVEF; $p < 0.0001$) and left ventricular end-diastolic diameter (LVEDD; $p < 0.0001$) significantly increased after 6 months, but not left ventricular end-systolic diameter. Right ventricular long and short-axis dimensions decreased (both $p < 0.0001$). Peak VO_2 and of peak O_2 pulse improvements correlated with both LVEF ($p = 0.0009$ and $p = 0.0019$, respectively) and LVEDD ($p < 0.0001$ and $p = 0.032$, respectively) increments. The decrease of both long and short-axis right ventricular dimensions positively correlated with both LVEF and LVEDD improvements. The improvement in LVEF correlated with Qp:Qs ($p = 0.0026$).
Conclusions: Transcatheter ASD closure leads to a significant improvement in cardiopulmonary function within 6 months, via an increase in peak O_2 pulse. An increase in left ventricular stroke volume and in cardiac output due to a positive ventricular interaction is the mechanism leading to an improved peak VO_2.

O10-2

Steps towards the percutaneous replacement of atrioventricular valves: an experimental study

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Background: Percutaneous valve replacement has recently been introduced and early clinical experience has been reported. To date, this technique is, however, limited to the replacement of pulmonary and aortic valves in selected patients. Here, we report our experience of percutaneous replacement of the tricuspid valve in animals.
Methods and Results: A newly nitinol stent, forming two large disks separated by a cylinder with a diameter of 18-mm, was specially designed for the purpose of the study. An 18-mm bovine valve was mounted in the central part of the stent. A PTFE membrane was finally sutured onto the ventricular disk. Ten ewes were, equally, divided into two groups according to the lasting of sacrifice time points (Group 1: one hour, Group 2: one month). 9 out of 10 devices were successfully delivered in desired position. In one animal, the device was trapped in tricuspid cordae leading to incompletely opening of the device. A trivial paravalvular leak was noticed in one animal from group 2 due to down-sized device as compared with annulus size. Mean right atrial pressure increased from 5 to 7-mmHg and did not change during the follow-up. At autopsy, examination confirmed the good position of the devices in successfully implanted animals.
Conclusions: Implantation of a semi-lunar valve in tricuspid position is possible in ewes through a transcatheter approach. A "downsize" disk-based nitinol stent is needed to allow valve implantation in atrioventricular position. Further improvements will make this technique feasible in humans in tricuspid as well as in mitral positions.

O10-3

Stent implantation in the ductus arteriosus – when, why, how?

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Objectives: Attempts of stenting the ductus arteriosus (DA) for continued patency started in 1991; human experience was described in hypoplastic left heart syndrome for bridging to heart transplantation (HTX) and as basis for combined Norwood stage II and I repair as well as for pulmonary blood flow in cyanotic congenital heart disease. We report on our current experience in ductal stenting, indications, technical aspects are presented.
Patients: From 6/96–1/04 ductal stenting was performed in 66 pts, with duct dependent systemic (n = 34) and pulmonary (n = 32) blood flow.
Methods: Ductal stenting in left heart obstructions (group A) was performed transvenously (5F introducer sheath/7F guiding catheter), in pulmonary atresia/stenosis (group B) we mostly used a transarterial approach (4F introducer sheath). Starting with hand crimped stents (Jo-med, Nir-flex, Corinthian) we switched to premounted stents (Tsunami, Omnilink, Tecnic) with a length of 10–20 mm.The final diameter was 6–9 mm in group A and 3–4,5 mm in group B.
Results: In left heart obstructive lesions ductal patency was achieved for up to one year, redilation was necessary occasionally. In this group one patient died after PA-banding, 9 pts had biventricular repair, 13 had combined Norwood I and II repair at the age of 3–7 mo, meanwhile 9 are completed to TCPC and 5 pts underwent HTX. In group B ductal patency was demonstrated for up to 5 years, a second stent was necessary in 10; an additional AP-shunt in 5 pts. The rate of reinterventions was higher and more complicated. Two pts with ductal stenting in a rescue situation, died despite successful intervention. In follow up palliative surgery (Glenn, TCPC) was performed in 8 pts (2 pts still waiting), corrective surgery in 9 pts (3 pts still waiting). Together with transcatheter opening of the right ventricular outflow tract, 5 pts have been cured by intervention alone. We had one procedural death in group A and none in group B.
Conclusions: Ductal stenting should be considered in selected patients with duct dependent systemic or pulmonary blood flow. Bridging to transplantation, new approaches of combined Norwood surgery or delayed surgery in severely compromised newborns on the one hand and right heart obstructive lesions requiring only temporary additional blood supply on the other make them candidates for this transcatheter approach.

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O10-4

Late fenestration of a Fontan circuit, including TCPC with extra-cardiac conduits

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Objectives: A fenestration can be life-saving in patients with a failing Fontan circuit. The wall between systemic and pulmonary venous blood may consist of atrial fibrous or muscular septum, pericardium, conduit tissue (Goretex), or composite (extra-cardiac conduits). It remains a challenge to create and maintain a well balanced fenestration in such circuits.
Methods: A fenestration was created or enlarged during 14 procedures in 11 patients a mean of 31 (SD 36) months after Fontan procedure. Indications were low cardiac output 4, PLE 4, bronchial cast 1, chylothorax 2. 4 pts had a pre-existing small fenestration; puncture (from ICV 6, SCV 1) with a Bronckebrough needle was required in 7 pts. After puncture, a 0.014″ stiff coronary wire was introduced; the small puncture hole was enlarged with either a Mullins sheat 4, Rotablator 1, and/or regular 3 or Cutting Balloon 2. A variety of techniques was used to open the fenestration: balloon dilation only 5, Wallstent 2, stent 3, covered stent 1, diabolo shaped stent 3 (Stumper technique with wire loop centered between balloon-stent). We aimed for a saturation of 80–85%. In 1 pt a stent was recanalised twice under temporary bilateral carotid occlusion. When punctioning through an extra-cardiac conduit, both walls layers were reapproximated by a modified Stumper technique: the distal half of the stent was first opened in the "left atrium" keeping the proximal half unfolded within the Mullins sheat; all is then pulled against the septum; the sheat is further withdrawn into the ICV; the stent is fully opened in diabolo across the septation.
Results: A significant R > L shunt was created in all patients: saturation decreased from 96 ± 2 to 84 ± 4%. In all 5 pts with balloon dilation only, the fenestration spontaneously closed within days-weeks; this was clinically sufficient in 3 pts with very early post-operative complications. In all patients with long–straight stents the fenestration regressed significantly within days–weeks, with total occlusion in 4/5 pts within 3 months. All patients with a short diabolo shaped stent had a lasting stable fenestration.
Conclusions: A fenestration can be obtained even in extracardiac Fontan circuits. Currently our preferred method is the modified Stumper technique which allows very accurate positioning across any septum, and reapproximates the different layers if required.

O10-5

Percutaneous closure of residual post surgical ventricular septal defects

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Background: Significant residual VSD may occur after surgical closure. Redo operation have higher rate of morbidity and mortality.
Aim: To report our experience with various Amplatzer VSD Occluder devices in closure of post-surgical residual VSD.
Patients and Methods: Between January 2000 and December 2003, 72 patients (pts) underwent percutaneous VSD closure. Eleven out of 72 had a residual postsurgical VSD. Eight pts had the patch closure of an isolated VSD, 2 had VSD closure in tetralogy of Fallot and 1 in REV procedure. They were operated on a median of 8 years before the percutaneous procedure. All pts had echocardiographic signs of left ventricle volume overload. Two different Amplatzer devices were used: The muscular VSD occluder (mVSD) and the eccentric VSD occluder (pmVSD). For closure of mVSD a circuit between the right internal jugular vein and the right femoral vein was created, while in pmVSD an artero-venous circuit between the right femoral artery and the right femoral vein was created.
Results: Median age at intervention was 13 years (range 1,5–64 yrs). Median weight was 55 kgs (range 9–80 kgs). Median QP/QS was 1,8/1 (range 1,5–2,5). Mean fluoroscopy and procedure times were 41 ± 31 and 135 ± 91 min, respectively. The median size of the VSD was 8 mm (range 6–18 mm). In 9 pts a mVSD device was used, while in 3 cases a pmVSD device was implanted. In one subject two procedures and devices were needed. No death occurred. Two pts required immediate external cardioversion (in one due to ventricular fibrillation, in another due to atrial fibrillation). In one subject due to the impossiibily to place the sheath towards the apex of the left ventricle the device was opened through the aortic valve. No significant aortic, mitral or tricuspid regurgitation occurred. A mild to trivial intraprotesic residual shunt was showed in three pts. One patient had a mild residual shunt between the two devices implanted. All subjects but one were discharged home 48 hours after the procedure. One subject due to post procedural fever was discharged 96 hours after the procedure. Median follow-up was 8 months (range 1–33 mts). Complete occlusion was shown in all subjects but one who had a mild residual shunt between the two devices implanted. No late complications occurred.
Conclusions: In the current era, percutaneous closure of residual post surgical VSD can be achieved safely and successfully.

O10-6

MRI guided transcatheter implantation of a prosthetic valve in aortic valve position: feasibility study in swine

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Objectives: To test the feasibility of MRI to guide transcatheter implantation of a prosthetic valve in aortic valve position.
Background: The ability of MRI to acquire real-time images at any arbitrary orientation and with high soft tissue contrast for visualization of the aortic valve and coronary arteries makes this technique attractive for transcatheter implantation of a prosthetic valve in aortic position.
Methods and Results: The endovascular intervention was performed in swine (n = 6) using a 1.5 Tesla scanner. The implanted device comprised a tricuspid teflon heart valve, which was sutured into a selfexpanding nitinol stent. The valved stent was compressed and front loaded into a 10F delivery system. Catheter visualisation was achieved by the use of small MR susceptibility markers, which were easily perceptible to the observer. MRI provided online detailed 3D information about the position of the delivery system and its surrounding soft tissue anatomy including the mitral valve, aortic valve and coronary arteries. Successful transcatheter implantation of the prosthetic valve was achieved in all cases. Postinterventional assessment of phasic aortic blood flow using velocity encoded cine MRI showed no aortic insufficiency in five cases. Only minor central valve incompetence with a regurgitant fraction of 6 ± 3% was detected in two cases. Aortic valve stenosis, obstructed coronary arteries or wall motion abnormalities were not observed. Postmortem examination revealed accurate valved stent placement and no evidence of vascular or myocardial damage. Accuracy of the valved stent placement was in the range of 1–3 mm.

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Conclusions: Transcatheter implantation of valved stents in the aortic valve position can be succesfully performed under MRI guidance.

Session 11: General Cardiology I

O11-1

Warfarin therapy improves clinical outcome of Kawasaki disease patients with giant coronary aneurysm

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Background: Giant coronary aneurysms (GA) increase the risk of sudden death due to myocardial infarction (MI) caused by massive thrombosis. Anticoagulation therapy using warfarin for Kawasaki disease (KD) patients (pts) with GA is controversial. We investigated whether warfarin therapy improve the clinical outcome of KD pts with GA.
Methods: We followed 2031 KD pts since 1973 at Kurume University hospital. Fifty-one pts (37 male and 14 female) were retrospectively studied. Giant aneurysms were diagnosed by coronary angiogram in all pts. Patients divided into 2 groups, Group A; consisted 29 coronary branches in 17 pts treated with combination of warfarin with aspirin, Group B; consisted of 60 coronary branches in 34 pts treated with aspirin alone.
Results: Mean age at diagnosis was not significantly different (3.1 year vs. 2.6 year, p = NS). Angiograms were performed 4 ± 2 times during follow-up period. Combination therapy of warfarin and aspirin significantly reduced the incidence of MI compared with aspirin alone (aspirin alone 25% vs. warfarin 3.4%, $p < 0.05$). Stenotic lesions developed in 31 branches, including 10 branches (34%) in Group A, and including 21 branches (35%) in Group B. The incidence of stenosis was not significantly different between 2 groups. The incidence of recanalization was not significantly different between 2 groups (Group A; 4 branches vs. Group B; 8 branches). The number of requiring CABG surgery (2 branches, 6.9%) in patients treated with combination of warfarin with aspirin tend to be smaller than those (10 branches, 16.7%) of patients treated with aspirin alone. However, there was no significantly different between 2 groups. In 1 patient treated with aspirin alone, sudden death occurred by MI due to coronary thrombosis in acute phase. However, no pts treated with combination of warfarin with aspirin died suddenly in acute phase.
Conclusions: The combination of warfarin with aspirin therapy improved the clinical outcome of KD pts with GA. The warfarin therapy reduced the incidence of MI and prevents the sudden death.

O11-2

Cyclosporine C2-blood-level may identify inadequate immunosuppressive effect in pediatric heart transplant patients

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Background: Inadequate Cyclosporine A (CyA) bio-availability is a significant risk factor for acute or chronic rejection and graft failure after organ transplantation. Dosage adjustment of Cyclosporine A is routinely controlled via the trough concentration (C0-level), which is the blood concentration before the next application, in most transplant centers. Studies in adult transplant recipients point out, that measurement of the blood level two-hours after oral application (C2-level) may present higher bio-availability and immunosuppression. The aim of this empiric study was to evaluate, whether a reduced C2 level is related to inadequate immunosuppressive effect of Cyclosporine and possible graft-rejection.
Method: 20 children were referred to the German heart center because of suspected graft rejection. Rejection was monitored in all patients using echocardiography and intramyocardial EKG (IMEG) and was confirmed by myocardial biopsy. Measurement of CyA blood concentration was performed just before (C0-level) and 2 hours after the oral application of Cyclosporine (C2-level). Blood concentration was analyzed in standardized matter using EDTA-blood sampling and mass spectometry (Applied Biosystems, USA).
Results: The mean age was 9 years (range 3–15 years) and the mean post-transplant time was 3 years (range 1.4–5.6 years). All patients received clinical rejection-monitoring, where only four of twenty patients had a biopsy-proven rejection (ISHLT-classification IIIA). In patients with rejection mean C2-levels were significantly lower than in those without rejection (367 ng/ml vs. 1056 ng/ml, $p = 0.02$). There was no significant difference in the measured C0-level between both groups (219 ng/ml vs. 158 ng/ml, $p = 0.78$).
Conclusions: Reduced C2 level rather than C0 level was related to graft rejection and may reflect more adequately bio-availability and immunosuppressive effect of oral Cyclosporine A-treatment in pediatric heart transplant patients.

O11-3

Infective endocarditis in pediatric and adult patients with congenital heart disease – Japanese nationwide survey

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This study aimed to clarify the practical prevention, management and prognosis of infective endocarditis (IE) in patients with congenital heart disease (CHD) in Japan through a nationwide survey. The clinical data on 239 patients with IE (age; 14.7 ± 15 years, 14 days–69 years) hospitalized during 1997 to 2001 in 66 institutions in Japan were analyzed. The most common underlying lesion was ventricular septal defect (38%) and tetralogy of Fallot (18%). 56% were post-repaired or palliated patients. 56/74 repaired patients with IE had some residual cardiac lesions after repair.

The most frequent procedure related to IE was dental procedure (38%) and cardiovascular surgery (29%). Blood culture was performed in 3.5 times/patient in average and positive in 86%. Causative organism was streptococcal species in 50% and staphylococcal species in 38%. Vegetation was found in 90% with mean diameter of 11 mm. Complication was observed in 49%. Antibiotics for IE treatment were penicillins and/or cefems in 85% with the duration of 29.7 ± 16 days. Antibiotics in 70% were changed to alternative antibiotics during active phase, and either carbapenems or polypeptides (40%) were frequently used for this purpose. Mortality in total was 8.8% (21/239), being 8.3% in patients with medical therapy only, 11.5% in those underwent surgery in active IE and 0% in those underwent surgery in healded IE. The mortality (21 deaths) was higher in patients those were lower age especially infants (6/21 = 28.5%) and those had a methicillin resistant staphylococcus aureus (11/21 = 52.4%), aortic valve (7/21 = 33.3%) and perioperative (6/28 = 21.4%) IE. IE in patients with CHD still carried significant mortality and morbidity in spite of recent improvements of antimicrobial therapy and diagnosis. The results of this nation-wide survey should be helpful in making future guidelines for prevention and management of IE in CHD.

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O11-4

Aortic elastic properties in patients with Marfan syndrome (MFS) – new diagnostic markers?

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Aortic wall dysfunction due to a loss of elasticity is the primary cause of premature death in patients with MFS. We sought to determine aortic elastic parameters noninvasively during a follow up of 30 months and to quantify their diagnostic power.

28 patients with MFS according to the Ghent criteria aged 16,5 ± 9,7 years were investigated prior to medical therapy and 30 months later under medication with a beta blocker. 28 age and sex matched healthy persons served as control group. Diameter measurements were obtained at the aortic bulbus, the ascending and the descending abdominal aorta out of M-Mode echocardiographic images by a special autocontour finding software. After simultaneous blood pressure determination several aortic elastic parameters were calculated.

At the first examination MFS patients showed larger mean end-diastolic aortic root diameters (33 vs. 24 mm) and ascending aortic diameters (26 vs. 20 mm) than controls. Ascending aortic distensibility and maximum systolic area increase (MSAI) showed to be 50% decreased in patients. A significant reduction (30%, $p < 0,05$) of distensibility and MSAI was observed also in the descending aorta while aortic stiffness index was increased by the same amount. Follow up investigations 30 ± 8 months later under oral medication with Atenolol (0,7 ± 0,5 mg/kg KG) revealed different results between a group of patients on a higher dose (1.1 ± 0.5 mg/kg KG, $n = 13$) and a low dose group (0,3 ± 0,3 mg/kg KG, $n = 15$). Patients on the higher dose showed a significant increase of mean ascending aortic distensibility from 31 to 45 $kPa^{-1} \cdot 10^{-3}$, while aortic elastic parameters did not change significantly in the low dose group. The diagnostic power of several aortic parameters was analyzed by logistic regression models. The model including ascending aortic distensibility as well as aortic root and ascending aortic diameters related to body surface area showed the highest sensitivity and specificity (95% each).

Loss of aortic elasticity can be accurately quantified out of M-Mode echocardiographic images in patients with MFS. Ascending aortic distensibility as well as the diameters of the aortic bulbus and the ascending aorta can help to evaluate the benefit of medical therapy, may influence therapeutic decisions and – implicated in logistic regression models – can serve as sensitive diagnostic markers for the presence of MFS.

O11-5

Can claims for medical negligence inform the practice of paediatric cardiology?

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In UK law an expert witness is answerable to the Court. A medicolegal report should reach the same conclusions, whether instructions are from lawyers acting for plaintiff or defendant. Civil cases ultimately concern money paid by insurance companies, despite dented medical pride when a claim is successful.

Causation and liability have been assessed in 80 consecutive cases. In 48 (60%) instructions were from plaintiff, in 29 (36%) from defendants and 3 were joint. Liability was admitted in 6 cases: Failure to give globulin in early Kawasaki disease, failure to perform pulmonary artery banding, dopamine via umbilical artery rather than vein, untreated high K, over heating at cardiac catheterisation and failure to record and review echocardiographic data. Four additional cases reached the court, 3 being successfully defended, alleged failure to arrange follow-up was upheld. Two further cases, too large a balloon for pulmonary valvotomy, and cardiac perforation at diagnostic catheterisation with angiography in the pericardial space are likely to be settled. Most claims rightly fail, but analysis indicates situations where vigilance may prevent problems in the future. 10 claims involved failure to diagnose congenital heart disease on an antenatal scan, 5 of which were diagnosable on a four-chamber view. 8 claims involved Down syndrome and 4 sudden deaths, preceded by at least one episode of syncope on exertion. Causes were primary pulmonary hypertension, catecholaminergic polymorphic ventricular tachycardia, Wolff–Parkinson–White syndrome and hypertrophic cardiomyopathy.
Conclusions: Litigation is increasing. Awareness and vigilance can safeguard patient and Cardiologist. Expert opinion should be obtained on all fetuses where a four-chamber view is not obtained. All children with Down syndrome should be screened by echocardiography on diagnosis. Syncope on exertion should always be investigated further. β-blocker to be given on induction for repair Tetralogy of Fallot. Echocardiographic as well as pacemaker checks to be performed on follow-up of congenital heart block. Consider anticoagulation for infants with SVT. Send copies of correspondence to parents as well as to colleagues.

O11-6

Does balloon septostomy performed outside paediatric cardiac centres affect outcome?

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Objectives: To compare the outcome of patients with transposition of great artery (TGA) undergoing balloon septostomy at peripheral neonatal units with those undergoing septostomy at tertiary cardiac centre.
Methods: Over a 76 month period (January 1997–April 2003), 147 patients with TGA were treated at Birmingham Children's Hospital (BCH). Out of these, 115 underwent balloon septostomy (BAS) [peripheral neonatal unit (PNNU) 14, BCH 64, other tertiarycardiac centres (OC) 34]. Interventional expertise was provided solely by trained Paediatric Cardiologists. All case records were analysed retrospectively. Eight patients older than 30 days at presentation to BCH were excluded. All patients from OC were excluded from analysis due to later referral and multiple co-morbid factors. Outcome measures were: age at admission, morbidity, mortality and length of hospital stay. Morbidity included significant acidosis, haemodynamic instability, difficult venous access, necrotising enterocolitis, sepsis and loss of peripheral pulses. All BAS were performed under ultrasound guidance at PNNU; while at BCH, 49% were by ultrasound and 51% by additional fluoroscopy.
Results: Comparing PNNU and BCH groups, there was no significant difference in the incidence of pre-BAS morbidity (PNNU 21%, BCH 14%, $p = 0.26$), post-BAS morbidity (PNNU 28%, BCH 33%, $p = 0.5$), age at BAS (median age: PNNU 1 day, BCH at birth) and duration of post-operative hospital stay at BCH (median duration: PNNU & BCH 8 days). The PNNU group was admitted significantly later to BCH (median age: PNNU

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3 days, BCH 1 day, p = 0.007) and had significantly shorter hospital stay at BCH (median duration: PNNU 11 days, BCH 14 days, p = 0.04). Eleven of 14 patients from PNNU could be extubated prior to transfer to BCH. There was no death in the PNNU group. One patient in the BCH group died. The patient had delayed transfer, was moribund on admission and failed to improve post BAS.
Conclusions: Balloon septostomy can be performed safely and effectively at peripheral neonatal unit. There is no difference in morbidity or mortality. Septostomy at peripheral neonatal unit allowed for elective transfer, extubation and reduced total duration of hospital stay at the tertiary cardiac centre.

Session 12: Functional Imaging

O12-1
Assessment of regional myocardial function in the long-term follow-up of children with history of Kawasaki Disease – results of a study using Tissue Doppler and Angiography

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Objectives: Kawasaki Disease (KD) leads to coronary aneurysm (CA) formation in 5–20% of all patients. Subsequently myocardial dysfunction due to persistent aneurysms may cause severe problems. Reports concerning regional myocardial function in the long-term follow-up of KD are rare. The study intended to assess regional myocardial function using Tissue-Doppler-Echocardiography (TDE) in comparison to conventional angiography.
Methods: Eighteen asymptomatic children aged 13 ± 4 years (range 6–23 y) with persistent coronary aneurysms following KD were included in this study. Eighteen age- and gender-matched healthy subjects served as a control group. Morphology of coronary arteries and left ventricular function was evaluated invasively by angiography during acute stage and at follow up. All children underwent echocardiography including TDE and Strain Rate Imaging (SRI) at rest and during a submaximal bicycle exercise test (HR 128 ± 8 bpm). Median follow-up period between acute KD and TDI was 9,8 years (4,4–21,5 y). TDE/SRI examination of the left ventricle (LV) was performed from long and short axis, apical 2- and 4-chamber views. Peak systolic (Vmax-S) and early diastolic (Vmax-E) velocities, peak systolic strain and peak systolic and diastolic strain rate, were calculated for all segments of the LV.
Results: Good quality recordings of TDE were received in 52% of all investigated LV segments. Segments supplied by normal and affected coronary arteries showed no differences concerning visible wall motion, systolic velocities, strain or strain rate both at rest and during exercise. At rest, Vmax-E was normal in all segments. During exercise, Vmax-E increased in unaffected segments and segments supplied by a coronary artery having an aneurysm (7,4 ± 3,9 cm/s to 8,7 ± 5,6 cm/s). However in segments supplied by arteries with an actually relevant stenotic aneurysm, Vmax-E decreased significantly during exercise (7,2 ± 3,9 cm/s to 5,0 ± 4,8 cm/s – p < 0,05).
Conclusions: In the long term follow up of KD systolic function is unimpaired even in segments with relevant coronary stenosis. In these segments an exercise-induced regional diastolic dysfunction can be revealed by TDE. This is potentially helpful in identifying developing coronary stenosis at long-term follow-up, even in asymptomatic children.

O12-2
Role of three dimensional transthoracic echocardiography in the management of patients with congenital heart disease

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Background: Currently, two-dimensional echocardiography is the cornerstone for non-invasive imaging in congenital heart disease, but in complex defects, there are limitations in the spatial information derived. Accurate anatomic definition is mandatory for successful surgical or non-surgical intervention. We reviewed the additional information provided by 3D echo to evaluate its role in the management of congenital heart disease.
Methods: We acquired a Philips Sonos 5500TM with 3D image acquisition software in 2003 and all patients who had both 2D and 3D echos from June to December were retrospectively reviewed by the cardiology and surgery consultant team. This information was compared with the intraoperative findings in all patients who had subsequent surgical intervention.
Results: Twenty-five 3D echos were undertaken during this period. Diagnoses included: TGA (5), AVSD (4), aortic stenosis (4), anomalous pulmonary venous drainage (2), abnormal mitral valve (2), complex intracardiac defects (2), VSD (2), ASD (2), Ebsteins anomaly (1) and pericardial effusion (1). Additional information over 2D echo was obtained all but one patient with supracardiac TAPVD. This information altered the proposed surgical intervention in 5 patients (20%), including the resection of a membrane in the left ventricular cavity, repair rather than palliation in a complex intracardiac defect, aortic valve repair rather than replacement, surgical repair of Ebstiens anomaly, and repair of complex AVSD. In 16 of the 25 patients 3D echocardiograms resulted in greater spatial undestanding of defects seen on 2D. This included definition of coronary arteries in TGA, AV valve morphology in AVSD and the detailed morphology of ASDs and VSDs. In aortic stenosis it gave clear definition of the aortic valve anatomy and extent of commisural fusion with valuable information on associated subvalvar pathology. It was most helpful in understanding the AV valve morphology and subvalvar apparatus. It identified the nature and extent of valve perforation in endocarditis and following subaortic membrane resection.
Conclusions: 3D echo evaluation of congenital heart defects is extremely helpful in defining the anatomy of complex congenital heart defects and in identifying valvar and subvalvar pathology. With improvements in software it will supersede 2D echocardiography in the forseeable future.

O12-3
Carotid-subclavian arteries Index (CSI), a new echocardiographic index to recognize coarctation of the aorta in neonates and infants

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Introduction: Coarctation of the aorta is a common congenital heart malformation. Mode of diagnosis changed from clinically to almost exclusively by echocardiogram and MRI. We claim to find a new echocardiographic index, based on simple and reliable morphologic

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measurements, to facilitate the diagnosis of aortic coarctation in the newborn. We reproduce the same procedure for older child to validate this new index.
Material and Methods: We reviewed echocardiographic studies of 47 neonates with diagnosis of coarctation who underwent cardiac surgery between January 1997 and February 2003 and compared them with a matched control group. We measured 12 different sites of the aorta, aortic arch and the great vessels on the echocardiographic bands. In a second time we reviewed 23 infants for the same measurements and compare them with a matched control group.
Results: 47 neonates with coarctation were analysed, age 11.8 ± 10 days, weight 3.0 ± 0.6 kg, body surface 0.20 ± 0.02 m^2. The control group was of 16 newborns aged 15.8 ± 10 days, weight 3.2 ± 0.9 kg and body surface 0.20 ± 0.04 m^2. A significant difference was noted in many morphologic measurement between the both groups, the most significant being the distance between the left carotid artery and the left subclavian artery (coarctation vs control: 7.3 ± 3 mm vs 2.4 ± 0.8 mm, $p < 0.0001$). We then defined a new index, the carotid-subclavian arteries index (CSI) as the diameter of the distal tranverse aortic arch divided to the distance left carotid artery to left subclavian artery being also significaly different (coarctation vs control: 0.76 ± 0.86 vs 2.95 ± 1.24, $p < 0.0001$). With the cut-off value of this index of 1.5 the sensitivity for aortic coarctation was 98% and the specificity of 92%. In an older group of infant with coarctation (16 patients) we apply the same principle and find for a cut-off value of 1.5 a sensitivity of 95% and a specificity of 100%.
Conclusions: The CSI allows to evaluate newborns and infants for aortic coarctation with simple morphologic measurement that are not depending of the left ventricular function, presence of a patent ductus arteriosus or not. Further aggressive evaluation of these patient with a CSI < 1.5 is indicated.

O12-4
Cardiac magnetic resonance imaging (CMR) combined with dobutamine stress in children and young adults after correction of tetralogy of Fallot (TOF)
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Introduction: Longstanding pulmonary regurgitation (PR) after correction of TOF leads to biventricular dysfunction. To study biventricular response to exercise, CMR combined with physical stress has been used. Pharmacological stress with an inotropic drug may be a good alternative. We used a low dose dobutamine protocol.
Methods: 30 patients operated for TOF (mean follow up 15,9 ± 4,6 y, mean age at repair 1,0 ± 0,6 y) underwent CMR at rest, and during dobutamine stress (max. 7,5γ). Monitoring was performed. Ventricular volumetric data was derived in short axis direction and EDVi, ESVi, SVi and EF were measured. Phase Velocity Cine MR was used to measure flow across the pulmonary valve orifice. After CMR a maximal incremental bicycle exercise test was performed.
Results: CMR results: RV measurements at rest and with stress: RVEDVi (ml/m) 137 ± 40 vs 133 ± 41 ($p < 0.01$); RVESVi 72 ± 29 vs 55 ± 25 ($p < 0,01$); RVSVi 65 ± 15 vs 77 ± 20 ($p < 0,01$); RVEF (%) 48 ± 8 vs 60 ± 7 ($p < 0,01$). LV measurements: LVEDVi 82 ± 12 vs 81 ± 15 (n.s.); LVESVi 35 ± 9 vs 25 ± 10 ($p < 0,01$); LVSVi 46 ± 7 vs 56 ± 9 ($p < 0,01$); LVEF 57 ± 7 vs 70 ± 8 ($p < 0,01$). PR measurements at rest and with stress: PRi (ml) 24 ± 13 vs 31 ± 20 ($p < 0,01$); PR (%) 34 ± 15 vs 34 ± 17 (n.s.). Bicycle-ergometry results: The test was stopped at submaximal levels in two patients. The percentage of predicted maximal workload was 88 ± 13%; max. HR 180 ± 17/min; max VO_2 37,9 ± 7,7 ml/kg/min; respiratory quotient at max exercise 1,21 ± 0,07. The achieved percentage of predicted maximal workload in patients with PR <30% differed from that in patients with PR ⩾ 30% (97 ± 8 vs 83 ± 13%, $p < 0,05$). Correlations with reached percentage of predicted workload: (Positive) LVEF at rest ($r = 0,43$ $p < 0,05$), LVSVi at rest ($r = 0,47$, $p < 0,05$) and RVEF at rest ($r = 0,41$, $p < 0,05$). (Negative) PR percentage during stress ($r = -0,39$, $p = 0,05$) and PR percentage at rest ($r = -0,46$, $p < 0,05$).
Conclusions: Our LV measurements are in agreement with earlier reports. However the normal systolic RV response and unchanged PR (%) are remarkable. Patients with PR over 30% perform significantly worse in maximal exercise testing. All patients with exexercise capacity below 80% of predicted had a PR (%) of over 30%.

O12-5
Myocardial ischemia and scars are not common in the systemic right ventricle at long-term follow-up
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Background: The systemic right ventricle (RV) in patients after atrial switch operation or with congenitally corrected transposition of the great arteries (ccTGA) often fails. Because some studies have found a high incidence of myocardial ischemia and scars, it has been proposed that myocardial ischemia and scars are a reason for the failure of the systemic RV. However, this is not in accordance with our clinical observations. The aim of this study was therefore to test the hypothesis that myocardial ischemia and scars are common in patients with systemic RV using newer imaging modalities (positron emission tomography, PET and late enhancement magnetic resonance imaging, LE-MRI) in consecutive patients with systemic RV.
Methods and Results: 40 patients with systemic RV were studied by PET and LE-MRI (25 patients 21.8 ± 4.4 yrs after atrial switch operation, age at operation: 1.3 ± 1.5 yrs and 15 patients with ccTGA, age: 31.1 ± 16.6 yrs, no previous operation). Two investigators analyzed PET and LE-MRI studies and were blinded to the findings of the other method . None of the patients after atrial switch operations had myocardial ischemia or scars. Only one patient with ccTGA had an anterolateral ischemia of the systemic RV and only one patient with ccTGA had a subendocardial scar of the systemic RV.

This study shows that the hypothesis that myocardial ischemia and scars are common in patients with systemic RV is not correct. Therefore myocardial ischemia and scars are not the reason of failure of the systemic RV. Note: This abstract has been partially presented as an oral presentation at the 2003 meeting of the German Association of Pediatric Cardiology.

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O12-6

Atrial baffles restrict rise in stroke volume in patients after atrial switch repair for transposition of the great arteries (TGA), when compared to their natural analogy of unoperated congenitally corrected transposition (ccTGA) – a study by magnetic resonance imaging (MRI)

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Objectives: Exercise tolerance in patients after atrial switch repair for TGA is known to be reduced. Debate is still ongoing whether right ventricular (RV) myocardial failure in face of systemic ventricular demand or diastolic inflow impairment by atrial baffles is the underlying mechanism. The present study should answer this question by comparing patients after atrial switch OP with unoperated patients with ccTGA, its natural analogy with the right ventricle in systemic position.
Material and Methods: 12 patients after atrial switch OP without baffle obstruction (median age 20 yrs. (15–28)) and 11 patients with unoperated ccTGA (median age 37 (6–59)) were studied under resting condition and during infusion of dobutamine 10 μg/kg/min. Using MRI (Philips Gyroscan NT, 1.5 T) short axis cuts of the ventricles for volume measurements were performed under both conditions. A multiphased steady state free precession technique was applied. In addition, aortic stroke volume was assessed by MR phase velocitiy mapping. The following parameters, indexed to body surface area, were assessed: RV volumes during enddiastole and endsystole, RV stroke volume (RV-SVI), RV ejection fraction (RV-EF), cardiac index, heart rate, systolic and diastolic blood pressure and RV mass.
Results: SVI under dobutamine rose significantly in the ccTGA group ($p < 0.01$) but not in patients after atrial switch OP ($44{,}16 \pm 8{,}38$ to $43{,}41 \pm 6{,}83$, $p < 0{,}01$) Under rest RV-EF was higher in the ccTGA group ($62{,}4 \pm 13{,}6\%$ vs. $48{,}2 \pm 22{,}6\%$, $p < 0{,}05$) than after atrial switch OP. This difference was abolished under dobutamine. All other parameters in the two groups did not differ statistically significant.
Conclusions: Under dobutamine stress RV-SVI can be increased in patients with ccTGA, but not in patients after atrial switch OP. Given a similar pressure load to both right ventricles, this difference can only be explained by diastolic inflow impairment at atrial level in patients after atrial switch OP.

Session 17: Pulmonary Vascular Biology

O17-1

VEGF expression is modulated by the redox-modifying enzymes catalase, glutathione peroxidase and superoxide dismutase in pulmonary artery smooth muscle cells: role of reactive oxygen species

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Pulmonary hypertension is frequently associated with vascular remodeling linked to progressive intimal and medial thickening and proliferation of vascular cells. Often, hypoxemia and a prothrombotic state are accompanying this disorder. Both stimuli can induce the vascular endothelial growth factor (VEGF), which is upregulated in pulmonary hypertension and may contribute to remodeling. Recently, reactive oxygen species (ROS) have been implicated as signaling molecules triggering cellular proliferation in response to a variety of stimuli. In this study we investigated whether modulation of ROS levels by redox-modifying enzymes affects the activation of the hypoxia-inducible transcription factor HIF-1alpha and the expression of its target gene VEGF in response to thrombin or the hypoxia mimetic $CoCl_2$ in pulmonary artery smooth muscle cells (PASMC).

Stimulation of PASMC with thrombin or $CoCl_2$ increased ROS production and enhanced HIF-1alpha protein and VEGF mRNA levels as well as HIF-dependent reporter gene activity. Treatment with antioxidants or overexpression of glutathione peroxidase or catalase decreased ROS production, HIF-1alpha protein and VEGF mRNA levels and HIF-dependent reporter gene activity, whereas in PASMC overexpressing superoxide dismutase enhanced ROS production, HIF-1alpha protein and VEGF mRNA levels and HIF-dependent reporter gene activity was detected. Similarly, treatment with H_2O_2 (50 μM) enhanced ROS production, HIF-1alpha protein and VEGF mRNA levels as well as HIF-dependent reporter gene activity and stimulated proliferation of PASMC.

These findings suggest that a "prooxidant" state associated with elevated ROS levels promotes activation of the HIF pathway and VEGF expression leading to enhanced proliferation of PASMC. In contrast, an "antioxidant" state with reduced levels of ROS prevents the activation of the HIF pathway. Thus, shifting the redox balance to a more reduced environment thereby preventing activation of the HIF pathway and limiting VEGF expression may be beneficial for treating remodeling processes during pulmonary hypertension.

O17-2

Human hypoxia-inducible factor 3-α (HIF3α) isoforms modulate the hypoxia response pathway in endothelial cells

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Chronic hypoxia can result in pulmonary hypertension and vascular remodeling due to increased proliferation and hypertrophy of vascular cells. The hypoxia-inducible transcription factor family HIF is activated by low oxygen levels and controls hypoxic gene expression. These heterodimers consist of an inducible α-subunit and a constitutive β-subunit. Whereas human HIF1α and HIF2α as well as rodent HIF3α have been shown to increase the expression of genes involved in pulmonary vascular remodeling including vascular endothelial growth factor (VEGF) and plasminogen activator inhibitor-1 (PAI-1), the role of human HIF3α in the vascular hypoxic response is not clear yet. A truncated isoform to HIF3α (inhibitory PAS domain protein, IPAS) has been found to inhibit the HIF pathway in the mouse. A human homologue has not been described yet. We aimed to identify human IPAS homologues and to determine their role in the hypoxic response in endothelial cells (EC).

Analysis of human HIF3α mRNA transcripts showed a number of isoforms, one of which, HIF3 transcript variant 3 (HIF3tv3) produces an amino acid sequence similar to IPAS. RT-PCR analysis revealed the presence of HIF3α and HIF3tv3 in EC and pulmonary artery smooth muscle cells. Expectedly, overexpression of HIF1α resulted in increased HIF-dependent reporter gene activity as well as elevated VEGF and PAI-1 promoter reporter gene activity under hypoxia in EC. Overexpression of HIF3α or

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HIF3tv3 reduced the hypoxic responses. Moreover, compared to controls, the proliferative activity was significantly decreased in HIF3α- and HIF3tv3-expressing cells. In contrast to HIF1α- and HIF2α-expressing EC, showing a marked increase in the angiogenic capacity in an in vitro angiogenesis assay, the angiogenic response was markedly decreased in HIF3α- and HIF3tv3-expressing EC. These data show, that in human EC not only the IPAS homologue HIF3tv3, but also HIF3α itself depress hypoxic gene expression, proliferation and angiogenesis suggesting that the human HIF-3α system may play an important role in regulating the HIF pathway and limiting the hypoxic response. Thus, enhanced expression of HIF3α may have a beneficial effect in controling pulmonary remodeling processes in response to hypoxia.

O17-3

Rac regulates thrombin-induced tissue factor expression in pulmonary artery smooth muscle cells involving the nuclear factor kappa B pathway

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Pulmonary hypertension is characterized by hypertrophy and enhanced proliferation of vascular cells, frequently associated with a prothrombotic state, which may play an important role in promoting vascular remodeling and thus the manifestation of dysfunction of the pulmonary vasculature. The primary link between vascular cells and the hemostatic system is provided by the cell surface protein tissue factor (TF) leading to the formation of thrombin. Intriguingly, thrombin itself has been shown to modulate the expression of TF in smooth muscle cells. However, the signaling pathways underlying such a thrombogenic cycle are not completely understood yet. The GTPase Rac has been shown to be essential for activation of the NADPH oxidase and subsequent production of reactive oxygen species (ROS), but can also act directly by activating downstream kinase cascades leading to modulation of transcription factor activities and gene expression. The transcription factor NFkB is a major regulator of redox-sensitive gene expression. We therefore investigated whether Rac and NFkB are involved in the regulation of TF expression in response to thrombin in pulmonary artery smooth muscle cells (PASMC).

Thrombin-induced TF mRNA and protein expression as well as TF-dependent surface procoagulant activity were abrogated in PASMC expressing dominant-negative RacT17N but were enhanced by active RacG12V. Thrombin or RacG12V also increased reporter gene activity mediated by the active human TF promoter (pTF636) and, although to a lesser extent, by the minimal TF promoter (pTF111), known to lack a NFkB DNA binding site. Indeed, NFkB-dependent reporter gene activity was enhanced by thrombin and RacG12V. Moreover, overexpression of the NFkB subunits p50/p65 revealed significantly elevated reporter gene activity mediated by pTF636, but not by pTF111. Finally, in RacG12V-overexpressing cells, pTF636-, but not pTF111-dependent reporter gene activity, was significantly abrogated by coexpression of dominant-negative mutants of the inhibitory IkB protein or the IkB kinase, which prevent NFkB activation. Since enhanced NFkB activity has been observed in patients with pulmonary hypertension, Rac-dependent activation of the NFkB pathway may be a critical element promoting thrombin-induced TF activity and thus a prothrombotic state in pulmonary hypertension.

O17-4

Urotensin II is a novel activator of the NADPH oxidase in pulmonary artery smooth muscle cells: role in the regulation of plasminogen activator inhibitor-1 expression

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Human urotensin II (hU-II) has recently been described as a potent vasoactive peptide which may be involved in a variety of cardiovascular diseases including pulmonary hypertension. However, the signaling mechanisms activated by this peptide are not well understood. Since reactive oxygen species (ROS) play an important role as signaling molecules in the vasculature and have been recently implicated in the pathogenesis of pulmonary hypertension and vascular remodeling, we investigated whether hU-II can stimulate ROS production, ROS-dependent signaling pathways and expression of the fibrinolysis inhibitor plasminogen activator inhibitor-1 (PAI-1) known to be upregulated in pulmonary hypertension, in pulmonary artery smooth muscle cells (PASMC) via activation of the ROS-generating enzyme complex NADPH oxidase.

ROS generation was significantly increased by hU-II in a time- and dose-dependent manner. This response was accompanied by elevated protein levels of the NADPH oxidase subunits p22phox, p47phox, NOX1 and NOX4. ROS production stimulated by 100 nM hU-II was abrogated by the antioxidant vitamin C, the flavininhibitor diphenyleneiodonium (DPI) as well as by transfection of a p22phox antisense vector. In contrast, overexpression of p22phox increased ROS production by hU-II. Stimulation with hU-II resulted in the activation of ERK1/2, p38 MAP kinase (p38MAPK) and protein kinase B (PKB) which was redox-sensitive and dependent on p22phox. hU-II also upregulated the expression of plasminogen activator inhibitor-1 (PAI-1) and enhanced the proliferation of PASMC. This response was dependent on the NADPH oxidase as well as on the p38MAPK, ERK1/2 and the PI3 kinase/PKB pathways.

These results show that hU-II is a potent activator of ROS generation by the NADPH oxidase in PASMC, leading via activation of p38MAPK, ERK1/2 and PKB to enhanced expression of PAI-1 and increased proliferative activity. Since elevated levels of PAI-1 have been associated with pulmonary hypertension, hU-II-mediated activation of the NADPH oxidase may thus provide a novel mechanism contributing to this disease.

O17-5

The serum- and glucocorticoid-regulated kinase Sgk1 promotes redox-sensitive upregulation of tissue factor by thrombin in pulmonary artery smooth muscle cells

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Pulmonary hypertension is a disorder frequently associated with a prothrombotic state which has been implicated in promoting vascular remodeling. Thrombin itself has been shown to increase the expression of tissue factor (TF), the regulator of the extrinsic coagulation cascade. However, the signaling pathways supporting such a thrombogenic cycle in the pulmonary vasculature are not well understood. The serum- and glucocorticoid-inducible kinase Sgk1 is a serine/threonine kinase which has been described as a stress-sensitive kinase regulated by growth factors and hormones at the

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transcriptional and posttranslational level. In this study we investigated the role of Sgk1 in thrombin-induced TF expression in pulmonary artery smooth muscle cells (PASMC).

Stimulation with thrombin (3 U/ml) resulted in a time- and dose-dependent increase in Sgk1 mRNA and protein levels which was inhibited by pretreatment with antioxidants or the NADPH oxidase inhibitor diphenylene iodonium (DPI). Redox-sensitive regulation of Sgk1 was further shown by enhanced levels of Sgk1 mRNA and protein in response to H_2O_2 (50 μM). Thrombin also increased TF mRNA and protein expression as well as TF-dependent surface activity in a redox-sensitive, NADPH oxidase-dependent manner. Overexpression of Sgk1 enhanced TF mRNA and protein expression and TF-dependent surface procoagulant activity under control conditions and in the presence of thrombin whereas expression of a dominant-negative Sgk1 mutant abolished these responses.

These findings demonstrate that thrombin regulates Sgk1 levels in a redox-sensitive manner in PASMC, and that this kinase is essentially involved in the control of TF expression and TF-dependent surface procoagulant activity. Thus, Sgk1 may play a prominent role in promoting a prothrombotic state and vascular remodeling processes in pulmonary hypertension.

Session 19: Interventional Cardiology II

O19-1
Transcatheter closure of post-infarct ventricular septal defects using the Amplatzer VSO – new hope for an otherwise dire outlook

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Introduction: Post-infarct ventricular septal defect (PIVSD) carries an abysmal prognosis which is not greatly improved by surgery. Many surgeons also prefer to operate a few weeks after septal rupture by which time many would have perished. New technology allows for transcatheter closure of acquired ventricular septal defects; we report our initial experience involving 10 centres led by a congenital heart interventionist as most adult cardiologists lack exposure to this technique.
Methods: Between September 2001 and November 2003, 19 patients were considered for catheter closure of a PIVSD using a proactive medical approach and implantation of an Amplatzer VSO device. Five (3 female) were not carried out because of old age (1), moribund state (1), bowel perforation (1), hospital decision (1) and inability to cross the VSD (1). Fifteen patients (8 male and 7 female) received 18 devices, 10 during the acute phase and 5 late after infarction. The age range was 57 to 78 years (Mean 65.4). The earliest implant was 3 days after the PIVSD and the longest 2.4 years. The 10 acute cases had 11 devices, all were on a balloon pump and 3 had coronary stenting. The intra-aortic balloon was weaned in all within 7 days. One patient had bypass graft (BG) and PIVSD closure prior to device closure. The 5 in the chronic group were in controlled failure and all had previous BG; 3 also had previous surgical VSD repair and 1 had an aneurysmectomy. Two had 2 devices inserted during different sessions.
Results: All patients survived the procedure but there were 4 late deaths due to septicaemia (1), leg gangrene from balloon pump (1), failure to close VSD (1) and tricuspid valve damage from the procedure (1). One developed a pericardial effusion and another a small aortic dissection. Three have small residual shunts, 2 in the acute and 1 in the chronic group. One of the transcatheter patients required subsequent surgery for tricuspid valve repair.
Conclusions: Transcatheter closure of PIVSD is feasible, the initial results are encouraging and provide hope for a condition with an otherwise dire outlook. A proactive approach with intensive care, balloon pump, early revascularisation and defect closure appear to be the key factors for success. The Amplatzer VSO device appears to result in closure in the majority but some PIVSDs may progress. Through experience in closure of congenital VSDs, paediatric cardiologists are in a strategic position to take this development forward.

O19-2
Aortic valve function after balloon dilatation in 184 pediatric patients after 17 years follow-up

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Background: Balloon dilatation of pediatric aortic valve stenosis is an established procedure and is regarded as the valid alternative for surgical management. Immediate and short-term results have been studied. However, long-term results remain unknown. Therefore the aim of this study was to study the long-term results of balloon dilatation of pediatric aortic valve stenosis and ist efficacy to prevent or postpone surgery.
Methods: Up to 17 years follow-up data of 184 patients who received a balloon dilatation of aortic valve stenosis in our institution between January 1986 and October 2003 were reviewed. The patients were divided into patients aged <1 month (group <1 mo, n = 63) and patients aged ≥1 month (group ≥1 mo, n = 121) at the time of balloon dilatation of aortic valve stenosis.
Results: Group <1 mo was 0.2 (0.0–1.0) months old and group ≥1 mo was 4.5 (0.1–22.0) years old at the time of the first dilatation. Six (10%) patients of group <1 mo, aged 3.1 (1.6–30.5) mo and 14 (12%) patients of group ≥1 mo, aged 8.8 (0.5–17.1) yrs, underwent a second dilatation. Eleven (17%) patients of group <1 mo, aged 1.2 (0.1–12.9) mo and 26 (21%) patients of group ≥1 mo, aged 13.5 (0.8–20.0) yrs, had aortic valve surgery later. The time between the first dilatation and the operation was 1.2 (0.1–12.9) yrs in group <1 mo and 4.8 (0.1–12.2) yrs in group ≥1 mo. Kaplan-Meier analysis showed that after 8 yrs 53% of group <1 mo and 68% of of group ≥1 mo was free of adverse events (second dilatation, opearation, death) and 60% of group <1 mo and 79% of of group ≥1 mo was free of operation. The change of aortic gradient and development of aortic regurgitation was also studied (data cannot be shown in abstract).
Conclusions: Balloon dilatation of pediatric aortic valve stenosis postpones aortic valve operation for more than 8 yrs in 60% of group <1 mo and 79% of of group ≥1 mo. Aortic regurgitation is not a major issue in group ≥1 mo.

O19-3
Total percutaneous Fallot tetralogy correction: early clinical experience and new experimental procedures

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We hypothesized that we could achieve total percutaneous Fallot tetralogy correction, if the right ventricular outflow (RVO) obstruction was corrected, since there are methods for malalignment ventricular septal defect (VSD) occlusion. We used an existing method for RVO manipulation (balloon valvuloplasty) in 2 patients and experimental methods (alcohol ablation, RVO patch placement) in 10 piglets.

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Clinical: Two cyanotic patients (4 and 7 years old, respectfully), with the Fallot tetralogy variant of dominant pulmonic valve stenosis, underwent total percutaneous correction using the transcatheter patch (TP) for the VSD occlusion and pulmonic balloon valvuloplasty, for the RVO obstruction. Balloon valvuloplasty was first performed, followed by balloon test VSD occlusion and TP VSD occlusion. The patch was released in 48 hours.
Experimental: Alcohol RVO septal injection was performed in 5 piglets using a transthoracic Doppler needle. The needle position was guided by a marker catheter placed in the RVO. An RVO patch was placed in 5 piglets; a large transthoracic sheath was used to create an anterior wall defect which was subsequently corrected by a TP. The TP was released 12–24 hours after implantation. The experimental procedures were guided by fluoroscopy and echocardiography and autopsies were performed in all animals. Both patients became acyanotic with mild residual infundibular gradients and are doing well on follow-up. The alcohol injection was accurate and caused a selective septal infarction. Percutaneous anterior defect creation was feasible as well their correction by a TP. One piglet developed hemopericardium. In autopsy the pericardium was thickened, but the TP was in place occluding the defects. In conclusion percutaneous Fallot tetralogy correction is feasible but is limited to the anatomic variants amenable to current RVO obstruction, relief methods (balloon valvuloplasty); it could be expanded though, with the development of new procedures. Additional experimental work is required.

O19-4
Stenting for congenital heart disease; a multi-center inventory of complications
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Introduction: Stenting for congenital heart disease is performed since 1992 in many centers. The number of pts/procedures is largely unknown. Stenting may be associated with serious complications. We performed a retrospective multi-center study on stent complications.
Methods: Retrospectively data were collected of all stent implantations between 1992 and 2003 from 7 centers in the Netherlands and Belgium. These included patient data, stent implantation sites and complications.
Results: Centers started with stenting between 1992 and 1997, in one center in 2001. 447 sites were stented in 409 pts, There were 15 different implantation sites. Age ranged form neonate to adult. All centers <40 implantations in the study period, except one center that performd 235. Top stent sites were left pulmonary artery (117), right pulmonary artery (81), MAPCA's (39), recoarctation (37), ductus (27), Mustard/Senning baffle (25) and native coarctation (21).

Complications occurred in 50/447: 5 procedure related deaths (1 duct stenting, 1 Fallot, 1 vessel rupture, 1 cerebral embolism, 1 myocardial infarction), 26 stent migrations (16/24 surgical or cath. retrieval), 10 malposition (incl. kinking or occlusion), 5 vessel dissection/rupture, 6 other major complications (arrhythmia, transfusion need, air embolism, thromboembolism etc). Relatively most complications occured in stenting right ventricular outflow tract (40%), pulmonary vein (20%), right-sided homograft (20%), MAPCA's (14%), shunts (11%) and aortic stents (10%).
Conclusions: Including one center which performed 50% of all stents, each induvidual center performed <7 stents/yr. Although mortality is low, complication rate is substantial (8–20%). 50% of complications are stent migrations or embolizations. Centers should join forces to reduce the complication rate.

O19-5
Transcatheter closure of ventricular septal defects using the Pfm VSD Nitinol Coil
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Over the past 10 years a variety of occluding devices has been used for transcatheter closure of ventricular septal defects (VSDs), but none has gained wide acceptance. Device embolization, AV-block, complete left bundle branch block, regurgitation of aortic and tricuspid valve are potential complications. We report our experience of transcatheter closure of muscular and perimembranous VSDs in 22 and 8 patients, respectively, with the the Pfm VSD Nitinol Coils. The minimal VSD diameter ranged from 3–7 mm. The implantation was abandoned in one patient with perimembranous VSD due to unproper device configuration. There were no procedural problems or complications at all in the remaining 29 patients. Within 24 hours after deployment of a single coil there was no residual shunt in 19 of 29 patients (65%). The total occlusion rate (single coil) after 6 months was 79%. Subsequent implantation of additional coils (1 coil in 4 patients and 2 coils in 1 patient) was achieved in 5 of 6 patients with residual shunt to obtain complete occlusion. On follow-up evaluation, there were no evidence of arrhythmias, device embolization, valve incompetence, endocarditis or hemolysis.

The Pfm VSD Nitinol Coil appears to be a promising device for transcatheter closure of VSDs in children and adults with a minimal diameter upto 7 mm. Further studies are required to document its efficacy, safety, and long term results in a larger patient population.

O19-6
A new design for a low profile and flexible device for transcatheter closure of atrial septal defects
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Objectives: For patients with an atrial septal defect, closing the defect with transcatheter technique has evolved to be the first line choice of therapy. So far, the devices available on the market have been of two major types: self-centering or non-self-centering. We have sought to develope a new device with less material and a very low profile. We also wanted it to have self-centering characteristics but with less radial force than the currently available self-centering devices, which stretch the myocardium at the edge of the defect. Our device, called SolySafe™, is constructed of 8 metal wires (Phynox), one plastic wire-holder at each end and a polyester membrane in the middle. This device is introduced through a 10 F sheath.
Methods: Our device was tested in a swine model against the Amplatzer ASO. An atrial septal defect was created with standard transseptal technique using static balloon dilation. The animals

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(10 in each treatment group) were randomized to either treatment. They were evaluated hemodynamically and with intracardiac echocardiography and fluoroscopy during implantation and with 24 h Holter recordings immediately after implantation. The animals were then re-evaluated at the end of the study (1, 2, and 6 months) with hemodynamic measurements, intracardiac echocardiography, ocular inspection and microscopy of the atrial septum and sections of the brain and the kidney.

Results: In all animals the respective device was implanted in good position. In one Amplatzer animal there was a pericardial tamponade from a standard delivery wire that perforated the left auricle. This was managed with a pericardial drainage. The rest of the procedures were without immediate complications and all the animals had uneventful recoveries. So far, there has not been any sign of residual shunting. With the sacrifice of the 6 month group still pending, we have only seen one long-term complication: In one animal, a wire on the left atrial side of a SolySafe device was broken and had embolized to the left ventricle. The rest of the results are still pending or only partially analyzed, but will be ready for presentation at the meeting.

Conclusions: We believe this device to be a promising new design for the closure of atrial septal defects. It has a very low profile and with properties that will place it somewhere in between a non- and a self-centering device.

Session 20: Fetal Cardiology

O20-1

Sensitivity and specificity of foramen ovale and ductus arteriosus restriction to predict postnatal status in fetuses with transposition of the great arteries

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Background: Although prenatal diagnosis of transposition of the great arteries (TGA) reduces neonatal mortality, the preoperative course can be complicated in infants with a restrictive foramen ovale (FO) or ductus arteriosus (DA) constriction. We sought to determine the specificity and sensitivity of prenatal features of physiological shunts in predicting postnatal clinical status in prenatally diagnosed TGA delivered in a tertiary care center providing all facilities for neonatal urgent care.

Methods and Results: The outcomes of 130 fetuses with TGA were reviewed over a period of 5.5 years. Restriction of the FO and/or constriction of the DA could be analyzed in 119/130 fetuses at 36 ± 2.7 weeks of gestation. Twenty-four/119 had at least one abnormal shunt (23 FO, 5 DA, and 4 both). Thirteen/130 neonates had profound hypoxemia ($PaO_2 < 25$ mmHg) and metabolic acidosis (pH < 7.15) in the first 30 minutes and required immediate balloon atrioseptotomy. Two of them who had abnormal FO and DA died despite aggressive resuscitation. The specificity and sensitivity of the fetal echo in predicting neonatal emergency were 84% and 54% respectively. The specificity and sensitivity of a combination of restrictive FO and DA constriction were 100% and 31% respectively.

Conclusions: Prenatal diagnosis of TGA does not eliminate the risk of death. Restriction of the FO and/or of the DA has a high specificity to predict the need for emergency care but the sensitivity is too low to detect all high-risk fetuses. Exceptional procedures should be considered for fetuses that have a combination of restrictive FO and DA constriction.

O20-2

Fetal pulmonary stenosis and pulmonary atresia with intact ventricular septum: determinants of biventricular repair

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Objectives: Fetal pulmonary stenosis (PS) and pulmonary atresia with intact septum (PAIVS) represent a spectrum of severity. Outflow obstruction may result in ventricular hypoplasia. In the era of fetal intervention, predictors of postnatal outcome are crucial to select which fetuses may benefit from and require antenatal intervention. We assessed the value of markers in predicting suitability for biventricular (BV) versus single ventricle (SV) repair in fetal PS and PAIVS.

Methods: Review of all cases of fetal PS and PAIVS, diagnosed at our centre since 1995. Inclusion criteria were: (1) pulmonary valve stenosis or atresia, (2) intact ventricular septum, and (3) serial follow-up to birth. The following parameters were assessed at diagnosis: ventricular dimensions and systolic function, flow direction in the arterial duct, the presence endocardial fibroelastosis (EFE) and right ventricular sinusoids. Analysis of inferior vena cava (IVC) Doppler flow to assess right ventricular (RV) diastolic function included integrated time velocity of systolic forward flow to diastolic flow ratio (S/D VTI) and the ratio of reversed to forward IVC flow [A/(S + D)]. Depending on postnatal intervention, 2 patient groups were created and parameters compared.

Results: Of 30 cases identified, 10 were excluded because of termination of pregnancy (n = 8), non-cardiac fetal or postnatal demise (n = 2). Gestational age at diagnosis for biventricular repair was 25.2 ± 6.6 weeks and for single ventricle repair was 29.5 ± 5.8 weeks (NS).

	BV repair (n = 14)	SV repair (n = 6)	P-values
PS⋆	10	0	
PAIVS or critical PS⋆	4	6	⋆0.01
RV/LV length ratio	0.82 ± 0.21	0.49 ± 0.1	0.001
Apex-forming RV	11/14	1/6	0.02
RV–EFE	2/14	4/6	0.04
TV/MV ratio	0.97 ± 0.2	0.5 ± 0.14	0.001
TV inflow duration (ms)	162.6 ± 34.4	110.62 ± 21.8	0.002
PDA flow reversal	10/14	6/6	NS
IVC S/D VTI ratio	4.16 ± 2.04	2.58 ± 1.24	0.04

Conclusions: At the time of prenatal diagnosis of PS and PAIVS, various anatomical and functional findings are useful in predicting postnatal outcome. Baseline RV/LV length ratio, TV inflow duration and IVC S/D VTI ratio provide useful markers of RV growth potential and suitability for biventricular repair.

O20-3

Clinical features, management and outcomes of children with fetal versus postnatal diagnosis of atrial isomerism (heterotaxy syndromes)

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Objectives: The aim of the study is to compare the features, management and outcomes of a large cohort with fetal (F) and postnatal (PN) diagnosis of left (LAI) and right (RAI) atrial isomerism.

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Methods: The records and echocardiographic studies of all cases with the diagnosis of atrial isomerism between 1990 and 2003 at one center were reviewed. Characteristics and outcomes were compared for F versus PN diagnosis and for LAI versus RAI.
Results: There was a total of 176 patients, 72 (41%) patients were diagnosed prenatally. LAI (n = 104) was associated with interrupted inferior caval vein in 92%, AV septal defect (AVSD) in 57%, right (31%) or left (22%) ventricular outflow obstruction (OTO), anomalous pulmonary veins (APVC) in 35%, double outlet right ventricle (DORV) in 29% and complete heart block (CHB) in 9%. RAI (n = 72) was mainly associated with AVSD in 86%, right OTO in 88%, APVC in 78%, and DORV in 47%. Major arrhythmias, left OTO and extracardiac defects were uncommon in RAI. Management and outcomes are described in Table. Survival estimates of live births with LAI were 67.3%, 61% and 53.9% at 1 month, 1 year and 5 years respectively, compared to RAI with 56%, 31.5% and 23.3% (p = 0.0004).

Table: Management and outcomes of atrial isomerism (heterotaxy syndromes).

	LAI (F) n = 47	LAI (PN) n = 57	RAI (F) n = 25	RAI (PN) n = 47	P value (F vs PN)	P value (LAI vs RAI)
Prenatal demise	22/47	N/A	15/25	N/A	N/A	NS
Extracardiac defects	14/47	11/57	0/25	4/47	–	<0.001
CHB	9/47*	0/57	0/25	0/47	–	<0.05*
Livebirth	25/47	57/57	10/25	47/47	–	–
Compassionate care	6/25	9/57	4/10*	9/47*	<0.01*	<0.05
Cardiac surgery	12/25	34/57	6/10	38/47	NS	<0.02
Non-cardiac surgery	4/25	7/57	0/10	0/47	NS	<0.01
Healthy, no intervention	5/25	7/57	0/10	0/47	NS	<0.01

Conclusions: Prenatal demise due to spontaneous fetal death or pregnancy termination was high in RAI and LAI. Compassionate care was more common (p < 0.01) in the RAI group. Disease and surgery related mortality was higher in RAI due to more severe lesions.

O20-4
Fetal arrhythmia complicated by hydrops: neurological follow-up

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Objectives: Fetal arrhythmias are associated with congestive heart failure and development of fetal hydrops, which may result in neurological morbidity and mortality. Limited data exists on the long-term outcome of hydropic fetuses.
Methods: A retrospective study on cognitive and neurological functioning of 16 infants, aged 0,5 to 12 years, who experienced fetal arrhythmias complicated by hydrops.
Results: Seven fetuses had supraventricular tachycardia, 3 had atrial flutter, 1 had ventricular tachycardia and 5 had congenital complete atrioventricular block (CCAVB). Mean GA at birth was 35 weeks and 5 days. Nine fetuses with tachycardia converted to sinus rhythm in a mean time of 7,9 days; resolution of hydrops was achieved in 6 of these patients in a mean time of 7,4 days. Neonatal cranial ultrasound was normal in 7 infants and all but one of these were normal at follow-up: one infant showed a focal thalamic infarction, first seen by the end of the first week, and developed multiple cerebral lesions as a result of a malignant LQTS and died at the age of 2 years. Five infants had flaring on neonatal cranial ultrasound, one associated with a subependymal pseudocyst, and one with calcifications due to a congenital CMV infection. Two of these infants were normal at follow-up, one died two days after birth as a result of withdrawal of therapy, and one infant showed mild global delay. One infant showed evidence of a parenchymal haemorrhage of antenatal onset, presenting as a unilateral porencephalic cyst. He developed a mild hemiplegia with normal cognition. Three infants with CCAVB without cranial ultrasounds were normal at follow-up.
Conclusions: Fetal arrhythmias complicated by hydrops are thought to predispose the unborn child to neurological damage. However, in this series 13 out of 16 infants were neurologically normal. Prognosis seems particularly good in case of successful treatment of tachycardia, delivery at term, and in case of CCAVB. Initiation of therapy should not be withheld or delayed on the assumption of poor neurological outcome.

O20-5
Prenatal diagnosis and outcome of transposition of the great arteries

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Objectives: To document the prenatal diagnostic features and outcome of transposition of the great arteries.
Setting: A tertiary fetal cardiology centre.
Study Design: Retrospective analysis between January 1994 and November 2003.
Patients: During the study period a total of 14,466 pregnancies were evaluated and 1835 cases of congenital heart disease were identified. Of these, 119 fetuses had ventriculo-arterial discordance. In 64 cases this was associated with complex disease including double inlet left ventricle, mitral atresia, tricuspid atresia, atrioventricular discordance or AVSD. These fetuses were excluded. 55 fetuses were identified with AV concordance and ventriculo-arterial discordance. This group was analysed in detail.
Results: Three pregnancies are ongoing. Six pregnancies ended in termination of whom two had an anticipated single ventricle type of repair and one family had a previous infant death due to TGA. 46 infants were liveborn of whom one was lost to follow-up. 24 infants had "simple" transposition, 9 TGA/VSD, 1 TGA/PS, 6 TGA/VSD/PS, 1 TGA/coarctation and 4 had TGA/VSD/coarctation. Discrepancies in prenatal and postnatal diagnoses were noted in 4 cases related to the presence of VSD (n = 3) or coarctation of the aorta (n = 1). In all cases, postnatal imaging confirmed the major prenatal finding of ventriculo-arterial discordance. On the basis of intention to treat, the 30 day survival was 42/45 (93%) and one year survival was 39/45 (87%). There was one pre-operative death of an infant with an associated left diaphragmatic hernia. Two children have a single ventricle physiology and one had a Senning operation. 23 infants had an arterial switch for simple TGA of whom 21 survived. Both of these deaths were related to coronary artery anatomy. All 9 babies with TGA/VSD survived, 4/6 infants with TGA/VSD/PS survived and 3/4 infants with TGA/VSD/Coarctation survived.
Conclusions: Prenatal diagnosis of VA discordance is very accurate. Despite good results for the arterial switch operation, there remains a significant mortality related to extra-cardiac abnormality, associated cardiac lesions and coronary artery anatomy.

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O20-6

Atrioventricular and ventriculo-arterial discordance: echocardiographic features, associations and outcome in 28 fetuses

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This retrospective study aimed to identify common features of atrioventricular and ventriculo-arterial discordance to help improve the prenatal detection and diagnosis of this condition. Associated cardiac malformations and outcome were also evaluated.

Material and Methods: The study population comprised of 28 fetuses with a prenatal diagnosis of double discordance made at a tertiary centre for fetal cardiology between January 1993 and December 2003. The diagnosis was confirmed in 23, either postnatally or at post mortem examination. In the other 5 cases permission for autopsy was refused. Videotape recordings of fetal echocardiograms were analysed to establish features common to this condition. These included reversed differential insertion of the atrioventricular valves, identification of the right ventricle via the moderator band, the origin and orientation of the great arteries. Associated cardiac abnormalities and outcome were also noted.

Results: 27 cases were referred with suspected congenital heart disease and 1 because of a family history. The mean gestational age at presentation was 20 weeks (range 15–26). Reversed differential insertion of the atrioventricular valves was identified in 21 cases, and the right ventricle, which was left sided in all cases, identified by the moderator band in 24. A parallel or abnormal orientation of the great arteries was seen in all cases, with the pulmonary artery being the first vessel seen moving cranially from four chambers in all. Only 4/28 cases were isolated. There was a ventricular septal defect in 18, Ebstein's anomaly in 4, pulmonary stenosis in 10, coarctation in 3 and tricuspid atresia in 1. Congential heart block was present in 1 case prenatally and a further 2 postnatally. The cardiothoracic ratio was increased in 2/28 cases, both with Ebstein's anomaly. There was mediastinal shift to the right in 7/28. Outcome: There are currently 17 survivors, one baby died in the neonatal period and 2 in infancy. The pregnancy was interrupted in 8 cases.

Conclusions: Abnormal orientation of the great arteries in association with the finding of a left sided right ventricle, identified by reversed differential insertion or the moderator band should alert a sonographer to this diagnosis. Isolated cases are rare prenatally, but short term survival in our series is good with 17/20 babies where pregnancy continued still alive.

Session 21: Adult Congenital Heart Diseases

O21-1

MRI controlled cardiac catheterisation for assessment of pulmonary vascular resistance: in-vivo validation and clinical application in patients with pulmonary hypertension

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Background: Current diagnostic tools for assessment of pulmonary vascular resistance (PVR) are limited in terms of accuracy and reproducibility. Aim of this study was to assess the suitability of a novel MRI method for measuring PVR.

Methods: PVR was determined with a MRI technique, in which pulmonary arterial (PA) pressures were assessed invasively with flow directional catheters and quantitative PA flow volumes were assessed with velocity encoded (VEC) MRI. For pressure acquisition flow directional catheters were guided in a 1.5 Tesla MRI unit under MRI fluoroscopy control into the main PA.

Results: In validating in-vivo studies (8 swine) PVR was determined with thermodilution technique and compared to MRI (0.9 ± 0.5 vs. 1.1 ± 0.3 Wood units$\star m^2$). Bland-Altman test showed agreement between both methods. However, interexamination variability was $7.9 \pm 2.2\%$ for thermodilution, but only $2.1 \pm 0.3\%$ for MRI measurements. After these validating studies, eight patients with pulmonary hypertension (PHT) and six control patients were studied. All patients were transferred after routine cardiac catheterisation to the MRI unit for further assessment of PVR. In patients with PHT measurements were performed at baseline and during inhalation of nitric oxide (NO), 20 ppm. Compared to control, PVR was significantly elevated in the PHT group (1.4 ± 0.8, 13.1 ± 5.1 Wood units$\star m^2$, respectively, $p < 0.001$) but decreased significantly to 10.3 ± 4.6 Wood units$\star m^2$ during inhalation with NO ($p < 0.05$). Interexamination variability of all MRI derived PVR was only $2.6 \pm 0.5\%$. In all experiments (in-vivo and clinical) catheter guidance was performed successfully under MRI control.

Conclusions: Catheter guidance maneuvers such as positioning flow directional catheters in the PA is feasible under MRI control. PVR can be measured accurately and reproducibly with the proposed MRI technique. This investigative approach seems to be well suited for assessment of PVR in a variety of patients with pulmonary hypertension and/or congenital heart disease.

O21-2

Emergencies in adults with congenital heart disease – a complex and interdisciplinary management is necessary!

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Background: The quantity of emergencies in adult patients (pts) with congenital cardiac disease (CCD) reaching is continuously growing.

The aim of this study was to determine, besides the reason and the nature of emergency situations, the personal and technological requirements for an adequate management of these pts.

Study Design: Prospective multi-centre-study, performed at 5 supraregional centers for adults with CCD.

Results: During 12 consecutive months 1033 admissions of adults with CCD occurred to one of the participating centers. A total of 201 admissions (20%) were classified as an "emergency". 70% ($n = 112$) of the pts had been previously operated on. The underlying CCD was: univentricular heart ($n = 45$), tetralogy of Fallot/truncus arteriosus ($n = 43$), transposition of the great arteries ($n = 28$), pressure shunts ($n = 26$), flow shunts ($n = 24$), right or left ventricular outflow obstruction ($n = 15$), others ($n = 20$). The majority of admissions (75%) were due to cardiac arrhythmias,

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congestive heart failure or for infectious reasons. The most often used technical procedures for the clarification of the emergency were: ECG (n = 201), laboratory tests (n = 201), transthoracic echocardiography (n = 154), chest radiography (n = 95), transesophageal echocardiography (n = 69), CT-/ or MRI-imaging (n = 29). In 63% consultants from non-cardiological departments had to be involved: surgery (n = 47), internal medicine (n = 42), neurology (n = 12), ophthalmology/ otorhinolaryngology (n = 11), gynecology/obstetrics (n = 5), and others (n = 11). For treatment 46 pts underwent urgent surgery (heart, brain, general surgery), 24 pts had an electrical cardioversion. Twelve pts deceased. The survivors were hospitalized for 11.3 ± 16.8 days.
Conclusion: The present study verifies for the first time the wide range of diagnostic and therapeutic procedures required during emergency situations in adults with CCD. For an adequate management of these chronically ill pts, especially experienced physicians and centers, but also special diagnostic and therapeutic tools are mandatory. A multidisciplinary diagnostic and therapeutic approach with close affiliation to medical subspecialties is indispensable. These data may provide a basis for guidelines for the treatment of emergency situations in adults with CCD.

O21-3
Risk stratification for heart failure in grown up with congenital heart disease in the long-time follow up after cardiac surgery
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Background: Heart failure (HF) is one of the main problems during long-time follow up of patients with congenital heart disease after cardiac surgery. The purpose of our study was to evaluate the risk of HF according to the individual heart defects and procedures in a group of patients with an age between 14 and 40 years operated in our institution.
Methods: Plasma brain natriuretic peptide (NT-proBNP, immunoassay) and maximal oxygen uptake (VO_2max, spiroergometer) were measured in 172 consecutive patients during a long-time follow up examination. Diagnosis of HF was assumed by the following criteria: elevated NT-proBNP level >100 pg/ml and a reduced VO_2max <25 ml/kg/min.
Results: 46 of 172 patients fulfilled these HF criteria. These patients were significantly older (29.4 ± 8.1 vs 23.9 ± 6.8 years, $p < 0.001$), had significant higher right ventricular (RV) pressures (33 ± 22 vs 20 ± 9 mmHg, $p < 0.001$) and enddiastolic RV diameters (36.8 vs 26.8 mm, $p < 0.001$) determined by echocardiography, but showed no difference in left ventricular shortening fraction (35 ± 7 vs 37 ± 6%, ns). Compared to this overall risk of HF, Odds ratios (OR) were determined for the 10 most frequent diagnoses and procedures for individual risk stratification. For example the risk of HF in patients with tetralogy of Fallot (OR = 2.28) operated with a transanular patch plastic (OR = 3.24) was 5.3 times higher than in those without transanular patch plastic (OR = 0.61).
Conclusions: Heart failure during long time follow up of operated congenital heart defects predominately depends on age and RV function. Our data help to identify risk factors of HF and permit a specific advice of the patients concerned.

O21-4
Employment and career advice in adults with congenital heart disease
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Objectives: To compare employment rates in adults with congenital heart disease (CHD) with controls and at different severities of CHD. To evaluate the career advice given to adults with CHD.
Methods: Two hundred and ninety nine adult CHD patients underwent a questionnaire based interview by a trained nurse. They were asked to give an identical questionnaire to a friend to act as a control. One hundred and seventy seven controls replied. CHD patients were classified into three categories based on disease severity.
Results: Excluding students, maternity and retirement, significantly more of the adults with CHD were unemployed than matched controls (51/156 (33%) vs 25/156 (16%), $p < 0.0001$) and more had been out of work for greater than a year (37/151 (25%) vs 5/151 (3%), $p < 0.0001$). More of the CHD group had gone onto educational/vocational courses (145/174 (83%) vs 123/174 (71%), $p = 0.004$). Similar proportions received careers advice with fewer of those with CHD finding this helpful (26/134 (19%) vs 41/134 (31%), $p = 0.037$) and more with CHD being given advice against certain occupations (71/169 (42%) vs 19/169 (11%), $p < 0.0001$). However a higher proportion of those with CHD who had received career advice were employed than those with CHD who had not (157/216 (73%) advised employed vs 25/54 (46%) not advised, $p = 0.0002$). Unemployment rates were not significantly higher in those with more severe disease (32/98 (32%) mild disease, 40/133 (30%) significant disease, 18/42 (43%) severe disease, $p = 0.57$).
Conclusions: Whatever the disease severity adults with CHD are more likely suffer from unemployment and long term unemployment than matched controls. They are more likely to receive negative career advice and advice that they find less helpful than controls. Education of patients and employers of the limitations that should be placed on those with CHD as well as advice from cardiologists and careers officers at adolescent and adult clinics may improve the employment outcome for adults with CHD.

O21-5
The effect of cigarette smoking on myocardial contractility, preload, and afterload in adolescents
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Tobacco is the most important preventable cardiovascular risk factor in all age groups. Both passive and active exposures to tobacco smoke have adverse cardiovascular consequences. This study assessed echocardiograhic measures of cardiac function in tobacco smoking and healthy adolescents to determine the association of cigarette smoking with myocardial contractility, preload, and afterload. To further investigate these effects, 30 healthy adolescents (M/F: 27/3) aged 16.1 ± 1.8 years and 30 tobacco smoking adolescents (M/F: 28/2) aged 16.2 ± 1.4 years were evaluated with 2-dimensional and M-mode echocardiography. The number of cigarettes smoked per day was 31 ± 7.1 and the mean duration of smoking was 3.1 ± 1.1 years. The rate-corrected velocity of circumferential fiber shortening (VCFc) and end-systolic wall stress (ESWS)

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relation were used as a load-independent estimation of contractility. Preload, afterload and ventricular mass were also measured. Despite similar shortening fractions and ventricular mass, the smoking adolescents group had significantly lower mean VCFc and higher ESWS than the healthy adolescents group (1.09 vs. 1.31 circ/s and 39.2 vs. 31.4 gm/m^2 respectively). There was not difference between two groups' for systolic and diastolic blood pressures and left ventricular ejection fraction. All these findings suggest that these adolescents are at high risk for cardiovascular disease during their lifetime. These findings may be important to determine the effect of smoking in adolescent ages. Unfortunately, tobacco use may be the most prevalent risk factor in youth and the most difficult to treat because of the addicting power of nicotine. There is an evidence that the prevalence of tobacco use is increasing in the adolescents. This means that the risk of cardiovascular complications may also be increasing among adolescents.

O21-6

Effectiveness of antibioprophylaxis for infective endocarditis in patients with congenital heart diseases

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Objective: This study reviews our long-term experience with infective endocarditis in congenital heart diseases and evaluates goodness of fit between protocols and risks, and patients and physicians education.
Material and Methods: From 1966 to 2001, 138 episodes occurred in 125 patients, at a median age of 10.3 years. Sixty-five cases with definite cardiac disease, had experienced a prior known event: 34 cases had applied adequate prophylaxis, 16 did not receive prophylaxis despite risk and 15 were given previous antibiotics toward a minor illness. In the seventy-three remaining episodes, prior event was unknown: of them, cardiac disease was definite in 43 and not suspected in 30.
Results: Repaired cardiopathies accounted for 38 cases, palliated complex cardiopathies for 34 and native cardiopathies for 66 (21 ventricular septal defects, 38 mitral and/or aortic valvulopathies and 7 others). Cardiac surgery, dental and cutaneous causes were the commonest sources of infection. Dental causes were the second most frequent in patients who received adequate prophylaxis and the most frequent in those who did not apply prophylaxis because of physician or patient underestimating the risk. All surgical cases were given prophylaxis. Of the 34 episodes related to dental causes, about 1/4 received adequate prophylaxis, 1/4 did not despite known event and 1/2 did not because of unknown event. Of the 16 episodes related to cutaneous causes, about 1/4 did not receive antibiotics despite known event and 3/4 did not because of unknown event or unsuspected cardiac disease.
Conclusion: Current protocols for endocarditis prophylaxis, although not always protective, seem to be more effective than in the past. Non-compliance and lack of patient and physician education continue to be significant problems which have potential to be improved. Cutaneous infections have to be taken into consideration as important causes of infective endocarditis. Prophylaxis should be emphasized in patients with mild cardiac lesions, in particular with small VSD, where the risk is likely to be underestimated by both patients and physicians.

Session 22: Interventional Cardiology III

O22-1

Breakable stent for interventions in infants and children: an animal study and histopathological findings

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Objectives: Use of stents in infants and children is limited due to growth of the stented vessel. Enlargement of the diameter of the stented segment by re-dilatation is restricted by design and stability of the stent. Therefore we developed a breakable stent that can be broken open by angioplasty when needed.
Material and Methods: Medical grade stainless steel stents (n = 16) were interventionally implanted in the subclavian or carotid artery in neonatal pigs (n = 7). Patency of the stented segments was demonstrated by repeated angiography. Re-dilatation was performed up to 4 times. Between 18 and 162 days after implantation the animals were killed and the vessel containing the stent removed. Besides standard histology we employed scanning electron microscopy (SEM) for biocompatibiliy screening.
Results: Implantation was performed without complications in all animals. "Breaking" of the stents was achieved by re-dilatation using a conventional angioplasty balloon catheter. During follow up patency of all stented segments was demonstrated angiographically. SEM demonstrated complete cellular coverage of the stent strats. At the site of breaking of the stent thinning of the vessel wall and partial rupture of the media was seen histologically. An only mild inflammatory reaction was detected.
Conclusions: The new breakable stent can be broken open by simple angioplasty. Feasibility and biocompatibility was demonstrated in an animal model. Surgery for removal of a stent in an infant or child due to disproportion of a formerly implanted stent and the vessel during growth may be postponed or even avoided by the use of a breakable stent.

O22-2

Stents in the treatment of aortic coarctation: initial results and intermediate-term follow-up

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Seventy four patients with different pathologies of the aorta were treated with stent implantation. The purpose of the study was to assess the value of balloon expandable intravascular stents in treatment of native and recurrent aortic coarctation. Between 1996 and 2003, 54 patients, 38 with native and 16 with recoarctation after various types of surgical repairs, were successfully treated with Palmaz (39) and CP (15) stents in three Polish cardiac centers. Mean age was 18,38 ± 12,12 years (range 5 to 52 yrs). In 19 pts aortic coarctation was diagnosed incidentally during standard pediatric examination. Mean stent to narrowest aortic diameter ratio was 3,24 ± 1,39. After stent implantation peak systolic pressure gradient decreased from 45,11 ± 18,93 mmHg to 7,39 ± 9,95 mmHg

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($p < 0.001$), narrowest diameter increased from 5,3 ± 2,25 mmHg to 15,2 ± 3,29 mmHg ($p < 0.001$). Technical complications occurred in 5 pts: stent dislocation during implantation – 2 pts, surgical iliac arteriotomy for balloon-stent removal in an incompletely expanded stent caused by a balloon rupture – 1 pt and minor complications related to artery puncture in 2 pts. Mean follow-up is 25,37 ± 24,03 months (range: 2 to 96 months). Nine patients underwent 12 successful stent re-dilations 3–54 months after initial implantation (neointimal hyperplasia, patient growth). In 2 pts CP stent fracture was detected 16 and 36 months after implantation with no clinical significance. Mean systolic blood pressure decreased from 149,26 ± 23,64 mmHg before stent placement to 122,4 ± 10,67 mmHg at follow-up ($p < 0.001$). Twenty seven patients (50%) require pharmacological blood pressure control. CT angiography performed in 27 pts showed normal location of stents with no evidence of aneurysm. In 4 pts neointimal hyperplasia and in 2 pts fractured stents were documented in CT during follow-up.
Conclusions: Balloon expandable intravascular stents implantation for native and recurrent aortic coarctation has good results in the short and intermediate terms. Successfull redilation of stents implanted to aorta is possible. Technical complications during stent implantation and during follow-up can occur. Long-term outcome of bigger number of patients has to be evaluated for determining the role of transcatheter stent therapy for native and recurrent coarctation of aorta.

O22-3
10 year experience of transluminal treatment of severe congenital stenosis – hypoplasia of branch pulmonary arteries or MAPCA's
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Background: Severe congenital stenosis or hypoplasia of branch pulmonary arteries (PA) or MAPCA's is frequently not amenable to surgical management. Some lesions can be treated with balloon dilation and/or stent implantation, however discrete segments are often resistant to high pressures or large balloons, which increases the risk of complications (dissection, aneurysm formation, disruption of the adjacent vessel wall).
Methods and Results: From 1993 to 2003 106 hypoplastic or congenitally stenosed PA or MAPCA's were dilated in 25 ps (Fallot variants 22; familial PPS 3) during 70 procedures (median age 5.25 y, range 0.7–27.2). Conventional balloons were used in 84 vessels (median diameter 6 mm, range 3–15); 18 vessels had stenoses resistant to high pressures (18 atm), moderate residual stenosis or severe recoil was seen in 20 cases. Severe aneurysm formation (>50% of desired diameter) was seen in 2 cases and significant dissection in 2 vessels (causing loss of segmental lung perfusion). After 2001 Cutting Balloons (CB Boston Scientific, current sizes 2–8 mm) were used in 22 vessels. Standard CB angioplasty (3–7 mm) was used in 18 cases; 4 vessels were treated with a CB catheter augmenting technique (2001–2003: 4 mm CB side-by-side to 5–6 F dilator). All vessels treated with CB angioplasty could successfully be dilated, including those who repeatedly had shown to be pressure resistant. During dilation no episodes of bleeding, perforation or rupture were experienced. Subsequent stent implantation (4–16 mm) was performed in 21 patients after conventional balloon angioplasty and in 16 patients after CB dilation. Indication for stent implantation was significant vessel recoil, severe intimal damage with aneurysm formation or dissection and risk of subsequent loss of vessel patency, and critical filiform long segment narrowing. Medium term follow-up of stents was excellent in short straight lesions, however 4 of 12 curved or long stents had thrombosed.
Conclusions: Conventional balloon angioplasty often gives suboptimal results predominantly due to pressure resistant stenosis. CB angioplasty is indespensable for treatment of pressure resistant lesions; it is currently also our preferred treatment for filiform or long segment stenosis. Stents give very satisfactory results in short and straight lesions.

O22-4
Retrospective analysis of periprocedural and follow-up complication rates of 708 consecutive pediatric transcatheter interventions
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Purpose: To evaluate the incidence of complications associated with transcatheter therapy in pediatric patients with congenital heart defects.
Methods: Retrospective analysis of cardiac catheterizations performed during 1.9.1997 to 31.8.1999. A total of 1245 catheterizations were screened and 708 interventions identified. Catheterization records, discharge summaries and reports of the first postinterventional outpatient visits were analyzed and possible complications recorded and classified.
Results: A total of 45 (6,4%) complications were identified. 23 (3,2%) were classified as 'major' 22 (3,1%) as 'minor'. 16 (2,3%) complications were identified within the periinterventional period, 29 (4,1%) were identified during the outpatient visit. Transcatheter therapy of valvar aortic stenosis was associated with the highest complication rate 14/30 (46%). 13/30 patients were diagnosed with a de-novo aortic regurgitation I–II, 1 patient had a severe de-novo aortic regurgitation. In patients with pulmonary atresia, radiofrequency perforation of the atretic pulmonary valve was associated with 2 pericardial effusions and 1 occlusion of the femoral artery. Stent-implantationen into the PDA led to the occlusion of the femoral artery in 2 patients, and was treated by transcatehter recanalization. Further procedures are discussed with corresponding acute and follow-up complication rates.
Conclusion: Despite more complex interventions potentially lead to an increased incidence of untowards events, transcatheter therapy of congenital heart defects is associated with acceptable complication rates. Vascular complications remain the most frequently encountered complications.

O22-5
Transcatheter treatment of ventricular septal defects: 8 years experience
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From 1995 to 2003, 21 patients (pts) with ventricular septal defect (VSD) received transcatheter treatment in our Department. The VSD was congenital in 14 pts and followed an acute myocardial infarction in 7 pts.

Congenital VSD

The mean age and weight of the 14 pts were 11 ± 12 years (range 2 months–44 years) and 28 ± 20 (range 3,4–67) Kg. The VSD was located in the muscular septum (MVSD) in 8 pts and in the perimembranous septum (PMVSD) in 6 pts. The mean QP/QS was 2.4 ± 1.6. The procedure was completed in all the 14 cases.

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The devices implanted were: 1 PDA Rashkind, 1 Cardioseal, 6 Amplatzer muscular in MVSD and 1 Amplatzer muscular, 5 Amplatzer Membranous in PMVSD. There were no major complications. At the end of the procedure the VSD was completely occluded in 6 pts (42%) and mild residual shunt was still present in 8 pts (58%). In 1 patient, uncomplicated surgical device removal and closure of the VSD was performed, because of device displacement a few hours after the implantation. The mean follow-up period of 13 pts is of 20 ± 32 months. Echo Color Doppler showed complete occlusion of the VSD in 11 cases (85%) and mild residual shunt in 2 pts (15%).

Post myocardial infarction VSD

The mean age of the 7 pts was 79 ± 7 years. In 5 pts the procedure was performed 7 ± 2 (range 4–9) days after the acute cardiac event. Two pts with residual shunt after surgical closure of the VSD were treated 4 and 6 months after the myocardial infarction respectively. The procedure failed in 1 patient for the too large size of the defect. The device (1 Cardioseal, 3 Amplatzer ASO and 2 Amplatzer muscular) was successfully positioned in 6 pts. Two pts died after device deployment for cardiac rupture. Of the 4 pts with successful closure, 2 died few days after the implant for complications unrelated to the procedure. Two pts (12%), show good result at follow up of 7 and 4 years respectively.

Conclusions: Percutaneous closure of congenital muscular and perimembranous VSD is safe and effective and should be considered the treatment of choice in selected patients. In the postinfarction defects the results strictly depend on the conditions of the pts and on the timing of the procedure: the subacute phase seems the best option for successful treatment.

O22-6

Transcatheter device closure of perimembranous ventricular septal defects: medium term follow up

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Introduction: Transcatheter device closure of perimembranous ventricular septal defects (VSDs) carries a theoretical risk of late complications including heart block, bundle branch block and aortic and tricuspid regurgitation.

Methods: A retrospective case note review was performed on a cohort of patients who underwent successful closure of VSD with a 17 mm rashkind ductal occluder between 1991 and 1996 at our institution.

Results: 14 patients had a device successfully implanted. 3 underwent device removal and surgical closure of their VSD at 1.5, 5 and 15 months post procedure for a significant residual VSD (2) and aortic regurgitation (1). 11 patients have been followed up for a median of 10 years (range 4–12 years). Age at implantation was median 84 months (range 1–300 months) and weight 3.5–69 kg. All are in NYHA functional class 1 and none have required further intervention. Each patient is in sustained sinus rhythm (right bundle branch block in 3) and has a normal PR interval (range 0.12–0.2 ms). All have qualitatively normal function on echocardiogram. 6 have small, haemodynamically insignificant VSDs, 1 has mild aortic regurgitation which has been unchanged in 120 months follow up and 4 have mild tricuspid regurgitation.

Conclusions: Medium term follow up of a small group of patients in whom a device effectively occluded a perimembranous VSD has not shown significant complications. This suggests that the current Amplatzer™ membranous VSD occluder is likely to be safe and effective in the medium term.

Session 23: General Cardiology II

O23-1

Outcome following preterm delivery of infants antenatally diagnosed with congenital heart disease

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Objectives: To determine outcome following delivery prior to 36 weeks gestation in babies diagnosed antenatally with significant congenital heart disease (CHD).

Material and Methods: Databases at each hospital were reviewed to identify the total number of women referred for fetal echocardiography over a 5 year period. Fetuses diagnosed with significant CHD were identified (minor lesions such as small muscular ventricular septal defects and mild ventricular disproportion were excluded). Details of the neonatal course and outcome were obtained for those live born prior to 36 weeks.

Results: Between January 1998 and December 2002, 9918 women were referred for a fetal echocardiogram at a median gestation of 21 weeks (range 14–33 weeks). Significant CHD was diagnosed in 1191 fetuses (12%), and of these, 46 (4%) delivered prematurely. The major indication for referral was a suspicion of CHD on routine ultrasonography (39/46). Other indications included extracardiac or karyotypic abnormalities (3), a family history of CHD (1), and maternal illness (3). Delivery was precipitated by preterm labour in 28, fetal compromise in 7, pre-eclampsia in 4, chorioamnionitis in 2, and severe polyhydramnios in 1 (cause not recorded in 4). Median gestation at delivery was 33 weeks (range 24–35 weeks), and median birth weight was 1.56 kg (range 0.50–3.59 kg). Extracardiac anomalies were found in 23 (50%), of which 3 also had karyotypic anomalies. Twenty four babies underwent neonatal surgery: 16 underwent a cardiac procedure, 5 a general surgical procedure, and 3 underwent both. Eight died in the immediate postoperative period (33%). Two babies with critical right heart obstruction underwent interventional catheters, following which both died. Overall, 33 (72%) of babies died. Extracardiac/karyotypic anomalies increased the relative risk of death by a factor of 1.36. Survival by diagnostic group is shown in Table 1. Mean (se) hospital stay for those surviving to initial discharge was 41 (7.1) days.

Table 1. Survival by diagnostic group (VSD, ventricular septal defect; PS, pulmonary stenosis).

Diagnosis	Survivors/ total	Diagnosis	Survivors/ total
Isolated VSD	1/2	Transposition and congenitally corrected transposition	2/4
Atrioventricular septal defect	0/6	Hypoplastic left heart syndrome	0/7
Ebstein's anomaly	0/1	Coarctation/interruption of the aorta	3/5
Tricuspid atresia	1/2	Common arterial trunk	0/3
Fallot/pulmonary atresia with VSD	3/6	Complex	2/6
Critical PS/pulmonary atresia with intact septum	1/3	Cardiac tumour	0/1
		Total	13/46

Conclusions: There is a very significant morbidity and mortality for babies antenatally diagnosed with significant CHD who deliver prematurely, particularly for those with extracardiac or karyotypic anomalies. The outlook for such babies must be very guarded, and

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this should be reflected when making decisions over elective preterm delivery, and when counselling parents both ante- and postnatally.

O23-2

Timing of presentation and postnatal outcome of infants suspected of having coarctation of the aorta during fetal life

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Objective: To report the timing of presentation and clinical profile of a cohort of fetuses with normal main cardiac connections but fetal echocardiographic signs suggestive of coarctation of the aorta.
Design: A retrospective observational study.
Setting: A tertiary fetal and paediatric cardiology centre.
Patients: Between 1 January 1998 and 31 December 2002, 174 fetuses were included, of whom 144 infants were liveborn.
Main Outcome Measures: Of the 144 liveborn infants, 43 had coarctation of the aorta, four had interruption of the aortic arch and one was managed as hypoplastic left heart syndrome. Two infants were diagnosed with hemianomalous pulmonary venous drainage. Three infants with coarctation presented late at 7–13 weeks of age, 6–12 weeks after closure of the arterial duct. The incidence of left superior vena cava and ventricular septal defects did not differ between false positive and true positive diagnoses of coarctation. Fetuses with cardiac asymmetry had a higher incidence of left superior vena cava than a control group. The 30 day and one year surgical mortality of infants having repair of coarctation of the aorta was two of 41 (4.9%, 95% CI 0.6–16.0). All cause mortality of liveborn infants with any abnormality of the aortic arch was 5/48 (10.4%, 95% CI 3.5–22.7) at 30 days and one year, which was heavily influenced by prematurity and extra-cardiac abnormalities.
Conclusions: Precise diagnosis of coarctation of the aorta during fetal life remains difficult. Coarctation of the aorta may present several weeks after closure of the arterial duct and sequential echocardiography is recommended.

O23-3

Poor sensitivity of routine fetal anomaly ultrasound screening for antenatal detection of atrioventricular septal defect (AVSD)

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Objective: To assess antenatal diagnosis rate in a cohort of children born alive with atrioventricular septal defect (AVSD).
Setting: The Yorkshire heart centre, the sole centre providing fetal echocardiography and specialist management of patients with congenital heart disease for our region (population of 5.25 million).
Methods: Identification of all patients with AVSD's born between 1996 and 2001 by a search of prospectively collected, comprehensive diagnostic information held in the departmental database. Information relating to detailed fetal echocardiography was retrieved from the same database.

A four chamber view of the heart (as minimum) was obtained as part of standard obstetric ultrasound for all pregnancies in all referring hospitals. Detailed fetal echocardiography was offered to all patients with abnormal, possibly abnormal or difficult four-chamber views as well as for patients with a family history, markers for aneuploidy or extracardiac malformations.

Results: 92 children were live born with AVSD's. 64 (69%) had complete defects, 11 (12%) partial and 17 (18%) AVSD in association with complex intracardiac defects. 27 of 92 live born cases (29.3%, 95 CI 18.4–35.6) were detected by routine obstetric ultrasound screening – all were referred for detailed fetal echocardiography. The detection rate for AVSD in association with complex congenital heart disease was higher than for those with AVSD alone, 9/18 (50%, 95% CI 4.8–13.2) vs 18/74 (24%, 95% CI 10.8–25) ($p = 0.03$). 49/92 patients (53%) had Downs syndrome. 12/49 (24.5%, 95% CI 6.1–17.9) of these patients (AVSD associated with Downs syndrome) were diagnosed antenatally at routine obstetric screening versus an antenatal diagnosis rate of 15/43 (34.9%, 95% CI 8.8–21.1) for chromosomally normal children (p = NS).
Conclusion: Antenatal detection rate for AVSD by standard ultrasound screening using the four chamber view is poor overall, with a trend towards being even poorer in the presence of trisomy 21.

O23-4

A population-based study of cardiac malformations and outcomes associated with dextrocardia

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Background: The incidence of dextrocardia (D) and its associated cardiac and non-cardiac malformations are not known. There is inadequate information about outcomes to counsel parents about prognosis.
Methods: A retrospective review of all diagnoses of D at our provincial tertiary care hospital between 1985 and 2001 was performed. Cardiac and non-cardiac malformations, follow-up, and outcomes were determined. Dextrocardia was defined as the heart's position being in the right hemi-thorax due to embryological development. The incidence of D was calculated using population statistics.
Results: Eighty-two cases were identified (48 ante-natally). The incidence of D was estimated to be 1 in 11,871 pregnancies. Twenty-seven cases were situs solitus (SS), 31 situs inversus (SI), and 24 situs ambiguus/isomerism (SA). Cardiac malformations were found in 26/27 cases of SS, 8/31 SI, and 24/24 SA. The most common were septal defects (ASD/VSD/AVSD) (n = 60), abnormal pulmonary arteries (n = 30), abnormal pulmonary valves (n = 25), and abnormal venous return (n = 24). There were 17 univentricular hearts. Non-cardiac malformations were identified in 11/27 SS, 6/31 SI, and 13/24 SA. The most common non-cardiac malformation was gastro-intestinal obstruction (n = 15). Vertebral abnormalities (n = 3), hydrocephalus (n = 2), pulmonary hypoplasia (n = 2), Meckel's diverticulum (n = 2) and renal abnormalities (n = 2) were less common. There were two cases of Kartagener's syndrome and no chromosomal abnormalities were detected. Eleven pregnancies were terminated, one aborted spontaneously, three were stillborn, and two chose compassionate care. All terminated cases were diagnosed with D prior to termination and all involved more than one cardiac anomaly; seven also had non-cardiac anomalies. There were 44 cases in the intention-to-treat group (SS = 21, SI = 11, SA = 12). Thirty-nine had at least one cardiac operation and 18 had at least three. Forty cases were alive at most recent follow-up and four were deceased (SS = 2 and SA = 2).
Conclusions: The incidence of D was 1 in 11,871 pregnancies. In our cohort, the number of cases of SS, SI, and SA were similar. Cardiac and non-cardiac malformations were most common in SA. Cardiac malformations were often complex in SS and SA. Despite this, 91% of the intention-to-treat cohort were alive at follow-up.

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O23-5
Survival and quality of life of children with an univentricular heart in a single institution
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Background: Management of patients with univentricular heart (UH) has changed dramatically over the last years, influencing survival and quality of life (QoL).
Objectives: Retrospective evaluation of survival and quality of life of patients with UH, treated in 1 institution.
Methods and Results: Between 1952 and 2002, 193 patients with an UH were identified and enrolled to the study. Medical records were reviewed. Written and telephone contact was taken to evaluate QoL, using the SF-36 score. 43% patients were female, 57% were male. The 'single' ventricle had a left morphology in 67%. Mean follow up was 7.7 + 10.8 years (median 3.9, range 0–49.8). 153 patients (79%) had surgery, including a total cavopulmonary connection (TCPC) in 44%. In the non-TCPC group (56%), 37 patients (43.5%) had a superior cavopulmonary connection. Overall mortality was 48% (92 patients), with a Kaplan Meier estimate of survival at 10 years of 51%. Mortality in the surgical group was 40.5%. The survival probability for the TCPC group at 5 years after surgery was 81% and at 10 years 77%. If we divide the TCPC group in those who were operated before and after 1991, the total mortality was 60% and 4.9% respectively, with a survival probability at 10 years after surgery was 45% and 95% ($p < 0.05$) and at 10 years after birth was 70% and 95% ($p < 0.05$). In terms of QoL, 21 out of the 45 patients eligible for the test (16 years old at the time of the study), accepted so far to answer. Mean age of this group was 24 + 8.3 years. 48% were male, 52% female. 86% patients assist regularly to school or to a paid job. The 3 remaining have a neurological problem and don't work nor study, with 1 of them living in an specialized institution. 57% live with their parents, but they all are less than 18.5 years old. With respect to scores in QoL, significant differences existed in physical activity ($p < 0.05$), physical health ($p < 0.05$), social adaptation ($p < 0.05$), and mental health ($p < 0.05$), compared to general population, but more than 70% of the patients were still within normal limits. No significant differences were found in pain perception, vitality, and emotional status.
Conclusions: Survival probability has dramatically improved with years. Although our results still show a difference in quality of life, our patients have a completely normal life in the most of cases.

O23-6
Shift of risks in neonatal congenital heart disease? Analysis of institutional experience with transposition of the great arteries
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Objectives: The mortality of corrective neonatal cardiac surgery decreased dramatically during the last years. An analysis of the institutional experience with transposition of the great arteries (TGA) should show which problems exist in these patients (pts) now.
Methods: Retrospective analysis of all 60 pts with both simple and complex TGA born from 08/1998 to 12/2003 and anatomically eligible for arterial switch. Pts requiring Rastelli or atrial correction were excluded. Surgery was performed by the same surgeon on all cases in hospital and follow up records were reviewed.
Results: Complete follow up was available with a median of 21 months (1 to 64 months). Mortality: 1 pt with simple TGA, extracardiac and chromosomal anomalies developed severe postnatal capillary leak syndrome of unknown origin and died at the age of 5 weeks. One pt with simple TGA died early after arterial switch (surgical mortality 1/59 or 1.7%). There were no late deaths. Preoperative morbidity: poor mixing despite balloon atrioseptostomy required continuation of prostaglandin in 14, re-septostomy in 2 and stenting of the restrictive duct in 2 pts. Surprisingly, there were 4 pts (6.7%) with simple TGA who were diagnosed only after resuscitation (2) or with critical cyanosis (2) at home, 3 of them were older than 4 weeks and had to have palliative banding and shunt prior to secondary switch. An elective secondary switch was performed in 4 pts with TGA, VSD and coarctation. Postoperative morbidity: four procedures of redo surgery had to be performed in 3 pts (5%). Cerebral sequelae are known in 3 pts (5%), 1 developed hydrocephalus requiring a ventriculoperitoneal shunt, 2 show impaired statomotoric development and seizures.
Conclusions: Surgical mortality of neonates was the main concern in the past but has decreased in this center as well as in the literature. However, there were some patients with critical preoperative morbidity due to late detection of the cardiac defect. Corresponding to alarming reports from neonatologists of the same region these cases might be the result of a rapidly changing culture with reluctant attitude towards in-hospital birth and the loss of a so far well-established evaluation system for newborns. This may require additional training and education efforts for all involved in neonatal care.

Session 24: General Cardiology III

O24-1
Arterial stiffness and endothelial dysfunction in HIV-infected children
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Background: The role of antiretroviral therapy on acceleration of atherosclerosis in HIV-infected adults is controversial, partly because of the confounding effects of the involvement of classic cardiovascular risk factors.
Objectives: To study the impact on vascular function of HIV infection and antiretroviral treatment in children.
Design: Transversal study in 49 HIV-infected children (34 receiving antiretroviral therapy and 15 never treated) and in 24 age- and sex-matched controls.
Methods: Automatic, computerized, ultrasonic procedure evaluation of geometric and mechanical properties of the common carotid artery, and of the endothelium-dependent dilation and endothelium-independent dilation.
Results: Relative systolodiastolic variations in diameter of the carotid artery in HIV-infected children were significantly lower than those in controls, but there was no significant difference in intima-media thickness. Cross-sectional compliance and distensibility were also significantly lower. Wall stiffness, assessed as the incremental elastic modulus, was larger in HIV-infected children. Endothelium-dependent dilation was lower in HIV-infected children but non-endothelium-dependent dilation was similar to that in controls. We did not find differences for any of the vascular variables between HIV-children receiving antiretroviral therapy and those never treated. All arterial variables were similar in children with and without dyslipidemia. No correlation was found between the duration of antiretroviral therapy with or without protease inhibitors and vascular dysfunction. Conversely, wall stiffness positively correlated with the length of time without antiretroviral therapy.

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Conclusions: Never treated HIV-infected children had a vascular dysfunction in the absence of cardiovascular risk factors. No additional detrimental effects were observed after a mean of 5 years of antiretroviral therapy.

O24-2

Incidence of shunt thrombosis/stenosis and/or effusions/seroma after an aorto-pulmonary shunt in patients with cyanotic congenital heart defects. Does Heparin make a difference?

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Objectives: Does an early postoperative dose of 5,000-10,000 units/m^2/d Heparin affect the incidence of aorto-pulmonary shunt obstruction/occlusion and of effusions/seroma in patients with cyanotic heart disease?
Patients and Methods: Between 1994 and 1999 98 patients with cyanotic congenital heart defects at a mean age of 34.5 months (range 1 day to 29.9 years) were treated with a total of 144 aorto-pulmonary shunts (BTA left 17, BTA right 19, central shunt 110) and four valveless RV-PA conduits. In a retrospective analysis we compared two groups of postoperative courses. Group I (56 consecutive courses) was treated without therapeutic heparin (only 100–150 units/day in the arterial line). Group II (89 consecutive courses) received Heparin 5,000–10,000 units/m^2/d until the central venous line was withdrawn, 3 pts. were not included due to incomplete data. All pts. were examined by echo.
Results: The incidence of effusions was significantly higher in group II (42% vs 25% respectively, $p = 0.03$). The incidence of isolated shunt seroma did not differ significantly (7% group I vs 10% group II respectively, n.s.). There was no difference in shunt obstruction/occlusion between the groups.
Conclusions: The incidence of postoperative effusions was significantly higher in pts. treated with 5,000–10,000 units/m^2/d after an AP-shunt operation compared to pts. treated without therapeutic doses of heparin. There was no difference in the incidence of shunt obstruction and the appearance of shunt seroma. Hence we suggest to avoid the routine administration of therapeutic doses of Heparin in the early postoperative couse after a shunt operation.

O24-3

Predictors of sudden death following the Mustard or Senning procedure

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Objectives: To identify predictors for sudden death (SD) in patients with transposition of the great arteries (TGA), having undergone atrial inflow repair.
Background: SD is the commonest cause of late death after atrial inflow repair of TGA. Little is known about the predictors of SD.
Methods: A retrospective multi-center cae controlled study was performed. We identified 47 patients after Mustard or Senning operation who experienced a SD event (34 SD, 13 near miss SD). Each patient was matched with 2 controls with the same operation, but without a SD event. Information on numerous variables prior to the event was obtained and compared with controls at the same time frame.
Results: Presence of symptoms of arrhythmia or heart failure at most recent follow-up and history of documented arrhythmia (supraventricular tachycardia (SVT) in particular) were found to increase the risk of SD. The resting ECG, chest X-ray, and Holter ECG findings were not predictive of SD. Neither medication nor pacing was found to be protective.
Conclusions: Presence of symptoms and documented SVT are the best predictors of SD in TGA patients following atrial repair.

O24-4

Early regional myocardial dysfunction in duchenne muscular dystrophy detected by ultrasonic strain and strain rate imaging

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Background: Most patients with Duchenne Muscular Dystrophy (DMD) develop dilated cardiomyopathy during adolescence. Early in life (below 12 years), global cardiac function is interpreted as normal using conventional echocardiographic techniques. The aim of the present study was to determine whether ultrasonic strain/strain rate imaging could detect early abnormalities in regional function in children with DMD before global cardiac dysfunction is recognized.
Methods: 22 DMD patients aged 7.4 ± 2.6 years and 29 age-matched normal controls were included. Standard echocardiographic measurements as well as Doppler myocardial imaging data were obtained. Doppler myocardial velocities, peak systolic strain rate and strain were estimated both in the radial and longitudinal directions from the LV lateral wall, interventricular septum and RV lateral wall.
Results: Conventional indices of left ventricular function were not different in the patient group compared to normal controls. Myocardial tissue velocities were significantly reduced in the LV lateral wall. A highly significant decrease in radial peak strain rate and peak strain was found in the infero-lateral wall (SR 3.0 ± 0.4-s vs. 4.3 ± 1.2-s, $p < 0.001$ strain 39 ± 12 vs. 62 ± 13, $p < 0.001$). Peak longitudinal and strain rate was significantly reduced in the three segments of the LV lateral wall. No significant changes in peak systolic myocardial velocities, strain and strain rate could be detected in the interventricular septum.
Conclusions: We conclude that strain rate and strain imaging can detect early regional changes in deformation parameters in young DMD patients. Especially the inferolateral wall is affected in the early stages of the disease. This could have interesting implications for the treatment of these patients.

O24-5

Magnetic resonance coronary angiography and late-enhancement myocardial imaging in children with arterial switch operation for transposition of the great arteries

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Objectives: To demonstrate the feasibility of MR coronary imaging and define myocardial damage in children who have undergone arterial switch for transposition of the great arteries.

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Background: In older subjects who have undergone the arterial switch operation, there remains concern about the development of late coronary artery complications. Furthermore, the degree of possible myocardial damage resulting from coronary surgery has not been defined in older asymptomatic subjects.
Methods: Sixteen asymptomatic subjects with arterial switch for transposition of the great arteries were studied (mean age 10.8 ± 1.3 years). MR coronary angiography, late-enhancement MR imaging, global ventricular function and regional wall motion were assessed. Fifteen children were awake during imaging; one was imaged under general anesthetic.
Results: Diagnostic images of the ostium and proximal coronary artery were acquired in 72% of coronary arteries imaged; this rose to 100% in subjects older than 11 years. No ostial stenoses were seen. In all subjects, the proximal course of the coronary arteries was visualized. Two subendocardial viability defects were detected, corresponding to known compromise of the artery supplying that territory. Global left and right ventricular function were preserved, with no regional wall abnormalities.
Conclusions: We have demonstrated that diagnostic MR coronary angiography can be performed in the majority of subjects who have undergone arterial switch for transposition of the great arteries. Furthermore, we have demonstrated that there were no unexpected areas of myocardial infarction, suggesting that patients surviving to this age did not have asymptomatic episodes of myocardial damage at the time of operation.

O24-6
Regional myocardial function in children with congenital aortic stenosis. A Doppler myocardial study

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Objective: Congenital aortic stenosis (AS) is a relatively common condition that causes left ventricle (LV) pressure overload leading to hypertrophy, diastolic dysfunction, exercise intolerance, heart failure and even sudden death. Subendocardial ischaemia is probably one of the first events occurring in the development of the disease. A sensitive indicator of dysfunction would be of value for optimising the best moment for intervention. Strain Rate (SR) (unit/s) and Strain (S) (%) imaging is a new technique that quantifies regional myocardial deformation. We aimed to determine whether or not S and SR imaging could detect regional myocardial dysfunction in patients with AS.
Methods: Regional and global myocardial function was assessed by conventional echocardiographic indices and using Myocardial Velocities (Vel), S and SR imaging in 16 patients, mean age: 12.8 ± 2.8 years, with moderate to severe AS. Data were acquired in the basal infero-lateral wall using a long axis view. Longitudinal S and SR were calculated for the basal, mid and apical segments of the septum and lateral wall from apical 4-chamber. The values were compared, by means of unpaired t tests, to 28 age-matched normal subjects.
Results: There was a significantly higher ejection fraction, wall thickness and LV mass in the AS group compared to normals, whereas LV end diastolic, end systolic diameters and septal ring motion were significantly lower. Peak systolic S was significantly lower in five out of six analysed LV segments. SR was diminished in the basal infero-lateral and the basal and mid segments of the septal wall. Especially remarkable is the significant reduction in longitudinal deformation in the interventricular septum.

	Peak systolic velocity (cm/s) Aortic Stenosis	Peak systolic velocity Normals	Peak systolic strain rate (s^{-1}) Aortic stenosis	Peak systolic strain rate Normals	Peak systolic strain (%) Aortic stenosis	Peak systolic strain Normals
Infero-lateral	3.8 ± 1.3	3.7 ± 0.7	3.2 ± 0.6*	3.7 ± 1.0	49.8 ± 13.5*	57.2 ± 11.8
Basal septal	4.8 ± 0.8	4.9 ± 0.8	−1.5 ± 0.4*	−1.9 ± 0.6	−17.6 ± 12.0*	−24.4 ± 6.1
Mid septal	3.4 ± 1.0	3.5 ± 0.8	−1.5 ± 0.4*	−1.9 ± 0.7	−19.5 ± 7.0*	−24.2 ± 7.4
Apico septal	2.1 ± 1.0	1.8 ± 0.9	−1.6 ± 0.8	−1.7 ± 0.3	−24.8 ± 7.9	−24.9 ± 5.2
Basal lateral	7.0 ± 3.1	7.3 ± 2.0	−1.8 ± 0.8	−2.2 ± 1.1	−14.9 ± 9.7#	−26.7 ± 11.6
Mid lateral	7.0 ± 3.1	6.5 ± 2.1	−1.6 ± 1.7	−2.1 ± 0.8	−17.4 ± 15.3*	−26.4 ± 8.6
Apico lateral	6.5 ± 3.7	5.0 ± 2.4	−1.8 ± 0.7	−2.1 ± 1.0	−19.5 ± 9.3*	−24.6 ± 7.7

Values are expressed as mean ± SD; *$p < 0.05$ vs. normals; #$p < 0.01$ vs. normals.

Conclusions: Regional differences in myocardial deformation are observed in patients with AS. Especially longitudinal shortening in the septal wall seems to be especially affected. This could be related to subendocardial damage, which causes reduced deformation of the longitudinally oriented subendocardial fibers.

Session 29: Interventional Cardiology IV

O29-1
Is balloon stability important in balloon aortic valvuloplasty? Rapid ventricular pacing, a safe and effective option

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Background: Infants and children with congenital aortic stenosis have been treated with surgery or via transcatheter route. Surgery is advantageous as the valve is dealt with under direct vision and there is less likelihood of incompetence. With the development of catheter techniques, balloon aortic valvuloplasty has offered an equally encouraging alternative. A problem with balloon valvuloplasty is balloon movement resulting in sub-optimal result with possible aortic incompetence. There are several techniques to overcome this problem but none proven satisfactory. An alternative method to achieve balloon stability has been pursued namely rapid ventricular pacing, which drops the cardiac output during the time of balloon inflation.
Methods: From May 2003 till November 2003, 6 patients with significant aortic valve stenosis were treated by this strategy. Age of the patients ranged from 1 month to 17 years with 5 male and 1 female. All patients had a bipolar pacing catheter placed in the right ventricle. Invasive systemic pressures were documented with a catheter in the descending aorta. Rapid ventricular pacing was initiated at the rate of 200 per minute and increased to a rate required to achieve a drop in systemic pressure by 50%. Balloon was inflated only after the pacing rate was reached and the blood pressure dropped. Pacing was continued until the balloon was deflated. This procedure did not show any to and fro motion of balloon during inflation thus achieving optimal valvuloplasty.
Results: The systolic gradient across the aortic valve before balloon dilatation ranged from 48–100 mmHg (mean systolic gradient 57 mmHg). The systolic pressure in aorta ranged from 51–82 mmHg (mean systolic pressure 71 mmHg). The pacing rate to drop the pressure by 50% ranged from 200 to 240 per minute. Balloon stability at time of inflation was achieved in all 6 patients. The gradient post ballooning of the aortic valve ranged from 14–80 mmHg (mean systolic gradient of 33 mmHg). Angiogram performed post balloon dilatation showed trivial

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aortic incompetence in 1 patient and none in 5 patients. There were no untoward complications during the procedure or following the procedure.
Conclusion: Rapid ventricular pacing to stabilise the balloon during catheter valvuloplasty appears safe and effective and may decrease the incidence of aortic incompetence.

O29-2
Late longitudinal stent fracture as a cause for re-stenosis

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Stent implantation has been shown to be a safe and effective treatment for native and postoperative vascular obstructions based on short and medium term follow-up. Systematic long term follow-up is still limited. Investigation of the occasional cases of late re-stenosis has revealed longitudinal stent fracture as one mechanism. Therefore, we reviewed our late follow-up after intravascular stent placement to characterize the role of late stent fracture.

Seven cases of complete longitudinal stent fracture were identified from a population of 69 stent implantations. All stent fractures were associated with significant re-stenosis. All patients had undergone previous surgery mostly for Tetralogy of Fallot. Patient age at implantation ranged from 3.0 to 24.8 years (median 11.1 years), weight at implantation was 11.8–94 kg (median 31 kg). The implanted stents were Palmaz P128 (1), P188 (5) and P308 (1) expanded with 12–18 mm balloons (median 15 mm). The stents were located in the right pulmonary artery (3), the left pulmonary artery (2), the proximal RV–PA conduit (1) and in the transverse aortic arch (1). The stenosis diameter increased from 5.5–12 mm to 11–17 mm. The median interval to the documentation of longitudinal stent fracture was 54 months ranging from 48 to 68 months except for the stent in the transverse aortic arch (10 months). At this time the re-stenosis measured from 3.4 to 11.1 mm (median 7 mm) and was successfully treated with stent-in-stent implantation.

Late longitudinal stent fractures appear to be partly related to pulsatile external compression and are associated with re-stenosis. This observation has implications for the concept of resorbable stents and warrants continued long term follow-up after stent implantation.

O29-3
Pulmonary balloon angioplasty after cavopulmonary shunt – is it effective?

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Aim: To assess the efficacy of pulmonary balloon angioplasty (PBA) in the treatment of stenosis of the left pulmonary artery (LPA)after bidirectional cavopulmonary shunt (CPS).
Design: Retrospective analysis of serial angiographic studies and procedural data. Single tertiary paediatric cardiology centre.
Methods: Of 231 survivors after CPS, 23 patients were identified having LPA stenosis requiring treatment (10%). Nine patients were excluded from analysis (bilateral SVC in 2, primary stent in 2, completion of Fontan shortly after PBA in 3, and incomplete data in 2). The outcome of 21 PBA procedures in 14 patients with follow up angiography was studied in detail.
Results: Percentage LPA stenosis [(1 − LPA min/LPA max) × 100%] reduced from 41 ± 15% to 21 ± 14% ($p = 0.0001$) but was not altered at follow up (42 ± 30%, $p = 0.8$). Two patients (9.1%) had late complete occlusion of the LPA presumed due to dissection and thrombosis. The degree of LPA hypoplasia [(1 − LPA max/RPA max) × 100%] did not change either immediately (27 ± 15% vs 24 ± 16%, $p = 0.6$) or late after PBA (27 ± 24%, $p = 0.9$). Three patients required further patch augmentation of the LPA at completion of Fontan, and three underwent stent implantation prior to Fontan.
Conclusions: LPA stenosis and hypoplasia is a common finding after CPS. Pulmonary balloon angioplasty can acutely improve the stenosis but does not provide long term relief or change in the degree of LPA hypoplasia.

O29-4
Extending the limits of Amplatzer Septal Occluder: Closure of atrial septal defects beyond 40 mm in diameter

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Objectives: The Amplatzer Septal Occluder (ASO) device can close atrial septal defects (ASD) with stretched diameter of 8 mm. However, it has not been described whether the device is suitable for ASD that are ⩾40 mm in diameter. Since the left disc is 16 mm larger than the waist in 38 and 40 mm devices, it may be possible to close ASD that are ⩾40 mm, if the device can be aligned to the septum optimally. The objective of this study was to evaluate efficacy of the ASO in defects >40 mm in diameter.
Methods: From January 2000 to December 2003, 14 patients with ASD stretched diameter ⩾40 mm (n = 10) and non-stretch diameter ⩾40 mm (n = 4) underwent catheterization for device closure, with 38 mm (n = 12) and 40 mm (n = 1) device. Age ranged from 25 to 76 years. Only one patient had CVA. The defects were closed under transesophageal echocardiography (n = 8) and intracardiac echocardiography (n = 6). Proper alignment of the left disc was obtained by keeping the device facing posteriorly by partially deploying the device in the left pulmonary vein and/or clockwise rotation of the sheath.
Results: Device placement was attempted in 13 patients. In one patient with dextrocardia and partial anomalous pulmonary venous return, device placement was not attempted. The procedure was unsuccessful in one patient with non-stretched diameter of 46 mm. The device was successfully deployed, but was removed because of assumed instability of the device. Placement was successful in the remaining 12 patients. There was no incidence of device embolization. There was no impingement of the atrio-ventricular valves in any patient. All defects were completely closed at 6 months follow up. Follow up was available for up to one year (n = 12).
Conclusions: The large ASD usually have no anterior or retro-aortic rim and may also have a small posterior rim. Alignment of the left disc parallel to the atrial septum results in excellent position of the device after deployment, and defects that have a stretched or non-stretched diameter larger than 40 mm can be closed. The 16 mm larger diameter of the left atrial disc than the self-centering waist helps stabilize the device. We recommend that all patients with secundum ASD size should undergo cardiac catheterization and attempt to close the ASD if the non-stretched diameter of the defect is ⩽44 mm.

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O29-5

Stent – implantation in patients with aortic coarctation (CoA)

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Background: Today there is increasing evidence, that stent implantation for residual aortic coarctation may be a good alternative to surgery. We report our experience with this method.
Material and Methods: Since February 1999 30 patients (f = 9, m = 21) with coarctation (re-CoA = 28, native CoA = 2) at a median age of 15.7 years (range 9.9–39.4 y) were treated with a stent. Eighteen patients had isolated CoA, 2 pts an additional VSD and 10 pts complex associated cardiac defects. Initial surgery (n = 28) for CoA (extended end to end anastomosis 21, Waldhausen operation 2, conduit 5) was performed at a median age of 2 months (range 3 days–19 y).
Results: A total of 33 stents were deployed (Plamaz 18, CP 8, Intrastent 7). The median invasive gradient was reduced from 22 mmHg (range 7–44 mmHg) to 1.4 mmHg (range −16–10 mmHg) ($p < 0.0001$). The stenotic aortic segment was widened from median 8 mm (range 6–12.9 mm) to 12 mm (range 9–16.7 mm) ($p < 0.0001$). One stent embolized into the abdominal aorta, was left there without negative consequences. In two cases, the balloon ruptured during inflation. Rapid manual balloon inflation enabled safe stent deployment. All patients shall be reevaluated in a structured out-patient visit.
Conclusions: Stent implantation in patients with Re-CoA is safe and effectively eliminates the stenosis and may evolve to the method of choice for treatment of this lesion. Nevertheless arterial hypertension persists in some patients and medical treatment is necessary in these cases.

O29-6

Non self-centering devices for transcatheter closure of multi-fenestrated atrial septal defects: Cribriform Amplatzer Septal Occluder (CSO) versus Helex Septal Occluder (HSO)

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Background: A large number of devices have been investigated for percutaneous ASD closure. Cribriform Amplatzer Septal Occluder (CSO)™ and Helex Septal Occluder (HSO)™ are relatively new and are used for closing multi-fenestrated defects. We evaluated our experience with these 2 devices.
Results: Twelve patients with multi-fenestrated ASDs had percutaneous catheter closure using HSO (n = 5) or CSO (n = 7) devices in our institution. Transoesophageal echocardiogram and fluoroscopy were used for accurate device deployment and assessing residual shunts. Two patients in the HSO group and 4 patients in the CSO group had an aneurysmal atrial septum.

The size of devices deployed ranged from 25 mm to 35 mm in both the groups. Qp:Qs ratio ranged from 1.13 to 2.8 (median = 1.2) in the HSO group and 1 to 1.92 (median = 1.14) in the CSO group, with normal pulmonary artery pressures in both groups. The median for procedure duration was 90 minutes (range 73–121) in the HSO group and 95 minutes (range 16–155) in the CSO group. The shortest procedure duration in the CSO group was 16 minutes vs.73 minutes in the HSO group. Device deployment was satisfactory in all the patients. In 2 with CSO "buttercup" deformity of the left atrial disc were noted with no haemodynamic effects. Aneurysmal atrial septa were well splinted between the left and right atrial discs for both device types. Three of 7 with CSO had tiny residual leaks and two of 5 patients with HSO had a tiny residual leak.
Conclusions: CSO and HSO devices are well suited for closure of multi-fenestrated atrial septal defects and both these are particularly useful in stabilizing the atrial septum in the presence of septal aneurysms. Although ease of deployment, shorter procedure times and device profile favours the CSO device, deformation of the left atrial disc in some remains of some concern particularly in those patients who referred for prior thromboembolic events.

O29-7

Reinterventions after balloon dilatation for valvular aortic stenosis in neonates and infants

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Objectives: Good early and midterm results after balloon dilatation (BD) for valvular aortic stenosis (VAS) in neonates and infants in the reintervention free group are well studied, but data about the type of reinterventions and the typical complications are sparse. A review of our single center experience is presented.
Material and Methods: Between 1993 and 2003, 27 neonates and 45 infants underwent BD for VAS. Besides mortality and reintervention rate, the type, indication and result of reinterventions were studied retrospectively. Efficacy and complication of BD was definied as follows: Ineffective BD: Doppler peak instantaneous systolic gradient (DG) >75 mmHg within 3 months after BD. Late restenosis: DG returned to >75 mmHg after 1 year. Early AI: AI gr III–IV° develops within 3 months after BD. Late AI progression (progr.AI): AI gr III–IV° develops more than 1 year after BD.
Results: Early mortality was 11/72 (15%), one of them was catheter related. Late mortality was 3/72 (4%). In survivors who have not required reintervention 42/72 (58%), 1,9 ± 1,7 years after BD: echocardiography 4,0 ± 2,6 years after BD revealed a DG of 41 ± 18 mmHg and moderate aortic regurgitation in 5 patients. 16/72 pts (22%) required reinterventions: repeat BD in 9 pts, surgical valvulotomy in 2 pts, Ross operation in 2 pts, Ross-Konno operation in 1 pt, surgical valvuloplasty in 1 pt, prosthetic mitral valve implantation in 1 pt. BD was ineffective in 3/72 pts (4%). ReBD because of late restenosis was effective in 2/72 pts (3%), and ineffective in 3/72 pts (4%). Early AI developed in 2/72 pts (3%), progr.AI occured in 2/72 pts (3%). In 2/72 pts the BD was repeated in 2 weeks with the same ballon size because of severe residual stenosis and they are free from reintervention 1 and 3 years respectively after reBD. 1 pt underwent prosthetic mitral valve implantation because of severe mitral insufficiency without reintervention on aortic valve.
Conclusions: 1. Reintervention rate after BD for VAS in neonates and infants is not negligible. 2. Ineffectivity, restenosis, early AI and AI progression occurs proportionally among indications for reintervention.

O29-8

Non-surgical management of babies with pulmonary atresia intact ventricular septum medium term follow-up

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Between 1996–2002, 39 patients with pulmonary atresia intact septum underwent laser/radiofrequency ablation of the pulmonary valve. Age range between 7 days–11 years, weight 2.7 kg–33.4 kg,

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there were 22 females and 17 males. Out of 24 patients underwent laser perforation, there were 22 successful procedures and there were 9 successful radiofrequency procedures. All babies continued on Prostaglandin infusion for a mean of 10 days. Three patients required surgical procedures in form of right ventricular outflow patch. Three had surgical valvotomies and resection of subpulmonary valve stenosis. Two patients had 1½ repair (Glenn Procedures). There was 1 surgically related death.
Conclusions: We conclude that the non-surgical approach for pulmonary atresia and intact ventricular septum is safe and effective and should be considered as first line treatment.

POSTER PRESENTATIONS

Session 1: Heart Failure

P1

Recovery kinetics of oxygen uptake is prolonged in adults with an atrial septal defect and improves after transcatheter closure

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Background: In adults with an atrial septal defect (ASD) transcatheter closure leads to an improvement of peak oxygen uptake (VO_2), but the kinetics of recovery of VO_2 after maximal exercise in this patient population, and the impact of transcatheter ASD have never been investigated.
Methods: 20 consecutive patients underwent a maximal cardiopulmonary exercise test both the day before and 6 months after transcatheter ASD closure. For comparison, an age and sex matched group consisting of 53 healthy adults was built. The constant decay of VO_2, CO_2 production (VCO_2), minute ventilation (VE), and heart rate (HR), expressed as the first degree slope of a single linear relation were calculated for the first minute of recovery.
Results: ASD patients had a prolonged VO_2-slope ($p = 0.0012$), VCO_2-slope ($p = 0.0003$) and VE-slope (0.013) when compared to control subjects. After 6 months from transcatheter ASD closure significant improvements of VO_2-slope ($p = 0.0043$) and of VCO_2-slope ($p = 0.0022$) were recorded, so that no difference was found when compared to the control group ($p = 0.1$ and $p = 0.06$, respectively). VE-slope and HR-slope did not change after closure. Spearman correlation showed a significant association between VO_2-slope and peak VO_2 in the group of ASD patients, both before ($r = 0.67, p = 0.0012$) and after ASD closure ($r = 0.71, p = 0.0004$).
Conclusions: A limited cardiopulmonary reserve in asymptomatic adults with an ASD appears to affect not only maximal exercise responses but also the recovery phase. Transcatheter ASD closure induces a significant improvement of the ability of recovering from maximal exercise and eliminates the difference with respect to a normal population.

P2

Mechanical cardiac support in children – ECMO or ventricular assist device?

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Comparative evaluation of clinical experience with ECMO (centrifugal pump, Biomedicus) and the Medos-VAD (displacement pump, pulsatile flow).
Patients: ECMO: n = 18, age: 12d–21yrs (median 6.5 months); postcardiotomy 9, primary organ failure after HTX 5, prophylactic ECMO after HTX in children with elevated PVR 3, rescue-ECMO in cardiac shock 1. VAD: n = 11, age: 20d–11yrs (median 23 months); postcardiotomy 3, rescue/under CPR 3, elective for heart failure/myocarditis 5; BiVAD 4, LVAD7.
Results: ECMO: Duration 2-21d (median 5.5 days); complications: bleeding 16, sepsis 1, thrombembolic 2, myocardial infarction 1; successfull weaning 15/18 (1HTX), late death 4/15. VAD: 1–30d (median 9 days); bleeding 8, sepsis 3, cerebral infarction 3; survival 5/11: 2 recovery after myocarditis, HTX 3; 6 died: ARDS/MOF 5, cerebral infarction 1.
Discussion: ECMO compensates for pulmonary hypertension, intracardiac shunts and pulmonary failure after cardiotomy. Coronary perfusion and myocardial recovery may be impaired, when ECMO has to replace cardiac function completely. Organ perfusion with pulsatile flow is more effective in low flow states. Rate of complications are less under VAD compared to ECMO during prolonged application (>10d); VAD allows patient mobilization and neurologic evaluation (HTX candidates).
Conclusions: ECMO is preferred in the postcardiotomy setting and in the rescue application. If myocardial recovery does not take place within 1 week a switch to VAD has to be taken into account. VAD is suitable to provide assist during an unpredictable long waiting period.

P3

Acute heart failure in pediatric patients treated with mechanical circulatory support (MCS) – experience with the first five patients in Innsbruck

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Objective: Between June 2000 and September 2003 five pediatric patients with acute heart failure where treated with MCS. In all patients standard intensive therapy failed.
Patients: Patients were aged 2 month to 17 years, three male and two female. Diagnosis was dilated cardiomyopathy (DCM) with suspected myocarditis in three, end stage chronic DCM in one and Long QT-Syndrome with cardiac arrest in one patient. Extracorporal membrane oxygenation (ECMO) was used as rescue therapy in three patients. Two patients were stable enought for semielective thoracotomy and implantation of an pulsatile ventricular assist device (VAD).
Results: Two of 3 ECMO-Patients were switched to VAD for long-term-support after 4 and 5 days. The third patient was weaned from ECMO after 6 days. VAD use ranged from 8 to 168 days. Two patients were weaned from VAD after 47 and 79 days of support. The remaining 2 patients received orthotopic heart transplantation after 8 and 168 days. All Patients were discharged from hospital. Follow up range is 4 month to 3 1/12 years. All patients are free of symproms (NYHA I).
Conclusion: Machanical circulatory is a cost intensive but life saving therapy for acute heart failure if other therapy fails and is available for pediatric patients. ECMO can be established as rescue therapy within a short period. For long-term-support results of VAD with pulsatile blood flow are better. VAD can be used in children for a long period (up to 168 days in our institution).

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P4

Quantitative assessment of homodynamic status of ventricular septal defect in infants: natriuretic peptides study

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Background: In the clinical management of patients with ventricular septal defect (VSD), quantitative assessment of the severity of the VSD is of major importance. Atrial natriuretic peptide (ANP) secreted by the atrium, and brain natriuretic peptide (BNP) secreted mainly by the ventricle, are circulating peptides that respond to intracardiac hemodynamic status. We investigated that plasma ANP and BNP levels could reflect hemodynamic status in patients with VSD.
Methods: Twenty-two infants with VSD, median age was 3 months (ranged from 1 to 46 months), were studied. Diagnosis included perimembranous VSDs in 14 patients, doubly committed subarterial VSDs in 4 patients, inlet VSDs in 2 patients, and muscular VSDs in 2 patients. All patients were performed cardiac catheterization study to evaluate the hemodynamic severity of VSD. Plasma ANP and BNP levels were measured simultaneously at cardiac catheterization. All patients with VSD showed various degree of congestive heart failure.
Results: Pulmonary artery systolic pressure (PAP) was 44.2 ± 22.1 mmHg (ranged from 14 mmHg to 94 mmHg), mean pulmonary artery pressure (mPAP) was 27.9 ± 16.1 mmHg (ranged from 11 mmHg to 73 mmHg), the ratio of pulmonary blood flow to systemic blood flow (Qp/Qs) was 2.5 ± 1.1 (ranged from 1.0 to 4.5). Plasma ANP levels showed significant positive correlation with severity of VSD, PAP ($r = 0.65, p < 0.05$), mPAP ($r = 0.55$, $p < 0.05$) and Qp/Qs ($r = 0.65$, $p < 0.01$). Also, plasma BNP levels showed significant positive correlation with hemodynamic status of VSD, PAP ($r = 0.59$, $p < 0.05$), mPAP ($r = 0.48$, $p < 0.05$), and Qp/Qs ($r = 0.51$, $p < 0.01$). Eight patients, including 4 patients with doubly committed VSDs and 2 patients with inlet VSDs and 2 patient with perimembranous VSD, underwent cardiac surgery. After surgery, ANP and BNP levels decreased within normal value associated with improvement of hemodynamics status.
Conclusions: Plasma ANP and BNP levels correlated with hemodynamic status in VSD. This result suggests that the monitoring in ANP and BNP levels may aid in noninvasive quantifying the severity of VSD and may be useful to observe during follow-up.

P5

Increased BNP/ANP ratio in children with systolic ventricular dysfunction

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The clinical use of the natriuretic peptides type B (BNP) and A (ANP) as markers of ventricular dysfunction has not yet been fully established. Aim: To study the relationship between P-BNP and P-ANP in children with systolic ventricular dysfunction and different types of hemodynamic overload of the heart. Methods: Samples of P-BNP and P-ANP were taken during medical check-ups, pre-operative investigations or interventions (surgery/catheter) in children with congenital heart defects or cardiomyopathy. The hemodynamic load was evaluated by echo-doppler investigation (echo) and/or during catheterisation. Hemodynamic overload of the heart was classified as: Pressure overload of the left (Pres LV) (AS, CoA) or right (Pres RV) (PS) ventricle, Volume overload of the left (Vol LV) (VSD, PDA) or right (Vol RV) (ASD) ventricle, sufficient to indicate surgery/catheter intervention according to local praxis. Systolic ventriular dysfunction (Dysf) was defined as a fractional shortening (FS) of <26% on echo. 23 children without heart disease aged 2 weeks – 8.3 years served as a control group for the natriuretic peptide measurements. (Reference intervals for BNP: 0–18.4 ng/L and ANP: 0– 43 ng/L.) Results: 76 patients were included (46 boys, 30 girls), mean age 4.5 years (3 months–16.2 years). The BNP/ANP ratio was significantly higher in the Dysf group mean 1.6 (0.6–3.4) ($n = 15$; 9 DCM, 6 congenital heart defects) as compared to the group of children with Pres LV 0.3 (0.6–0.8) ($n = 15$), Pres RV 0.3 (0.1–0.8) ($n = 11$), Vol LV 0.6 (0.1–2.3) ($n = 16$), Vol RV 0.4 (0–2.0) ($n = 19$) and controls 0.2 (0–0.6) ($p < 0.0001$). Of the remaining groups, Vol LV was the only with significantly higher BNP/ANP ratio than the controls ($p = 0.04$). FS was significantly decreased in the Dysf group mean 14% (9–24) but within the normal range in the remaining groups ($p < 0.0001$). The BNP/ANP ratio showed an inverse correlation with the FS ($p < 0.0001$). Conclusion: The BNP/ANP ratio is less than 0.5 in healthy children. During increased hemodynamic load of the heart the BNP/ANP ratio seems to be preserved as long as the ventricles are able to compensate for the increased strain. As the cardiac overload overwhelms the compensatory capacity of the heart the BNP/ANP ratio increases and a value above one may indicate systolic ventricular dysfunction.

P6

Plasmatic natriuretic peptides in neonates and children with congenital heart diseases: correlation with ventricular function, heart failure symptoms and cardiac anatomy

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Objectives: To determine plasma levels of atrial natriuretic peptide (ANP) and brain natriuretic peptide (BNP) in control children and in children with congenital heart disease (CHD). In addition, to correlate ANP and BNP levels with ventricular function, heart failure symptoms and cardiac anatomy.
Methods: ANP and BNP were determined (a) in 163 children (86 male; median age 188 days, range 2 days–6 years) with CHD admitted to our Unit between December 2002–August 2003 and (b) in 35 healthy controls matched for age and sex. Left and right ventricular function were measured by a blinded echocardiographer as ejection fraction (EF) according respectively to Simpson and prolate ellipsoid model. Symptoms of heart failure were ranked using Ross score (RS) (Ross. Pediatric Cardiol 1992; 13: 72). A higher score (range 0–12) corresponds to more severe symptoms. Cardiac anatomy was distincted in 5 anatomic subgroups: cardiomyopathies (A) (22 pts), pressure left ventricular overload (B) (33 pts), volume left ventricular overload (C) (53 pts), pressure right ventricular overload (D) (33 pts), volume right ventricular overload (E) (22 pt). ANP and BNP concentrations were determined by immunoradiometric assays.
Results: ANP and BNP levels in pts with CHD were significatively higher than in controls (55.7 ± 32 v 19.0 ± 6 pg/ml ($p < 0.05$) and 167.7 ± 220 v 3.8 ± 2 pg/ml ($p < 0.001$)). ANP and BNP in pts were negatively correlate with EF ($r = -0.36$, $p < 0.001$ and

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$r = -0.52, p < 0.001$) and positively correlated with RS ($r = 0.52$, $p < 0.001$ and $r = 0.56$, $p < 0.001$). ANOVA analysis shows no direct correlation between the 5 anatomic subgroups and natriuretic peptides: ANP pg/ml: (A): 68 ± 32; (B): 51 ± 35; (C): 67 ± 31; (D): 47 ± 28; (E): 41 ± 20. BNP pg/ml: (A): 277 ± 417; (B): 302 ± 625; (C): 105 ± 233; (D): 150 ± 70; (E): 52 ± 23. *Conclusions:* ANP and BNP are elevated in a large population of neonates and children with CHD. ANP and BNP levels reflect the severity of symptoms of heart failure and the impairment of ventricular function and not necessarily the anatomy substrate. ANP and BNP may be used as a helpful adjunct to monitor clinical and functional status in such pts.

P7

Perioperative plasma cardiac troponin I (TnI) and N-terminal pro-brain natriuretic peptide (N-proBNP) profile in children with rheumatic valvular disease and congestive heart failure (CHF) undergoing surgery

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Objectives: Both TnI and more recently N-proBNP were identified to monitor myocardial damage and severity of heart failure in adults and children. However, in neither cases, the relation to functional recovery was evaluated.The aim of this study is to determine plasma profile of TnI and N-proBNP in children CHF secondary to rheumatic valvular disease undergoing surgery, and to analyse relations to echocardiographic measurements.
Methods: Prospective study including twenty consecutive children (mean age of 11.7 ± 2.4 years, 12 boys) Measures were taken before and after (6, 12, 24 hours, 2–4 weeks) valvular surgery requiring cardiopulmonary bypass. Measures were expressed as mean ± SD; Repeated measures were compared using the Friedman's test and correlation determined pre-op, 24 hours and 2–4 weeks post-op by Spearman rank correlation coefficient. A p value < 0.05 was considered as significant.
Results:

	Before surgery	After surgery			
		6 hours	12 hours	24 hours	2–4 weeks
TnI [ng/ml]#	0.023 ± 0.019*	6.218 ± 3.979	5.320 ± 3.597	4.172 ± 2.273	0.038 ± 0.023**
N-proBNP [ng/l]#	3166 ± 6192***	3759 ± 7423	4361 ± 7566	4887 ± 9797	1792 ± 4290****

#normal values: TnI < 0.04 ng/ml; N-proBNP: 150–450 ng/l; *p < 0.05: pre-op versus 6, 12, 24 hours, 2–4 weeks post-op; **p < 0.01: 2–4 weeks versus 6, 12, 24 hours post-op; ***p = NS: pre-op versus 6, 12, 24 hours, 2–4 weeks post-op; ****p < 0.01: 2–4 weeks versus 6, 12, 24 hours post-op.

	TnI	N-proBNP
Shortening fraction	$-0.55, p < 0.001$	$-0.36, p = NS$
Left ventricular mass index	$0.03, p = NS$	$0.38, p = 0.04$
End-systolic wall stress (WS)	$0.07, p = NS$	$0.44, p = 0.03$
Left atrium to Aorta diameter ratio	$-0.09, p = NS$	$0.28, p = NS$

Conclusions:

- TnI was increased immediately after repair undersigning acute myocardial necrosis and progressively returned to normal values after 2–4 weeks.
- N-proBNP was markedly increased compared to normal values. It peaked at 24 hours after repair suggesting ongoing adaptation to modified ventricular loading condition as demonstrated by the correlation with end-systolic wall stress.
- N-proBNP progressively decreased to levels lower than preoperatively at 2–4 weeks and may be useful to monitor functional recovery.

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P8

Arrhythmogenic right ventricular dysplasia presenting with severe right heart failure findings including abdominal ascites in a child

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Arrhythmogenic right ventricular dysplasia is characterized by the progressive replacement of the myocardium by fatty or fibrofatty tissue leading to cardiac electrical instability and progressive ventricular dysfunction. While the presenting symptom is commonly related to ventricular arrhythmias, severe right ventricular failure findings are unusual during childhood period. This report describes a 12-year-old girl with arrhythmogenic right ventricular dysplasia, presenting with only severe right ventricular failure symptoms including fatigue, abdominal distension resulted from hepatomegaly and abdominal ascitis as an unusual presenting symptom.

The case has multiple criteria which satisfy the diagnosis of ARVD. Major criteria fulfilled by the case include the fibrofatty infiltration of the RV myocardium with either atrophic or hypertrophic remaining myocytes surrounded by fibrous tissue, and severe dilatation and reduction of right ventricle ejection fraction demonstrated by echocardiographic evaluation and cardiac catheterization. Minor criteria fulfilled by the case include non-sustained LBBB-type ventricular tachycardia and frequent ventricular extrasystoles on Holter monitoring.

This case demonstrates an unusual presentation of ARVD with severe high heart failure leading to abdominal ascites, which can be the presenting symptom even in a child at a school age period. Furthermore, to the best of our knowledge, abdominal distension secondary to peritoneal ascites was not reported before as the presenting symptom in children. Therefore, the diagnosis of ARVD should be considered in school age children with peritoneal ascites secondary to severe right heart failure, even they don't have any complaints associated with ventricular tachycardia such as palpitation or syncope episodes.

P9

Are elevated levels of N terminal pro-brain natriuretic peptide in children a valid marker for heart failure in the perioperative period?

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Objectives: To investigate whether N terminal pro-brain natriuretic peptide (NTpro-BNP) is elevated in children with heart failure due to congenital heart disease preoperatively and after corrective or palliative surgery.

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Methods: In 18 children (median age 70 days; range 6–507 days) who underwent surgery for congenital heart disease plasma NTpro-BNP levels were measured pre- and/or 6 h and 12–48 h postoperatively using an automated enzyme immunoassay (Roche).
Results: Plasma NTpro-BNP levels were highly elevated in children with heart failure due to congenital heart disease preoperatively (median 2082 pg/ml, range 203–55697 pg/ml) and 6 h and 12–48 h postoperatively (median 17016/12935 pg/ml). The preoperative values in children within the first 3 months of life showed a wider distribution (median 6156; range 1897–55697 pg/ml) and significantly higher median NTpro-BNP plasma levels compared to children over 3 months of age (median 813, range 203–14124 pg/ml). The preoperative plasma levels correlate with the age at time of blood sampling ($r = 0.75$, $p = 0.001$) with the highest preoperative values in children within the first month of life (median 10134; range 5416–55697 pg/ml). Six hours postoperatively there was an increase in plasma NTpro-BNP levels compared to preoperative values followed by a decrease 12–48 h postoperatively. No correlation between NTpro-BNP plasma levels at 6 h and 12–48 h postoperatively and age could be observed ($r = 0.55$, $p = 0.483$/$r = 0.17$, $p = 0.807$).
Conclusions: The findings of this study suggest that plasma levels of NTpro-BNP are highly elevated in children in the pre- and postoperative period. In the preoperative period we observed a correlation between age and NTpro-BNP plasma levels – as described by Rauh and Koch for healthy neonates. Postoperatively we did not find an age dependency of plasma NTpro-BNP. Considering the physiological decrease within the first months of life plasma NTpro-BNP seems to be an applicable marker in children with ventricular dysfunction during the perioperative period. Further investigations are needed to elucidate a possible correlation of NTpro-BNP and age, ventricular morphology or volume load.

P10
Comparison of the New York Heart Association (NYHA), Ross, and Pediatric Heart Failure Index (PHFI) classifications in children with left ventricular dysfunction secondary to valvular disease. Biochemical and echocardiographic correlations

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Objectives: Classification of congestive heart failure (CHF) severity in children is not well established. Beside the NYHA classification that is used in adults, both the Ross classification and the PHFI were developed in children but never compared using correlation with echocardiographic measurements and biochemical markers of heart failure such TnI and N-proBNP.
Methods: 20 children (mean age: 11.7 ± 2.4 years, 12 males) were prospectively included and their pre- and 2–4 weeks post-operative characteristics studied. Correlations were determined by Spearman rank correlation coefficient.
Results: No correlations could be established between the three scoring systems and echocardiographic measurements (left ventricular shortening fraction, left ventricular mass index, left atrium to aorta diameter ratio and end-systolic wall stress).

	TnI	N-proBNP
NYHA	−0.20, p = NS	0.27, p = 0.025
Ross classification	0.08, p = NS	0.33, p = 0.001
Pediatric heart failure index	−0.13, p = NS	0.57, p < 0.001

Conclusion:

- In this study, no correlations between standard echocardiographic measurements and CHF scoring systems could be established.
- N-proBNP but not TnI correlates with clinical severity and may be a suitable way to biochemically monitor heart failure in children.
- Out of the three scoring systems, PHFI had the best correlation with N-proBNP and may be a suitable way to clinically score CHF in children.

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P11
N-terminal B-type natriuretic peptide and B-type natriuretic peptide levels in children with history of dilated cardiomyopathy or myocarditis

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Background: B-type natriuretic peptide (BNP) and the N-terminal segment of its pro-hormone (N-BNP) are markers for cardiac dysfunction. The relative significance of BNP and N-BNP in children with dilated cardiomyopathy (DCM) has not been elucidated. Moreover, it is unclear whether echocardiographic resolution of cardiac size and function is accompanied by normal levels of these peptides, which may suggest complete recovery.
Objectives: To correlate echocardiographic status and natriuretic peptide levels in pediatric patients with history of myocarditis or DCM.
Methods: Cardiac evaluation was performed and the levels of BNP (Beyer, USA) and N-BNP (Roche, Germany) were measured in 23 children who have a history of myocarditis or DCM (no myocardial biopsy performed). The mean follow-up interval was 4.8 y (3 m–10 y).
Results: Nine patients (5 males) had evidence of LV dysfunction and/or dilatation at the time of the study (DCM group), while 14 (9 males) had none (recovery). Data at presentation is shown in Table 1.

Table 1. Data at presentation.

Groups	Age (m)	LVEDD%	FS (%)
DCM	41 ± 69	120 ± 30	18.0 ± 5.6
Recovery	17 ± 33	120 ± 40	19.3 ± 6.4

Data at the time of the study is shown in Table 2.

Table 2. Data at the time of the study.

Groups	Age (m)	LVEDD%	FS (%)	BNP (pg/ml)	N-BNP (pg/ml)
DCM	76 ± 79	134 ± 31	20.7 ± 8.8⋆	138 ± 131⋆	3154 ± 2858
Recovery	90 ± 40	95 ± 9	37.9 ± 3.8	8.2 ± 5.6	122 ± 75

Values are mean ± SD; LVEDD% = LV end diastolic diameter as % of 90th% tile of normal. FS = fraction of shortening. ⋆ = $p < 0.05$ vs recovery group. There was excellent correlation between BNP and N-BNP levels ($R^2 = 0.89$, $p < 0.0001$). The areas under the ROC curves for the diagnosis of persistent DCM were: N-BNP = 0.984, BNP = 0.908, FS = 0.976. N-BNP levels of children who recovered (122 ± 75 pg/ml) were not different ($p = 0.45$) from the levels of 56 age-matched controls (113 ± 96 pg/ml), and none of the recovered patients had N-BNP level higher than the upper limit of normal.

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Conclusions: Both BNP and N-BNP are good markers for persistent DCM in children who had myocarditis or cardiomyopathy. N-BNP levels are normal in children who recover echocardiographically, suggesting no residual cardiac disease.

Session 2: Pulmonary Hypertension

P12

First experience with the Amplatzer fenestrated atrial septal device in the treatment of children with pulmonary hypertension in the United Kingdom

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Background: Primary Pulmonary Hypertension (PPH) is a rare but debilitating disease. Despite newer medical treatments, patients may continue to deteriorate towards right heart failure or recurrent syncope. The creation of an atrial communication by catheter intervention is known to improve symptoms, but experience is limited, particularly in children. There is a known risk of spontaneous closure of catheter created inter-atrial communications, especially created by graded balloon atrial septostomy. The Amplatzer fenestrated atrial septal device is designed to maintain patency in this setting and we report our initial experience.
Methods: A fenestrated device was manufactured to order for each named patient. Cardiac catheterisation was performed under general anaesthesia and pulmonary vascular resistance (PVR) was measured. Atrial septal needle puncture was performed and the puncture was balloon dilated appropriately. A standard delivery system was used to deploy and release the device, under echocardiographic and fluoroscopic control. Patients were anticoagulated after the procedure.
Results/Patients: Five children (age 18 m–17 y mean 8.4 y) with severe PPH underwent the procedure. All were in NYHA III and IV, had severe exercise intolerance, and were receiving specific medical treatment. Four patients had syncope or pre-syncope, and 1 patient was in right ventricular failure. Mean systemic arterial oxygen saturation (SaO_2) was 98.6%.
Cardiac Catheterisation: Mean pulmonary arterial pressure approached or exceeded mean systemic blood pressure, and mean pulmonary vascular resistance PVR was 12–21 U·m^2, mean 15 U·m^2, fixed in all cases. The procedure was successful in all patients. Two patients with the most severe disease needed intensive care for 9 to 14 days after the procedure.
Follow Up: After 1–6 m, mean 3 m, all patients are alive with patent fenestrations. Mean SaO_2 at follow up decreased by 3 to 15%, mean 7.6%. All have shown symptomatic improvement and resolution of syncope or pre-syncope.

Patient	Indications	Sex	Age (yrs)	PAP/ SAP	Pre-Saturation	Post-Saturation	PVR U·m^2
1	Syncope	F	12.5	61/72	98	94	16.9
2	RV Failure	F	17	71/61	98	83	21.4
3	Syncope	F	4.3	24/58	99	95	12
4	PreSyncope	M	5.2	54/55	98	87	14
5	PreSyncope	M	1.3	53/56	100	97	14.4

Haemodynamic and demographic data in patients having fenestrated atrial septal device inserted. RV: Right ventricle, PAP: Mean Pulmonary Artery, SAP: Mean Systemic Artery, PVR: Pulmonary Vascular Resistance.

Conclusion: The application of fenestrated atrial septal devices is feasible in children with severe PHT and is associated with clinical improvement. Long term patency of the atrial communication has yet to be confirmed, and optimum timing for the procedure has to be determined.

P13

Sildenafil influence on cerebral oxygenation measured by near infrared spectroscopy in infants after cardiac surgery

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Background: Sildenafil (Viagra®) is claimed to be a specific and effective pulmonary vasodilator and being used in newborns and infants with pulmonary hypertension after cardiac surgery. However, it is unclear whether there are side effects on cerebral vasomotility in this very vulnerable patient group.
Objectives: To investigate the effect of sildenafil medication on cerebral oxygenation in infants with elevated pulmonary vascular resistance after cardiac surgery using near-infrared spectroscopy (NIRS).
Method: Sildenafil was applied in three steps of 10 minutes with cumulative doses of 0.1, 0.3 and 0.6 mg/kg. We examined the changes of oxygenated haemoglobin (HbO_2), deoxygenated haemoglobin (HHb), total haemoglobin (tHb) concentration and cytochrome oxidase (CytOx) oxygenation and cerebral tissue oxygenation index (TOI) in 13 children (median age 4.5; range 2.1–20.9 months, median weight 5.5; range 3.5–10.6 kg).
Results: A significant increase in cerebral O_2Hb ($\Delta 2.3 \pm 0.6$ μmol/L) and tHb (Δ 0.9 ± 0.2 μmol/L) at the beginning of intravenous sildenafil administration with a decrease in HHb ($\Delta 1.3 \pm 0.4$ μmol/L) was observed. These changes lead to a significant elevation in cerebral TOI from 63.4 ± 2.5% to 65.7 ± 2.8%, while mean systemic arterial pressure and arterial oxygen partial pressure tended to decrease.
Conclusions: We conclude that an increased cerebral haemoglobin and oxygen supply during sildenafil application might indicate cerebral arterial vasodilatation, thus overriding normal cerebral autoregulation. Its use in young children should be guarded with caution.

P14

Hypoxic Pulmonary Hypertension: Aqueous Oxygen better than Prostaglandins

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Objective: To evaluate the effects on pulmonary circulation of Prostaglandins versus Aqueous Oxygen with induced hypoxic pulmonary hypertension.
Methods: Sixteen calves, 2-month-old, 73 ± 7 kg, underwent general anaesthesia, mechanical ventilation and median sternotomy. Catheters for continuous pressure and blood gas measurements were inserted in carotid and femoral arteries, left atrium, right atrium and pulmonary artery (PA), and a flow-probe placed around the PA.

After baseline measurements, hypoxic ventilation reduced the mean arterial PO_2 from 289 ± 98 mmHg to 42 ± 12 mmHg

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($P < 0.0001$). At this point, without changes in the hypoxic ventilation (the mean arterial PO_2 was maintained at 42 ± 13 mmHg), the effects of 3 ml/min of hyperbaric aqueous oxygen (AO = oxygen diluted in saline solution) infused directly into the PA for 30 minutes (n = 11) were compared with Prostaglandins infusion (0.1–0.2 μg/kg/min, n = 5), with continuous reading of the monitored parameters.

Results: Hypoxic ventilation raised significantly the values of systolic ($P < 0.01$), diastolic ($P < 0.05$) and mean ($P < 0.05$) PA pressure, PA/systemic pressure ratio for systolic ($P < 0.0001$) and mean ($P < 0.001$) pressures and Pulmonary Vascular Resistance ($P < 0.001$), while the Pulmonary Blood Flow decreased ($P < 0.01$).

AO infusion reduced significantly ($P < 0.005$) the values obtained with hypoxic ventilation with systolic (26 ± 6 vs 36 ± 7 mmHg), diastolic (11 ± 4 vs 16 ± 3 mmHg) and mean (16 ± 4 vs 24 ± 4 mmHg) PA pressure, PA/systemic pressure ratio for systolic (0.27 ± 0.07 vs 0.47 ± 0.09) and mean (0.27 ± 0.08 vs 0.49 ± 0.13) pressures and Pulmonary Vascular Resistance (3.42 ± 0.31 vs 6.89 ± 0.87 U), while the Pulmonary Blood Flow increased (3.6 ± 0.4 vs 2.7 ± 0.4 L/min). The only significant ($P < 0.05$) improvement obtained with Prostaglandins was the reduction of Pulmonary Vascular Resistance (5.36 ± 0.81 vs 6.67 ± 0.24), while all other parameters remained not statistically different the values obtained with hypoxic ventilation before Prostaglandins infusion. In one animal ventricular fibrillation occurred immediately after beginning of the infusion of Prostaglandins.

Conclusions: Acute infusion of hyperbaric AO solution into the PA can completely reverse all the negative effects induced by acute hypoxia on the pulmonary circulation; in the same situation, the effects of Prostaglandins are limited to a reduction of Pulmonary Vascular Resistance.

P15
Comparison of the acute pulmonary vasodilator effect of beraprost sodium and nitric oxide in congenital heart disease: Thailand multicenter study

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Background: Generally the cardiac catheterization was performed preoperatively to determine the acute vasodilator response in patients who had congenital heart disease with pulmonary hypertension (PHT). We compared the acute pulmonary vasodilator effect of orally administrated beraprost sodium with Nitric Oxide (NO) and 100% oxygen.

Methods: Patients (pts) who had congenital heart disease with severe PHT that required cardiac catheterization were enrolled in our study. Standard left and right cardiac catheterization were performed. Patients were tested with 100% oxygen, 40 ppm NO and 1 mcg/kg/min of beraprost sodium (Dorner®) given orally.

Results: There were 90 pts who had PHT undergone cardiac catheterizations. Their mean age was 15.6 ± 16.4 years old and weight was 26.1 ± 18 kg. There were 25 ASD, 44 VSD, 5 DORV and 12 other complex congenital heart lesions. Their base line hemodynamic data showed mean pulmonary pressure (mPA) 66.9 ± 17.1 mmHg, Qp:Qs = 1.9 ± 1.2 and pulmonary arteriolar resistance (Rpa) 14.7 ± 12.9 unit m^2. After 100% O_2 was given for 10 min the hemodynamic data were changed to mPA 61.7 ± 18.1 mmHg, Qp:Qs = 5.1 ± 8.1 Rpa 8 ± 9.5 unit m^2. NO (40 ppm) showed mPA 60.6 ± 16.7 mmHg, Qp:Qs = 3.2 ± 3.4 Rpa 9.3 ± 9.7 unit m^2. At 30 min after beraprost sodium was given, the data were mPA 62.1 ± 18.2 mmHg, Qp:Qs = 2.9 ± 2.5 Rpa 9.5 ± 8.9 unit m^2. There was a significant change in mPA, Qp:Qs and Rpa when compared room air and 100% O_2 condition (all $p < 0.001$). When compared to 100% O_2, NO at 40 ppm appeared to show no difference in the vasodilator effect in all measurements. Using beraprost sodium we found that there was a comparable pulmoanary vasodilator effect with 100% O_2 in lower Rpa but not in Qp:Qs.

Conclusions: It appeared that the acute pulmonary vasodilator effect of beraprost sodium was comparable to 100% O_2 and NO 40 ppm in preoperative evaluation of patients with pulmonary hypertension for congenital heart surgery. The benefit of using beraprost sodium is that it is readily available when compared with NO. It also gave more accurate Qp:Qs calculation when compared with 100% O_2. We could also use the results as a postoperative guideline management for this group of patient.

P16
Neonatal pulmonary embolism

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Background: Pulmonary embolism is very rare in the neonatal period. Controlled trials on this subjects are not feasible. Literature research has yielded only a few case reports.

Patients and Methods: We report on two newborns who presented with cyanosis. Both of them had been diagnosed with persistent pulmonary hypertension of the newborn (PPHN) when referred to our center. The first infant presented with persistent postpartal cyanosis even after intubation and mechanical ventilation. Echocardiography showed a moderate tricuspid valve regurgitation, a significantly dilated right atrium as well as only minimal blood flow across the pulmonary valve. Cardiac catheterization revealed bilateral thrombosis of both pulmonary artery lower segment branches. After 4 days of thrombolytic therapy with rt-PA echocardiography showed a normal pulmonary artery flow pattern. The radionucleid scan then presented with a regular pulmonary artery perfusion on all sides. The hypercoagulation screen detected an elevated level of "lipoprotein a" which has been linked to increased risk of thrombophilia in childhood. The second infant, a full term newborn, showed a sudden onset of dyspnea during breast feeding on day 7. The baby was presented to the local children's hospital and needed intubation and mechanical ventilation. Assuming congenital heart disease a therapy with PGE2 was started. By echocardiography we demonstrated a right ventricular dilatation with significant reduction of right ventricular function, moderate tricuspid valve regurgitation as well as right-left shunting across a persistent foramen ovale. In addition a suspicious structure thought to be a thrombus was found in the right pulmonary artery. Cardiac catheterization confirmed marked embolism of the bilateral pulmonary arteries. Local therapy with rt-PA was followed by a systemic therapy with rt-PA. After a 3 day treatment without any side effects pulmonary embolism could not be detected any more. There were no risk factors for thrombophilia. In both children an anticoagulant therapy with warfarin was initiated.

Conclusions: In newborns presenting with clinical signs of PPHN the rare condition of a pulmonary embolism should be considered as a differential diagnosis. We were able to demonstrate sufficient thrombolytic therapy with rt-PA in two patients without any complications.

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P17

Vascular endothelial growth factor, IL-6 and IL-8 levels in congenital heart disease

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Objectives: Increased pulmonary flow in congenital heart diseases with left to right shunt lesions and hypoxia in the congenital heart diseases with right to left shunt lesions, constitute the main course cause of the morbidity and mortality.
Methods: Patients (31 F, 22 M; 5 months–16 y, mean: 4.31 ± 4.19 y) were divided into two groups according to their shunt lesions: acyanotic group 1 (group 1a: pulmonar artery systolic pressure > 30 mmHg and group 1b < 30 mmHg) and cyanotic group. The same patients were then divided into two other groups: Group PA1 (pulmonary artery systolic pressure >30 mmHg) and group PA2 < 30 mmHg; Group Rp1 (pulmonary resistance < 2 Ü/m^2) and GroupRp2 (>2 Ü/m^2).
Results: The VEGF level (pg/ml) was increased in Group 1 (219.2 ± 32.8 vs 125.2 ± 207.1), Group 1a (300 ± 425 vs 117 ± 93), GroupPA2 (244.8 ± 374 vs 132.0 ± 173) $p > 0.05$ and in Group Rp2 (320.8 ± 411.6 vs 102.8 ± 128.9) $p < 0.05$. The IL-6 level (pg/ml) was increased in Group 2 (38.3 ± 128.0 vs 4.09 ± 6.12), Group 1a (5.43 ± 8.02 vs 2.41 ± 0.99), Group PA2 (27.6 ± 103.4 vs 2.62 ± 1.70), $p > 0.05$. The IL-8 level (pg/ml) was increased in Group 2 (44.1 ± 68.1 vs 27.0 ± 68.4), Group 1a (43.4 ± 89.5 vs 6.42 ± 4.91), Group PA2 (48.8 ± 83.3 vs 14.10 ± 35.07) $p > 0.05$, and Group Rp2 (43.8 ± 89.31 vs 7.72 ± 7.51) $p < 0.05$. Positive correlations were found between VEGF and IL6, IL8, pulmonary artery systolic and mean pressures. Arterial 02 saturations showed
Conclusions: VEGF plays an important role in the development of irreversible pulmonary hypertension; therefore evaluation of VEGF levels could be guiding in the management of the treatment.

P18

Prophylactic ECMO after heart-transplantation in children with elevated pulmonary vascular resistance

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Elevated pulmonary vascular resistance (PVR) secondary to left heart failure and pulmonary venous hypertension may cause donor right heart failure after orthotopic heart transplantation. We report of 3 children with elevated PVR, who were placed on ECMO immedeatly after HTX as a prophylactic treatment against right heart failure.
Patients: Age at HTX: 9, 20, 34 months. Diagnoses: Aortic Stenosis with Endocardial Fibroelastosis (2), Dilated Cardiomyopathy (1). Preop. PVRindex: 11.2, 12.8, 11.7 WU m^2, PVR/SVR-ratio 0.73, 0.73, 0.43. Inhalative prostacycline reduced PVR in 1 of 3 children.
Results: ECMO-duration. 46, 62, 72 hrs. Weaning with inhalative-NO, iv-Prostacycline and inotropic support. PAP/SAP-ratio after ECMO-expl.: 0.4, 0.4, 0.3. PVRindex 6 months after HTX: 3.5, 3.9, 2.3 WU m^2. Uneventfull follow up over 7, 15, 18 months.
Discussion: ECMO allows the right ventricle to recover from ischemic disorder following HTX and to adapt to elevated pulmonary pressures. In young children a marked reduction of elevated PVR due to left heart failure could be expected after HTX. Failing response to inhalative prostacycline in the preoperative testing does not exclude this change in PVR. The level at which PVR becomes an absolute contraindication against HTX in children remains unknown.
Conclusions: With prophylactic use of ECMO HTX is feasible in children with highly elevated PVR.

P19

Bosentan for pulmonary arterial hypertension in infants and young children with congenital heart disease: first experience

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Introduction: Bosentan, a dual endothelin-receptor antagonist, has been shown to be an effective treatment option in patients with the idiopathic form of pulmonary arterial hypertension (PAH). We used Bosentan as a compassionate treatment option in infants and young children with congenital heart disease (CHD) who had PAH preoperatively representing a contraindication to corrective surgery or persisting PAH after corrective surgery causing right heart failure and reduced exercise tolerance.
Methods: 7 consecutive pediatric patients with PAH due to CHD (age range; 6 months to 4.7 years) received 3 mg/kg Bosentan in two daily doses. Clinical, echocardiographic and hemodynamic parameters were measured before treatment, and during steady state while on treatment. Routine liver function parameters were monitored monthly.
Results: Bosentan treatment times ranged from 4–19 months. During Bosentan therapy, there were no significant adverse events. The clinical status remained stable in all patients. NYHA-class changed from 2.6 ± 0.7 to 1.6 ± 0.5 ($p < 0.05$). We found a mean reduction of the right ventricular systolic pressure (RVSP) from 96 ± 11 mmHg to 72 ± 26 mmHg ($p < 0.05$).
Conclusions: Treatment with Bosentan in infants and young children with PAH due to congenital heart disease was tolerated without significant side effects and resulted in stabilisation of clinical status. A significant reduction in echocardiographic RVSP could be demonstrated. These results suggest that the dose regimen used may be appropriate and secure for the treatment of young pediatric patients with PAH, resulting in a reduction of pathologically increased pulmonary vascular resistance.

P20

Decreased numbers of T lymphocytes and predominance of macrophages-granulocytes in the walls of peripheral pulmonary arteries from patients with pulmonary hypertension secondary to congenital cardiac shunts

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Objective: Inflammatory cells have been described in lesions of primary Pulmonary Hypertension (PH). Their role in secondary PH has not been investigated.
Methods: We analyzed lung biopsies collected from 26 patients with PH secondary to congenital heart defects (mean age 32.3; median 21.5 months) and 11 necropsy lung fragments from controls (mean age 23.4; median 10 months). In the PH group we classified the arterial lesions according to the Heath-Edwards system: HE 1 – isolated medial hypertrophy and HE ⩾ 2 – presence of intimal proliferation. Histological sections were submitted to immunohistochemistry in

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order to label inflammatory cells as follows: CD3 and CD20 monoclonal antibodies for T and B lymphocytes respectively and Mac387 for recently recruited macrophages and granulocytes (MAC-GR). With the aid of an image analysis system we quantified the inflammatory cells present in the walls of arteries $> 50\,\mu m$ in diameter and expressed the results as: (a) cell density/area of the adventitia and (b) indices of cells crossing the medial and intimal layers (number of crossing cells × number of positive arteries/total number of arteries).
Results: No difference was found between the HP and control groups regarding the sum of densities of adventitial inflammatory cells ($p > 0.05$). However, the proportion of each cell type varied within each group (RM ANOVA, $p < 0.004$). MAC-GR prevailed in PH patients, while T lymphocytes were the predominant cells in the controls and decreased in the PH group. Comparing PH samples according to the absence or presence of intimal proliferative lesions, there was no prevailing cell in the first (HE 1; $p = 0.217$), but in the second (HE ⩾ 2), MAC-GR were prevalent over total lymphocytes (RM ANOVA, $p < 0.02$). The indices of inflammatory cells crossing the arterial wall was significantly greater for MAC-GR when compared to lymphocytes in PH patients but did not differ within the control group.
Conclusion: The predominance of recently recruited MAC-GR in PH patients is compatible with ongoing inflammatory reaction in the arterial walls and may reflect the participation of such cells in the pathogenesis of PH lesions. On the other hand, the decrease in T lymphocytes may indicate a defective maturation of these cells in the lungs of patients with congenital cardiac shunts.

P21
Advantages of aerosolized iloprost in acute exacerbation of pulmonary hypertension secondary to vein of Galen arteriovenous malformation
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Objectives: Neonatal arteriovenous malformation of the vein of Galen is a rare disease. Neuroradiological therapy consists of embolization with coils and adhesive liquids. Despite of advancing interventional strategies the outcome is still complicated by cerebral lesions and residual av-shunts. Pulmonary hypertension (PH) leading to right heart failure is the most severe extracerebral complication. Treatment of PH with prostacyclin analogues is becoming a promising option.
Methods and Results: We report about two female neonates with early development of PH and different clinical outcome. The first underwent neuroradiological interventions twice with only few residual feeding vessels, that were not assumed to be responsible for persistent PH. Cardiac catheterization revealed suprasystemic pressure in the pulmonary artery without response to oxygen and nitric oxide (NO). Aerosolized iloprost (AI) was used as rescue treatment and caused short time improvement of oxygenation. The patient died at the age of 90 days, postmortal studies of the lungs revealed microembolie and intimal proliferation. The second patient had to be treated three times with coil-embolization. Prior to the last intervention PH was refractory to all used measures including intravenous prostacyclin. AI potently improved oxygenation without a decline of systemic arterial pressure. It was applied in intervalls of 2 hours until the shunt-volume could be effectively reduced by the final intervention. PH ceased and therapy with AI could be stopped.
Conclusions: PH in arteriovenous malformation of the vein of Galen can be complicated by microembolic events and can end up in fixed PH at early age. Acute exacerbations of shunt-induced PH can effectively be treated with aerosolized iloprost. It seems to be less successful in PH complicated by thrombembolic events.

P22
Children and adults with the Eisenmenger Syndrome may benefit from therapy with bosentan (Tracleer®), an endothelin receptor antagonist
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Background: Patients with the Eisenmenger Syndrome (EM) may benefit from enhancement of pulmonary blood flow (Qp), thus augmenting systemic oxygen delivery. This may be achieved with preferential reduction of pulmonary vascular resistance (PVR). Bosentan, a dual endothelin receptor antagonist, has been shown to effectively decrease PVR. We used bosentan in a small patient group with EM and clinically observed its effects.
Patients and Methods: 7 patients with congenital shunting defects and the EM syndrome (age, 14–62, mean 33.4 years) received bosentan at a final dose of 3 mg/kg/day in 2 doses. At regular intervals, vital parameters, NYHA class, 6-min walk tests, echocardiograms, and laboratory parameters were controlled.
Results: The medication was well tolerated in all cases without significant laboratory parameter changes. NYHA-class improved (2.7 ± 0.5 to 2.1 ± 0.4, $p = 0.03$), transcutaneous saturation increased (80 ± 9 to $87 \pm 6\%$, $p = 0.02$), which was associated with a trend in improvement of the 6-min-walking distance (298 to 386 meter, $p = 0.11$) and a trend in decrease of right ventricular systolic pressure by echocardiography (116 ± 36 to 108 ± 16 mmHg, $p = 0.09$).
Conclusions: Patients with the Eisenmenger Syndrome may experience clinical improvement and increase in systemic oxygen saturation when treated with bosentan, an endothelin receptor antagonist.

Session 3: New Technological Developments

P23
MRI-guided dilatation of aortic coarctation and stent placement: feasibility and comparison to x-ray guided catheter techniques
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The aim of the study was to assess the feasibility of MRI guided dilatation of artificially created coarctation of the aorta (CoA) and stent placement using intravascular miniature antenna guidewires in a porcine model. The results where directly compared to conventional x-ray guided catheter interventions.
Methods: In eight healthy farm pigs, a coarctation of the aorta was created by operative banding of the aorta at the site of the ductus arteriosus (60 to 90% stenosis). After 2–8 weeks all animals underwent conventional x-ray guided catheterization for a complete hemodynamic and anatomical evaluation of the CoA and 4 pigs had balloon dilatation. The other 4 pigs were transported to a dedicated cardiac MR-scanner (Sonata, Siemens). Anatomical and hemodynamic MR imaging was performed and compared to

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x-ray. Real-time MRI was performed with a steady-state free precession pulse sequence ("true FISP"; TR = 1.4 ms, TE = 1.15 ms, flip angle = 40°, 50–60 phase-encodings, 128 by 128 image matrix, 7–15 frames/sec, 1400 Hz/pixel, 35 mm slice thickness). With the real-time pulse sequence running under interactive user-control a costumized delivery sheath (AGA medical) was tracked towards the stenosis. A self-expanding nitinol stent was delivered and a balloon dilatation was performed. Following the procedure both MR and x-ray measurements were compared to intraoperative measurements and pathology.

Results: Real-time MRI can be used to follow the position of an antenna guiding catheter within the aorta and its branches. Successful deployment of the stent and balloon dilatation of the CoA could be monitored in all cases. The correlation of MR measurements of the diameters of the stenosis to intraoperative measurements was ($y = 1.01x - 0.24$, $r = 0.96$) and ($y = 0.90x + 0.24$, $r = 0.62$) for x-ray measurements. There was no significant difference in gradient measurements (MR: 52 ± 23 vs x-ray: 51 ± 18 mmHg; $p = 0.13$). Procedure time was significantly shorter using x-ray (23 ± 12 vs. 35 ± 28 minutes; $p < 0.05$) including a learning curve.

Conclusions: These data demonstrate that real time MR guided balloon-dilatation and stent placement is feasible in aortic coarctation. As compared to x-ray MRI has advantages in regards to radiation exposure and anatomical imaging but for the costs of higher procedure times.

P24
Adjustable pulmonary artery banding in children with muscular VSD for long-term palliation

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Objectives: Single or multiple muscular VSDs with significant left to right shunt require banding of the pulmonary artery in the first months of life. An optimized flow restriction can rarely be achieved or maintained over time. We report our experience with the Flow watch™ PAB (Endoart,Ch) banding device that can be adjusted completely noninvasive according the hemodynamic requirements.

Patients: Between 6/03 and 1/04 three patients with an age of 25–93 dd (mean 59) and a weight of 2,7–4,2 kg (mean 3,4) had implantation of the Flowwatch™ PAB. Diagnoses were muscular VSD in all and systemic pulmonary artery hypertension; associated lesions were: interrupted aortic arch (n = 1), additional perimembranous VSD (n = 1) and multiple non cardiac anomalies as microcephalus, 5th grade vesico-urethral reflux, hydronephrosis, butterfly vertebrae, cleft palate. One patient had dislocation of a conventional banding before.

Methods: The Flowwatch™ PAB was placed through median sternotomy around the pulmonary artery trunk. Postoperatively the device was tightened by external telemetric adjustment guided by echo and hemodynamic measurements.

Results: Implantation of the Flowwatch™ PAB was uneventful in all patients. Effective restriction of pulmonary blood flow was achieved by a degree of 50–75% stroke in the early postoperative period, corresponding to Doppler gradients between 45 and 65 mmHg. Within the first ten days after surgery a stepwise final adjustment was performed to reduce the pulmonary artery pressure to at least 50% of the systemic blood pressure what was very well tolerated by the patients. A release of the banding was necessary in one patient 12 hours after surgery with immediate stabilization of cardiac function. Up to now no further adjustment was necessary. One patient was operated on cleft palate and antireflux surgery 5 months later without any cardiac problems.

Conclusions: In patients with multiple VSD and pulmonary hypertension the placement of a Flowwatch™ PAB, surgical closure can be delayed. Gradual adaption of the right ventricle can reduce morbidity. Especially in patients with multiple organ problems, the device offers the opportunity of a staged repair without risk of fixed pulmonary hypertension. The completely noninvasive telemetrically adjustment is the main advantage to other banding devices.

P25
Decreasing ratio of plasma N-terminal pro-B-type natriuretic peptide and B-type natriuretic peptide according to age

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Recent studies suggest that B-type natriuretic peptide (BNP) and its N-terminal fragment (NT-proBNP) may be useful diagnostic tools in children with heart disease. Recently, we have reported normal concentrations of both parameter from infancy to adolescence (Heart 2003, 89:875-8, Clin. Chem. 2003, 49:1563-4). BNP levels are relatively stable after a rapid decrease during the first weeks of life, but NT-proBNP concentrations are decreasing with increasing age.

In 46 healthy subjects (22 males, 24 females) aged 0.4 to 17.5 years and 30 patients with congenital heart disease (17 males, 13 females) aged 0.2 to 18.4 years, plasma levels of BNP and NT-proBNP were measured in the same venous blood sample. BNP was measured by sandwich immunoassay (triage BNP assay, Biosite®), and NT-proBNP by the electrochemiluminescent immunoassay system Elecsys 2010 (Roche Diagnostics®).

The range of BNP plasma levels was 5 to 32 pg/ml in healthy subjects and 5 to 1300 pg/ml in the patient group, the range of NT-proBNP was 10 to 298 pg/ml and 30 to 18966 pg/ml. In both groups, the ratio NT-proBNP/BNP decreased with increasing age: Linear regression equation for healthy subjects: $\ln(\text{NT-proBNP/BNP}) = 2.332 - (0.083 \times \text{age})$, $r = -0.582$; $p < 0.001$. Linear regression equation for both groups: $\ln(\text{NT-proBNP/BNP}) = 2.747 - (0.109 \times \text{age})$, $r = -0.613$; $p < 0.001$.

ProBNP is cleaved into the two fragments NT-proBNP and BNP. However, the ratio of plasma NT-proBNP to BNP is not stable. There is a decreasing NT-proBNP/BNP ratio with increasing age. Differences in the metabolic clearance of both peptides during childhood may cause the different distribution of NT-proBNP/BNP in plasma according to age. Therefore, results of BNP studies should be reported with annotation of the assay used.

P26
Atrial resynchronisation after heart transplant in childhood

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Objectives: Exercise capacity is known to be reduced in clinically stable cardiac transplant recipients, there is a substantial attenuation

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in the peak heart rate response to a maximal exercise effort due to the lack of direct sympathetic innervation of the donor SA node. Recipient SA node can still respond to this stimulation and we hypothesised that resynchronisation of both atria could improve exercise tolerance in these patients.
Methods: We report our experience with two patients transplanted in childhood following end stage cardiomyopathy presenting incapacitating discomfort in accomplishing simple daily tasks (awakening, climbing few steps) despite good graft function without evidence of rejection or active arrhythmia. Objectively, they both show chronotropic incompetence during a treadmill exercise test (maximal upper rate 131 and 141 bpm). Two atrial bipolar screw-in endovenous leads were implanted, one in the remnant receiver's right atrium A1 (amplitude 1.0 mV and 1.6 mV) and the other in the right atrial appendage of the donor's heart A2 (amplitude 5.2 mV and 3.0 mV). Atrias were synchronised, triggering stimulation of A2 after sensing A1 by using a standard DDD pacemaker (Medtronic Kappa KDR931) programmed in DDD mode with AV delay (A1A2 delay) of 30 msec, and where A1 was in the atrial port and A2 in the ventricular port.
Results: Both patients felt subjectively much better and graft acceleration could be appreciated even with daily activities and confirmed by treadmill test (maximal upper rate 163 and 168 bpm respectively).
Conclusions: A1A2 resynchronisation after heart transplant in childhood is feasible, with no adverse effects and results in significant improvement of quality of life and exercise capacity due to restored chronotropic competence. It should be considered in active heart transplanted patients.

P27
Evaluation of remote stethoscopy for the assessment of heart murmurs in children
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Objectives: To assess the accuracy of "live" remote stethoscopy in the assessment of heart murmurs in children.
Methods: Consecutive new patients referred for assessment of a heart murmur were consented. Two groups of patients were studied. In the first group, all children were examined independently by two blinded consultant paediatric cardiologists "face-to-face" (FF). In the second group, all children were examined "face-to-face" by one cardiologist, and then evaluated by the second cardiologist remotely using an electronic stethoscope, with heart sounds and video images transmitted "live" utilizing an on site 384 Kbps telemedicine system (TM). A telemedicine facilitator assisted with stethoscope position. Main outcome measures included agreement between examiners, agreement with echocardiographic diagnosis, and frequency of request for further investigations.
Results: 75 patients were enrolled with 39 in group 1 (FF/FF), and 36 in group 2 (FF/TM). Inter-observer agreement was satisfactory for presence/absence of heart disease (FF/FF, $\kappa = 0.92$; FF/TM, $\kappa = 0.92$), and for clinical diagnosis (FF/FF, $\kappa = 0.85$; FF/TM, $\kappa = 0.92$). In group 1, there was strong agreement between clinical examination and echocardiographic assessment for presence/absence of heart disease ($\text{sensitivity}_A = 1.00$, $\text{specificity}_A = 0.93$; $\text{sensitivity}_B = 1.00$, $\text{specificity}_B = 1.00$). There were no cases of heart disease missed by TM assessment (sensitivity = 1.00, specificity = 0.93). Echocardiography request rates were similar between patients assessed by TM (30%), and in the FF controls (25%).
Conclusions: Remote stethoscopy appears to provide an accurate method for evaluating children referred for assessment of a murmur. It may be a useful modality for supplying an outreach outpatient service.

Session 4: Arrhythmia and Electrophysiology

P28
Autocapture with epicardial leads: impact of the lead position on the evoked response amplitude
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Objectives: A high evoked response (ER) signal is mandatory to ensure correct functioning of modern threshold tracking algorithms like SJM Autocapture™ which improve pacing safety and therefore the patients safety. The influence of the position of epicardial leads on the ER amplitude has not been investigated systematically.
Method: After median sternotomy the poles of bipolar epicardial steroid eluting leads (Medtronic CapsureEPI 4968) were fixed on the hearts of 10 piglets (21 ± 3.5 kg) at 4 different positions, which were RV = right ventricular outflow tract, RV + LV = right and left ventricular anterior wall, LV1 = left ventricular anterior wall, LV2 = left ventricular anterior and posterior wall. ER signals were measured with a SJM Microny SR + 2524T pacemaker at 5 different pulse widths (0.21, 0.37, 0.49, 0.7, 1.0 ms).
Results: The Table shows all summarized ER-signals at the 4 positions (mean ± SEM).

ER-signal	RV	RV + LV	LV1	LV2
mean ± SEM	6.68 ± 0.34	5.48 ± 0.33	12.39 ± 0.82	16.8 ± 1.13

Conclusions: The data indicate that fixing the poles of bipolar epicardial leads on the left ventricular anterior wall or on the anterior and posterior wall will result in higher ER amplitudes.

P29
Effect of betablockers in children with the long QT1 syndrome
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Objective: To evaluate the effect of beta-blockers (β-) in children with the long QT1 syndrome (LQT1) we reviewed the outcome of 61 children with a single KCNQ1 mutation. Patients and Methods: Among 122 children who were diagnosed with LQTS between 1984 and 2002, 118 were genotyped and 60/118 had a single mutation in KNCQ1. Age at diagnosis ranged from 1 day to 15 years. 25 pts had syncope or cardiac arrest and 4 neonates presented with isolated bradycardia The remaining 31 pts were diagnosed after familial screening. ECG showed a prolonged QTc of 450–580 ms in 47 pts, and 13 pts had QTc <450 ms with torsade de pointes documented in one case. All pts were treated with

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β-, propranolol in 11 pts (3–5 mg/kg) nadolol in 42 pts (50 mg/m^2) and acebutolol/atenolol in 7 pts, aiming at a maximum heart rate of 150 bpm <2 years and 130 bpm >2 years, annually checked by exercise testing and/or 24-h ECG monitoring.
Results: A 15 year-old girl diagnosed at 9 years after a syncope had a QTc of 420 ms and died suddenly after the family had stopped the treatment; another death was unrelated to LQTS. Follow-up of survivors was 2–17 years (7.5 ± 4.5 years). There were two side effects of β-: one pt had hypoglycemia and another had Raynaud phenomenon which disappeared when nadolol was changed to bisoprolol. During follow-up, propranolol was changed to nadolol in 5 pts for drug administration convenience, so that a total of 46 pts were on nadolol at the end of follow-up. All the 58/60 surviving pts are alive and free of symptoms, with properly beta-blocked sinus rhythm.
Conclusions: Treatment with β- alone was effective in all compliant pts with a single KCNQ1 mutation, with no cardiac event and no significant side effect. However, all genotyped LQT1 pts should be treated, even if they have a normal QT interval on ECG.

P30
Clinical usefulness of the combination of late ventricular potentials and QT dispersion for predicting ventricular tachycardia in children with mitral valve prolapse: a prospective study
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Mitral valve prolapse (MVP) is the commonest cardiac valvular abnormality in children. Several studies have shown a high prevalence of complex or frequent ventricular arrhythmias (VA) in these patients. The aim of this study was to examine prospectively the QT dispersion (QTd) and late ventricular potentials (LP) as risk markers for ventricular tachycardia (VT) in children with MVP.
Material and Methods: 151 children with MVP (age ± SD: 12.2 ± 3.1 years) were examined. All the children underwent a clinical examination, standard 12-lead resting ECG, signal-averaged ECG, 24-hour ambulatory ECG monitoring and echocardiography. The children with MVP were followed prospectively for a mean of 64 months. Follow-up studies were performed by clinical examinations, standard resting ECG, and by 24-hour ambulatory ECG monitoring at maximum intervals of 6 months.
Results: VA were recorded in 41.7% of patients with MVP. LP were more common (27% vs. 10%, $p < 0.02$) and QTd was significantly greater (44 ± 19 vs. 27 ± 14 ms; $p < 0.00001$) in children with MVP and VA compared with those without VA. During follow-up, 24 children with MVP developed nonsustained VT (3.1/100 subject-years). The sensitivity, specificity, positive predictive value (ppv) and negative predictive value (npv) of a QTd of ⩾50 ms in the identification of those children with MVP who are at high risk of developing VT were 79.2%, 85%, 50% and 95.6%, respectively. 14 out of 27 patients with MVP who presented with VT during study period had an abnormal signal-averaged ECG. To identify children with MVP who developed VT, the presence of LP had a sensitivity of 52% and a specificity of 90%, a ppv of 50% and a npv of 91%. A combined algorithm using both QTd ⩾50 ms and LP was the best predictor for VT (sensitivity: 83.3%; specificity: 95.2%; ppv: 66.7% and npv: 98.0%).
Conclusions: The combination of QTd and an abnormal signal-averaged ECG could be a useful index for identifying those children with MVP who are at risk for VT. A high specificity and high negative predictive value indicate that those children with MVP with QT dispersion below 50 ms and without LP on signal-averaged ECG are at low risk of developing VT.

P31
Atrio-atrial dual chamber pacing for restoration of sinus node dysfunction in a heart transplanted child
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Sinus node dysfunction (SND) of a transplanted heart is a well known phenomenon. Using the normal chronotropic function of the patients own remaining sinus node seems to be reasonable.
Patient: 11 years after successful heart transplantation for dilated cardiomyopathy a 13 year old teenage girl (164 cm, 54 kg) suffered from sustained typical atrial flutter (Afl) in the transplanted heart with consecutive impaired LV-function. Isthmic ablation terminated the Afl but SND with chronotropic incompetence resulted in continuous VVI pacing (epicardial rejection control device) while the patients own sinus node showed a normal heart rate variability. Therefore a dual chamber pacemaker (Medtronic AT500) with bipolar sensing capabilities for little signals in the atrial and ventricular channel, respectively, was implanted transvenously fixing 2 bipolar screw-in leads into the transplanted right atrium (ventricular channel) and the remaining part of the patients right atrium (atrial channel). Sensing and threshold parameters were satisfying. Programming a DDD-mode allowed correct tracking of the patients own sinus rate while pacing the transplanted atrium.
Conclusions: The concept of implantation of an 'atrio-atrial' DDD-pacemaker in a heart transplanted child can restore sinus node dysfunction of the transplanted heart.

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Rhythm abnormalities in patients with isolated ventricular noncompaction
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Isolated noncompaction of the ventricular myocardium (IVNC) is a rare congenital heart disease that results from an abnormal arrest in endomyocardial embryogenesis. The disorder is clinically accompanied by depressed ventricular function, endocardial clot with systemic embolization, and arrhythmias. This study consists of eleven patients followed in pediatric cardiology department with IVNC and cardiac rhythm abnormalities. Age at presentation ranged one day to 16 years. Clinically overt left ventricular dysfunction was present in six patients. Ventricular arrhythmias were detected in four patients. Two of whom admitted with syncope, and two with palpitation. In these patients different abnormalities were detected in surface electrocardiogram (ECG). Intracardiac electrophysiologic study revealed ventricular fibrillation in two, and ventricular tachycardia in other two of these patients. Implantable cardioverter defibrillator (ICD) was implanted in three of them. Four patients

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had sinus bradycardia; one was seen for the first time at 32 weeks' gestation and the fetus had moderate hydrops and heart rate was in the 60 s and regular. On delivery, she had sinus bradycardia of about 54 beat/min and severe spongioform cardiomyopathy. Transvenous pacing was not stimulated ventricular myocardium. Bradycardia and poor cardiac function lead to death within the first day. Three had isolated sinus bradycardia with a mean heart rate of 52, 54, and 58 beat/min. Two patients had AV block; one had intermittent first-degree, and Mobitz type II heart block with a heart rate of 45 to 55 beat/min. Pacemaker was implanted but he had died due to heart failure awaiting heart transplantation. The other one admitted with heart failure and bradycardia, and had complete A-V block and junctional rhythm with a ventricular rate of 50 beat/min. She had favorable two-year follow-up with an epicardial pacemaker. One patient had supraventricular tachycardia secondary to Wolf–Parkinson–White syndrome in whom tachycardia attacks was controlled with sotalol. Cardiac rhythm abnormalities are seen more commonly in patients with IVNC and ICD may be the life-saving mode of treatment in patients with refractory ventricular arrhythmias.

P33

Transesophageal electrophysiologic study in children

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Transesophageal electrophysiologic study (TEEPS) is a semi-invasive method of atrial stimulation and recordings. The aim of the study is to report our experience about TEEPS in children. A total of 67 TEEPS's were performed in 64 consecutive patients aged between 6 months to 18 years (mean 10.2 years) with the following indications; evaluation of palpitation, syncope and palpitation-syncope – 51 patients, risk assesment of Wolf Parkinson White syndrome (WPW) – 11 patients, follow-up of radiofrequency ablation procedure (RFA) – 2 patients and control of antiarrhythmic therapy effectiveness – 3 patients. Four patients had structural abnormality of heart. Standart electrocardiogram was obtained from all patients – 52 patients were normal, 12 patients had WPW syndrome pattern. Holter monitoring was performed in 54 patients-two of them had SVT, three of them had SVE. Exercise testing was performed in 47 of 64 patients – all were normal. In 1 patient who had WPW syndrome pattern had lost preexcitation during exercise testing. During procedure, a quadripolar electrode catheter was introduced through the nose into the esophagus of each patient and fixed in a position where the simultaneous recording of intraesophageal unipolar electrocardiographic derivations showed the greatest P wave potentials. A similar pacing protocol was performed in each patient. The protocol consisted of; (1) single and thereafter double extrastimuli at progressively closer intervals after an 8 beat pacing train at 500 and 430 msec cycle lengths and (2) incremental atrial pacing to the point of second-degree atrioventricular block. If tachycardia was not initiated, protocol was repeated after isoproteranol infusion. Tachycardia was initiated in 28 of 67 studies; 18 (27%) patients were AVRT, 8 (12%) patients were AVNRT, 1 (1.5%) patient was EAT and 1 (1.5%) patient was SNRT. Transesophageal atrial stimulation is effective in the initial evaluation and management of patients with arrhythmia and useful for investigation of palpitations, syncope, palpitation-syncope, follow-up of RFA, risk assesment of WPW syndrome and control of antiarrhythmic therapy effectiveness in children.

P34

Implantable Cardioverter-Defibrillators (ICD): mid-term results in children and adolescents

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Objectives: Since the introduction of ICD-therapy at the end of the eighties there has been a rapidly increasing experience with this treatment in the adult population. However experience with this treatment is still limited in the pediatric age group. The purpose of this retrospective study was to analyse and evaluate our results of ICD-therapy in children and adolescents.
Material and Methods: Between March 1998 and July 2003 14 patients underwent ICD-implantation in our tertiary referral centre. The mean age at implantation was 15.6 years (age range 10–21 years). The underlying cardiac disorders included primary electrical abnormalities in 6 patients (long QT-syndrome in 5 patients, idiopathic ventricular fibrillation in 1 patient), primary structural abnormalities in 6 patients (dilatated cardiomyopathy in 5 patients, hypertrophic cardiomyopathy in 1 patient), congenital heart disease (pulmonary atresia with ventricular septal defect after Rastelli repair in 1 patient, mitral valve prolapse in 1 patient). According to the ACC/AHA-Guidelines 8 patients had class I, 6 patients class IIb-indications for ICD-therapy. In all patients the clinical arrhythmia was inducible by programmed ventricular stimulation during electrophysiological testing. All patients received a transvenous ICD-system. VVI-ICD in 5 patients, DDD-ICD in 9 patients. We implanted 5 GEM DR II, 2 GEM I, 3 Marquis DR, 2 Micro Jewel (Medtronic Inc.) and 1 Ventak Prizm VR, 1 Vitaly DR (Guidant Inc.)
Results: The mean follow up was 32 months (range 3 to 65 months). During this period we observed neither severe complications nor mortality. There were no infections related to the device, thromboembolic complications or lead-perforations. 3 patients received appropriate DC-Shocks (21%), 1 patient received an inappropriate DC-Shocks (7%) 11 patients (79%) had no recurrence of their clinical arrhythmia under medical therapy. 3 patients (21%) needed a replacement because of lead-dysfunction. In 2 patients with DCM the device was explanted at the time of orthotopic heart transplantation.
Conclusions: According to our results progress in device- and lead-technology has resulted in a significant decrease of complications in paediatric ICD-therapy. Nevertheless a strict patient selection, using the ACC/NASPE-guidelines, is recommended.

P35

Ventricular arrhythmias in infants

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The aim of the study was to evaluate clinical presentation, long term follow-up (f-up) and prognosis in 16 patients (pts) with normal QTc and significant ventricular arrhythmias (VA) diagnosed between 1 day and 11 mo of age. In 5 pts (31%) arrhythmia was suspected prenatally, and VA confirmed by ecg at first day of life. In Holter-ecg VA presented 0,1%–55% (m = mean 18%) of total QRS/24 h. Single VEB had 8 pts, VEB pairs – 2 pts, nsVT with mean rate 202/min – 6 pts (37,5%). VA in 81%pts had LBBB morphology. At initial examination 6 pts had no obvious pathology of the heart. Because of clinically suspected myocarditis (myo) remaining 10 pts

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(62,5%) received immunosupressive (P-prednisone – 4 pts) and/or immunomodulation therapy (immunoglobuline-Ig – 4 pts, Ig + P – 2 pts), including 4/5 newborns with arrhythmia diagnosed prenatally. During 2,5–16,3 yrs (m = 8 yrs) of f-up VA disappeared in 13 pts (81%), in 69% during first 6 mo. In 2 pts (12,5%) after >5 yrs of f-up single VEB progressed to complex VA. Antiarrhythmic drugs (AAD) received 11 pts (69%), in 9 pts AAD were stopped in 1 year of life when VA disappeared. At the end of f-up pts without VA had normal x-ray, echo, signal averaged ecg, ventricular ejection fractions (EF) in radionuclide angiography. Endomyocardial biopsy (EMB) and magnetic resonance (MRI) were performed at age 10 yrs in 2 pts with progressive VA. First pt with idiopathic simple VA in infancy had MV prolapse and normal EMB in late f-up, but MRI suggested fatty tissue in RV wall. Second pt after neonatal myo treated with steroids had during f-up persistent abnormal ST-T in ecg, clinical symptoms of cardiomyopathy (abnormal LV size/function, II of mitral insufficiency) and immunosuppresive therapy was instituted because biopsy proved persistent myocarditis.
Conclusions:

1. VA diagnosed <1 year of life in 1/3 of pts were recognized prenatally.
2. Significant VA in newborns and neonates probably appeared during a course of myocarditis in 62,5% of pts.
3. Prognosis of VA in newborns and infants seems to be good because in 81% of pts arrhythmia disappeared.
4. In small percentage of pts (12,5%) progression of VA was observed.

P36

Efficacy of telemonitoring with patient-activated external loop recorders in children with transient episodes of arrhythmias

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Background: Establishing a firm electrocardiographic diagnosis for transient episodes of arrhythmias in pediatric patients is difficult, because they may disappear before an ECG is obtained. Patient-activated external loop recording (ELR) permits the storage of ECG at the time of symptoms and avoids the disadvantage of subcutaneous device implantation.
Aim of the Study: To evaluate the efficacy of ELR in establishing an electrocardiographic diagnosis in pediatric patients with transient symptoms of arrhythmia.
Methods: Prospective observational study of pediatric patients examined for 4 weeks with an ELR (Eventfoon or Holterfoon, Hartis, IMA B.V.) All patients were referred to our pediatric cardiology center in order to evaluate unexplained symptoms of arrhythmia. The ECG's stored at the time of symptoms were analyzed for presence or absence of arrhythmia.
Results: 34 patients (1.1 to 15.8 years of age, median 12.2) were included in the study. The heart was structurally normal in 32 patients. One patient had mild aortic stenosis and 1 was post arterial switch for TGA. 12-lead ECG revealed preexcitation in 5 pts, and first degree AV-block in 1 pt. Previous diagnostic tests including 12-lead ECG, echocardiography and 24-hour-Holter were not diagnostic. Indications for ELR were palpitations in 28 pts., syncope in 2 pts., and other complaints in 2 pts. During the 4-week period of loop recording 123 ECG's (median 3 ± 1 per pt.) were registered by the patients. Pathologic findings included SVT in 26% (9 pts.), asystole in 3% (1 pt.), ventricular ectopy in 6% (2 pts.). In 26% of patients the result of ELR led to therapeutic consequences during follow-up of 6.3 (median ± 5.3 months): RF-ablation in 2 pts., transvenous pacemaker-implantation in 1 pt. Five patients started drug treatment and two of them are awaiting RF-ablation.
Conclusions: Patient-activated external loop recording is an effective and convenient non-invasive tool in diagnosing transient episodes of arrhythmias in pediatric patients.

P37

Assessment of VDD/DDD versus VVIR pacing by cardiopulmonary exercise test in children and young adults

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We used a cardiopulmonary exercise test to assess the exercise tolerance during VDD/DDD versus VVIR pacing in random sequence in 13 patients (age: 14.4 ± 3.9, range 7–23 years). VDD/DDD pacemakers were implanted for atrioventricular block in 12 patients and sick sinus syndrome in one patient. The effect of pacing modes on effort tolerance has been assessed by using symptom-limited bicycle exercise protocol and gas exchange was measured on a breath by breath basis. Two exercise tests were performed on the same day: the VVIR mode and the VDD/DDD mode. Half of patients (six) were examined in the VVIR mode and the second half of the patients (seven) were examined in the VDD/DDD mode. After allowing patients to rest for over an hour, the second exercise test was implemented in other mode. VDD/DDD pacing, compared to VVIR, was associated with higher maximal heart rate (157 ± 13 vs 131 ± 18 bpm, $p = 0.000$), higher maximum oxygen consumption (VO_2 max) (23.5 ± 7.6 vs 21.8 ± 7.6 mL/kg/per min, $p = 0.028$), higher maximum work (115 ± 41 vs 105 ± 35 watt, $p = 0.007$) and longer mean exercise duration (11.5 ± 4.2 vs 10.3 ± 3.4 min, $p = 0.01$), without significant differences in mean peak minute ventilation (VE) (65.9 ± 21 vs 59.8 ± 20 L, $p > 0.05$) and exchange ratio (VCO_2/VO_2) (1.17 ± 0.08 vs 1.15 ± 0.06, $p > 0.05$). The increase in VO_2 max obtained in VDD/DDD versus VVIR was significantly related to the increase in maximal heart rate ($r = 0.708, p < 0.01$). In conclusion, VDD/DDD pacing, allowing the maintenance of atrioventricular synchrony, induces a significant improvement of exercise capacity, in comparison to VVIR pacing, related to the ability to reach higher heart rates during exercise. Atrioventricular synchrony might be useful for patients requiring physical activity.

P38

Autonomic dysfunction in children with mitral valve prolapse

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Heart rate variability (HRV) represents a noninvasive parameter for studying the autonomic control of the heart. The purpose of this study was to evaluate HRV in children with mitral valve prolapse (MVP) and to estimate the relation between HRV parameters and prevalence of ventricular arrhythmias (VA) in these patients.

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Material and Methods: 151 consecutive children with MVP (age ± SD: 12,2 ± 3,1 years) and 165 healthy subjects (age ± SD: 12,3 ± 3,7 years) as a control group were examined. HRV was assessed in both time and frequency domains using 24-hour ambulatory ECG recordings. Time domain indices included mean RR interval, SDNN, SDNNi, SDANNi, rMSSD and pNN50. Frequency domain HRV during the night and daytime was calculated using fast Fourier transformation analysis. The high frequency power (HF) and the normalized unit of the HF (HFNU) served as markers of vagal modulation. Low frequency power (LF) and the normalized unit of the LF (LFNU) were markers of sympathetic modulations. The LF/LH ratio was an index of sympathovagal balance.
Results: All the time-domain measures of HRV were significantly smaller in children with MVP than in controls. The MVP group demonstrated lower HF (daytime: $p < 0.05$) and HFNU (night: $p < 0.00002$; daytime: $p < 0.02$) values, higher LF (night: $p < 0.02$) and LFNU (night: $p < 0.00001$; daytime: $p < 0.009$) values and higher LF/HF ratio (night: $p < 0.00001$; daytime: $p < 0.0002$) than in controls. Children with MVP and ventricular arrhythmias (VA) in 24-hour ambulatory ECG recordings had significantly lower HFNU values (night: $p < 0.008$; daytime: $p < 0.005$), higher LF (night: $p < 0.02$; daytime: $p < 0.03$) and LFNU (night: $p < 0.004$; daytime: $p < 0.004$) values and higher LF/HF ratio (night: $p < 0.03$; daytime: $p < 0.02$) as compared with these without VA. None of the HRV measures was significantly related to the degree of mitral regurgitation in MVP group.
Conclusions: Our data suggest alterations in sympathovagal balance with reduced vagal and increased sympathetic activity during night and daytime in children with MVP. Increased sympathetic and reduced vagal tone is associated with higher prevalence of VA in these patients.

P39

Brady-arrhythmias in anorexic adolescent patients

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Mortality in Anorexia Nervosa (AN) is suggested to be associated with cardiac arrest. Specification of ECG abnormalities and their relationship with clinical characteristics facilitates risk assessment and provide clinical guidelines. Therefore a retrospective (n = 92) and prospective (n = 39) study in adolescent AN patients was performed. Included are clinical characteristics, laboratory parameters, 12 leads ECG, 24 hrs ECG and 2D Doppler echocardiogram. A control group consisted of 166 healthy girls. Linear regression analysis was used to describe the relationships between QT interval and heart rate.

95% of the patients were female, mean age was 15,6 ± 1,9 years, and mean Body Mass Index was 15,1 ± 1,4 kg/m^2. ECG showed bradycardia 56 ± 14 bpm (29–100 bpm) ($p < 0,0005$), right deviated QRS axis 74 ± 26° ($p = 0,01$) and a lower amplitude of the R wave in lead V6 0,9 ± 0,4 mV ($p < 0,0005$). ECG abnormalities included supraventricular arrhythmias, AV nodal blocks and repolarisation disorders. The QT interval was in 19% prolonged over the 95% confidence interval. Exercise abuse resulted in an even lower heart rate and prolonged QT interval. Mean minimum heart rate (HR) on the 24 hrs ECG was 37 ± 6 bpm (26–189 bpm). Rapid weight loss correlated with pronounced bradycardia ($p = 0,001$).

Conclusion: Although adolescent AN patients have potentially life-threatening bradycardias and prolonged QT intervals. ECG and Holter-recordings showed only various benign supra-ventricular rhythm disturbances and one non-sustained VT (3 beats). A high rate of weight loss contributed to a sharp decrease of the HR. Despite these findings, no mortality or life-threatening events were encountered. Based on these data no cardiac intervention in these patients seems to be warranted, in extreme bradycardic situations (forced) refeeding seems to be the most plausible solution.

P40

Experience with permanent pacing in cTGA patients

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Congenitally corrected transposition of great arteries (c-TGA) is a rare cardiac defect. The lack of trabecular network of venous ventricle, improper axial lead pressure and associated anomalies would not provide good stability for endocardial lead. For this reason most of the centers prefer to use pericardial leads. Progressively increasing threshold make late epicardial failure more common.

The aim of the study was to analyses the problems with permanent cardiac pacing in c-TGA children. Between 1980–2003 in 19 children (13 boys and 6 girls) with c-TGA pacemaker was implanted. Associated anomalies were found in 11 children: VSD – 9, ASDII-7, PS – 8, functionally single ventricle – 3, systemic valve atresia – 2, dextrocardia – 3, dextroversion – 1. Ten children had congenital complete atrio-ventricular block, 7 – post cardiac surgery complete or Mobitz II av block, 2-had acquire complete av block. The age at initial implantation range from 2 weeks to 18 years (mean 7.5 yr). Follow up period range from 6 weeks to 14 years (mean 6.5 yr). There were 3 deaths: 2 due to heart failure, 1 was sudden. Two patients were lost for follow up. For the first implantation 15 children received epicardial system: VVI – 3, VVIR – 8, DDD – 4, endocardial system was used in 4 pts: VVI – 1, VVIR – 1, DDD – 2. During the implantation endocardial approach was failure to attempt in 2 pts, because of initially high threshold and problems with lead stability. In one child endocardial DDD system was implanted successfully, but ventricular screw-in lead displaced after 12-hours. There were 7 reoperation in 4 children due to leads malfunction. The high threshold (4,8–6V) was in 4 pts, and there were 3 lead fractures.

Endocarial lead implantation in cTGA patients is difficult, but possible. High threshold and lead fracture are the main reason for reoperation.

P41

Slow pathway modulation: A safe and effective therapy of AV-nodal-reentry-tachycardia (AVNRT) in children and adolescents

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Objectives: AVNRT is the second most common type of supraventricular tachycardia in children and adolescents. The principles of dual AV nodal pathways and AVNRT have been understood for many years although the anatomical correlates remained unclear

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until recently. The posterior extension of the compact AV nodal structure seems to be the "slow pathway" region. This part of the AVNRT circuit is situated within the right atrium and can be ablated or modulated without risk of damaging the compact AV node itself. Radiofrequency modulation of this area was demonstrated to prevent the initiation of AVNRT without destroying the slow pathway. We report our experience with this treatment in the pediatric age group.

Material and Methods: Indications for electrophysiologic testing and slow pathway modulation in our institution included a history of several episodes of SVT refractory to appropriate medical antiarrythmic treatment and a minimum age of 5 years. After basic electrophysiologic testing with demonstration of dual pathway characteristics of the AV node (AH jump, AVNRT initiation) a combined anatomical and electrocardiographic guided approach was used to modulate the slow pathway. The endpoint for successful modulation was the inability to induce AVNRT both before and during orciprenalin infusion.

Results: During the period between 1997 and 2003 36 pediatric patients underwent slow pathway modulation in our tertiary referral centre. The mean age was 12.7 years (age range 5.6–22.6 years). The "typical" slow–fast AVNRT was seen in 31 patients, fast–slow AVNRT in 4 patients, slow–slow AVNRT in 1 patient. The mean follow up interval was 3.0 years (0.4–5.8 years). The primary success rate after the 1st procedure was 91%, in 3 patients (9%) AVNRT relapsed. A 2nd procedure was successful in both patients who underwent repeat modulation. There were no complications as AV-Block or vascular problems during the procedure and follow-up period.

Conclusions: According to our experience slow pathway modulation is a safe and effective treatment for AVNRT in children and adolescents. In experienced centres it should be associated with minimal risks and therefore offers an attractive alternative to long-term antiarrhythmic drug therapy.

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Ventricular preexcitation associated with dilated cardiomyopathy: a causal relationship?

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Background: In patients with Wolff Parkinson White (WPW) syndrome, dilated cardiomyopathy (DCM) is usually the result of recurrent, sustained tachyarrhythmic episodes. We report the association of WPW syndrome and DCM in the absence of tachyarrhythmias.

Patients: Four children (3 boys), aged between 12 months and 13 years, presented to one of the above institutions. Presenting complaints were failure to thrive associated with a systolic murmur (n = 2), isolated systolic murmur (n = 1), first episode of supraventricular tachycardia (n = 1). All patients had WPW syndrome on standard ECG, with the delta wave axis suggestive of a right sided pathway. All had evidence of DCM, with left ventricular end-diastolic diameter >97th centile for weight, and shortening fraction of between 13% and 25%. Metabolic screening, viral studies and other investigations to define the possible cause of DCM were negative. Three of the 4 patients had undergone annual ambulatory Holter recordings, without any episode of tachyarrhythmia being recorded.

Follow-up: During a follow-up of between 3 and 66 months, 2 patients underwent diagnostic catheterization and biopsy which were negative. Two patients underwent RF catheter ablation of accessory pathway, with normalization of myocardial function following the procedure. Another patient had spontaneous disappearance of preexcitation, with resolution of DCM. One patient is awaiting ablation.

Conclusions: Reversible DCM is associated with manifest ventricular preexcitation, even in the absence of sustained arrhythmias.

P43

Cardiac dysrhythmias after transcatheter closure of ASD with Amplatzer device

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Transcatheter closure of ASD has been as an alternative method to open heart surgery. Although, transcatheter closure of ASD with the Amplatzer septal occluder is a safe and feasible method in pediatric patients, there is little published data on arrhythmia analysis following transcatheter device closure of secundum ASD. We evaluated cardiac dysrhythmias with 24 hour Ambulatory ECG monitoring after transcatheter closure of ASD with Amplatzer device.

A total of 85 consecutive patients with ASD underwent transcatheter closure of secundum ASD with Amplatzer device between October 1998 and December 2003. The study involved 65 of these patients assessed by 24 hour ambulatory ECG monitoring. Seven patients were reevaluated by a second time 24 hour monitorization.

We didn't observe rhythm problem in ECGs before procedure. During procedure, transient complete AV block was seen in 2 patients. One of them returned to normal sinus rhytm in cath lab and the other returned to normal sinus rhytm in two hours. Transient junctional rhythm was observed in another patient during the device placement. Ambulatory Holter montorization was performed on all patients after a mean 4 months period (1–12 months). There was no rhytm abnormalities in ECGs after ASD closure. Holter recordings demonstrated rare supraventricular extrasystole in 2 patients, rare ventricular premature beats in 2 patients and intermittant sinus arrest with sinus pause, lasting <1.5 seconds was seen in 1 patient, with total of 5 patients (7.6%).

In conclusion, dysrhythmias after transcatheter device closure of secundum ASD with Amplatzer device are rare and benign. We need further long term follow-up for regarding late dysrhythmias after the transcatheter device closure of secundum ASD.

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Heart rate variability in children with congenital sensourineural deafness

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In our recent study consisting of approximately 800 children we found that there were no major abnormalities for repolarization parameters (QT/QTc duration, and QT/QTc dispersion) in children with congenital sensourineural deafness, when compared to hearing counterparts, if heart rates were similar. The completely unexpected result of that study was the finding of the significantly lower mean heart rate in congenitally deaf children. Decreased sympathetic tone as a result of the absence of auditory stimuli on autonomic nervous system in deaf children might be an explanation. To test that hypothesis, we obtained time- and frequency-domain heart rate variability (HRV) data from 24-hour Holter

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ECG recordings of 23 congenital sensourineural deaf children (male/female: 13/10, mean age: 12,7 ± 2,4) and compared them to those of age and sex matched 18 healthy school children (male/female: 11/7, mean age: 12,1 ± 4,2). HRV was measured by calculating time-domain and frequency-domain indexes from 24-hour recordings and from 6 hours of recordings obtained while subjects were sleeping (between midnight and 6 AM). Six time domain indices were examined: (1) Mean RR: mean of all normal sinus R-R intervals, (2) SDNN: standard deviation of all normal sinus R-R intervals over 24 h, (3) SDNN-i: mean of the standard deviation of all normal sinus R-R intervals for all 5-min segments, (4) SDANN: standard deviation of the averaged normal sinus R-R intervals for all 5 min segments, (5) rMSSD: root mean square of the successive normal sinus R-R interval difference, (6) pNN50t: percentage of successive normal sinus R-R intervals longer than 50 ms. Three frequency-domain indices were examined: (1) LF: low frequency index, (2) HF: high frequency index, (3) LF/HF ratio. There were no significant differences for either time- or frequency-domain HRV parameters between deaf children and healthy counterparts (Table). Since a 24-hour and 6-hour sleeping HRV data were highly correlated for all measurements we only presented 6-hour sleeping HRV data in Table.

Variables	Deaf children (n:23)	Control group (n:18)	P value
mean RR	887,48 ± 113,0	874,22 ± 181,32	0,77
SDNN	117,08 ± 45,32	117,25 ± 45,65	0,99
SDNN-i	97,47 ± 41,73	99,89 ± 41,92	0,85
SDANN	50,70 ± 21,05	47,45 ± 19,50	0,61
rMSDD	86,61 ± 51,90	93,70 ± 53,19	0,67
pNN50t	38,36 ± 21,49	40,03 ± 23,60	0,81
LF	27,38 ± 11,60	28,30 ± 12,47	0,80
HF	27,88 ± 14,12	28,79 ± 15,87	0,84
LF/HF	1,11 ± 0,39	1,08 ± 0,36	0,85

HRV parameters of deaf children and control group.

We conclude that, although our previous study indicated that decreased sympathetic activity or increased vagal tone might be responsible for lower mean heart rate; autonomic dysfunction does not seem to be present in patients with congenital sensourineural deafness.

P45
Acute haemodynamic compromise due to sustained arrhythmia after atrial septal device insertion

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Aim: Transient atrial arrhythmias are commonly encountered after the insertion of devices to close inter-atrial communications and are not thought to be important. We wish to describe our experience of significant arrhythmias in this situation.
Patients and Methods: Retrospective review of the cardiology database revealed 5 patients who sustained significant arrhythmias after atrial septal device insertion out of a total population of over 300. There were 2 children aged 11 days and 14.6 years, the former had insertion of a 9 mm Amplatzer septal occluder (ASO) for continued heart failure after coarctation repair and the latter insertion of a 25 mm Amplatz PFO occluder for transient ischaemic attacks in the setting of Ebstein's anomaly. There were 3 adults aged 26, 27 and 36 years. One had insertion of a 38 mm ASO, 1 had insertion of a 35 mm PFO occluder for desaturation in pulmonary atresia with intact ventricular septum and 1 had a custom made fenestrated device for a hypertensive left atrium in the setting of double inlet left ventricle and dual chamber pacing.
Results: The patient with Ebstein's anomaly had a sudden increase in medically refractory AVRT and went on to have unexplained sudden death. The remaining 4 patients developed atrial tachycardia. In the patient with DDD pacing, the atrial tachycardia conducted 1:1 as the mode switch had not been activated. This was not tolerated and resulted in VF arrest. The neonate who had ASD occlusion remained in a low cardiac output state for 2 days until successful DC cardioversion after loading with amiodarone. The adult who had ASD occlusion collapsed with broad complex tachycardia after the administration of intravenous flecainide and required resuscitation. The remaining patient has had arrhythmia requiring frequent DC cardioversion in addition to 2 ablation attempts.
Conclusions: Significant and potentially life threatening arrhythmias do occur after closure of inter-atrial communications presumably due to alterations in atrial geometry and dynamics. In our experience there were 5 patients including 2 requiring resuscitation and 1 sudden death. Before such procedures careful consideration has to be made of the underlying anatomical substrate, patients must be warned and any sustained atrial arrhythmia promptly treated.

P46
Catecholaminergic ventricular tachycardia: a primary indication for implantable intracardiac defibrillator (AICD)

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Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an uncommon arrhythmia in children and adolescents with a structurally normal heart. CPVT patients are exposed to syncopal events and sudden cardiac death due to stress-induced ventricular tachyarrhythmia. We report 2 such patients.
Case 1: An 8.5-year-old boy developed syncope a 1/2 hour after playing football. Personal and family history was non-contributory. Physical examination, ECG, QTc and chest X-ray were normal. Doppler/echocardiography examination revealed no abnormality. Signal averaged ECG detected no late potentials. Holter examination showed bi-directional ventricular ectopy, short run of polymorphic VT at higher heart rate, also evident during exercise testing. Cardiac CT scan showed no fatty infiltration of the right ventricle. Nadolol therapy effectively diminished the degree of ectopy with exercise. A de novo mutation on the cardiac Ryanodine 2 receptor gene of chromosome 1 was identified. Despite adequate beta blockade, the patient died suddenly from ventricular fibrillation at 14,5 years of age following an emotional event.
Case 2: An 11-year adolescent girl experienced syncope while swimming. Personal and family history was non contributory. Physical examination, ECG, QTc and chest X-ray were normal. Doppler/echocardiography examination revealed no abnormality. Late potentials not detected on signal averaged ECG. Holter and exercise testing identified isolated ventricular ectopy (VE) becoming multifocal followed by bi-directional ventricular tachycardia. She was treated with nadolol with marked reduction in VE and no relapse of syncope. A DDD pacemaker was implanted for bradycardia. For preventive reasons the unit was replaced 5 years later with an intracardiac defibrillator. A few weeks later, on adequate

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beta blocker therapy, whilst dancing, ventricular tachycardia occurred resulting in lifesaving delivery of burst overdrive followed by shock.
Conclusions: CPVT is reported to respond favorably to beta-blocker therapy. In these 2 patients compliance was believed good. Whilst the degree of arrhythmia may be diminished with adequate beta blockade, this disease may in some cases remain refractory and lethal. We strongly recommend implantation of an intracardiac defibrillator in these patients as additional primary therapeutic option.

Session 5: Basic Science and Genetics

P47

Differential proteomic profiling and hemodynamic characterization of the pressure overloaded right ventricle in young rats: a novel approach

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Background: RV failure is an important problem in congenital heart disease. We sought to determine the mechanisms underlying the progression from compensated RV hypertrophy to failure using an alternative approach. Therefore, we studied alterations in myocardial protein expression in relation to RV function. Here, we report the results after 6 weeks of RV pressure overload.
Methods: Male Wistar rats (age 8.1 ± 0.4 wk) underwent Pulmonary Artery Banding (PAB) (n = 6) or sham (n = 6) operation. Load-(in)dependent RV and LV function was characterized *in vivo* by means of the end-systolic pressure–volume relation (ESPVR) using the conductance catheter technique. Myocardial cytoplasmatic protein expression was determined by proteomic analysis of individual homogenates of RV myocardium (6 gels per group i.e. sham and PAB).
Results: In PAB animals the ratio of RV and RA to body weight was increased (0.58 ± 0.02 vs 1.03 ± 0.05 RV, $p < 0.001$; 0.13 ± 0.01 vs 0.21 ± 0.03 RA mg/g, $p < 0.05$). *Hemodynamics:* Hypertrophic RV end-systolic and end-diastolic pressures were increased (31 ± 1 vs 65 ± 5 and 4.3 ± 0.5 vs 10.5 ± 1.1 mmHg, $p < 0.001$). After PAB cardiac output decreased significantly, but heart rate was unchanged. Load-dependent indices of RV systolic performance (dPdtMax and Stroke Work) increased significantly ($p < 0.001$) as a result of PAB, as well as the load-independent slope of the ESPVR (0.09 ± 0.03 vs 0.29 ± 0.02 mmHg/μL, $p < 0.001$). Indices of LV function were not affected. *Proteomics:* Several stress- and metabolic proteins significantly differing in intensity were identified. A general downregulation was observed in beta-oxidation pathway proteins. Glycolytic pathway proteins showed an increased expression. Three spots were identified as HSP27 (+1.7–3.5 (in PAB), $p < 0.05$), suggesting HSP27 modifications.
Conclusions: Our results indicate that 6 weeks of PAB leads to compensated RV hypertrophy in rats. Despite the enhanced RV contractile state, cardiac output falls, suggesting initial transition from compensated state to failure. In these conditions, specific alterations and modifications in metabolic and stress related protein expressions were present in the hypertrophic RV. We expect that alterations in myocardial protein expression (in various subfractions) associated with the progression to RV failure will be disclosed in further studies.

P48

The methylenetetrahydrofolatereductase genotype as a risk factor for congenital heart defects

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Background: The etiology of congenital heart defects (CHD) has been partly illuminated, whereby genetic and environmental risk factors play a role. Because of the involvement of neural crest cells it can be hypothesized that the pathogenesis of CHD is similar to a pathophysiological mechanism of neural tube defects (NTD). Hyperhomocysteinemia, folate deficiency and mutations in methyleentetrahydrofolatereductase (MTHFR)-gene are related to higher risk of NTD in offspring. In a pilot study Kapusta et al (Pediatrics 1999) maternal hyperhomocysteinemia seemed to be a risk factor for CHD.
Objectives: We studied the MTHFR 667 C–> T polymorphism in children as a genetic risk factor for CHD, both in neural crest related and non-neural crest related heart defects.
Methods: We included 128 children with a CHD. The control group consisted of 218 children. The children with a CHD were recruited through the list of children admitted for a cardiac intervention in our hospital and invited to take part in this study. The control group was recruited in a normal population. One venous blood sample was taken for DNA extraction for genotyping the MTHFR 677 C–> T polymorphism.
Results: Of these 128 children 101 (79%) had a neural crest related heart defect and 27 (21%) had a non-neural crest related heart defect. The Odds Ratio for MTHFR 677TT genotype was 2.1 (95% Confidence Interval 1.0–4.3) for the whole group. For the neural crest related and non-neural crest related heart defects the calculated Odds Ratios for MTHFR 677TT genotype were 2.2 (95% CI 1.0–4.7) and 1.7 (95% CI 0.5–6.3), respectively.
Conclusions: The point-estimates of the risk for CHD due to MTHFR 677TT genotype are in line with the hypothesis that the MTHFR 677TT genotype is a risk factor for congenital heart defects, particularly for the neural crest related heart defects.

P49

The NADPH oxidase subunit p22phox is regulated by a redox-sensitive pathway in endothelial cells

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Endothelial dysfunction, hypercontraction and proliferation of vascular cells are associated with pulmonary hypertension. Endothelial dysfunction is characterized by increased levels of reactive oxygen species (ROS) and decreased NO availability. NADPH oxidases have been reported to contribute to ROS production in the endothelium and to be associated with endothelial dysfunction, hypercontraction and increased proliferation. However, the mechanisms regulating ROS production by this enzyme complex in the endothelium are not well understood. Since ROS generation can occur

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rapidly within minutes but also delayed within hours of stimulation, we investigated whether ROS play a role in the regulation of the NADPH oxidase in endothelial cells.

Two photon confocal microscopy showed that ROS generation was closely related to the expression of the NADPH oxidase subunits p22phox and gp91phox coupled to an enhanced cyan fluorescent protein. Thrombin stimulation resulted in a rapid, but transient increase in ROS generation peaking within 15 minutes, but also in a delayed elevation of ROS production which was maximal after three hours of stimulation and which was accompanied by an upregulation of p22phox, but not gp91phox protein levels. These responses were abrogated by treatment with antioxidants suggesting the involvement of ROS. Increasing ROS levels by exogenous application of H_2O_2 or endogenously by overexpression of superoxide dismutase elevated p22phox, but not gp91phox protein levels in a time- and dose-dependent manner. Stimulation with H_2O_2 activated ERK1/2, p38 MAP kinase (p38MAPK) and protein kinase B (Akt). Upregulation of p22phox protein levels and ROS production in response to H_2O_2 were dependent on p38MAPK and the PI3 kinase/Akt pathways. Finally, low concentrations of H_2O_2 or overexpression of p22phox stimulated the proliferation of endothelial cells.

These data show that p22phox, but not gp91phox protein levels are regulated in a redox-sensitive manner in endothelial cells. ROS generated by rapid activation of the NADPH oxidase may further lead to the induction of this enzyme resulting in the prolonged elevation of ROS which may be responsible for endothelial dysfunction and enhanced proliferation of vascular cells in pulmonary hypertension.

P50

Human vascular cells express a distinct NOX5 isoform: Role in the generation of reactive oxygen species and the proliferative response

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Pulmonary hypertension is frequently associated with vascular remodeling linked to intimal and medial thickening and proliferation of vascular cells. Recently, reactive oxygen species (ROS) have been implicated as signaling molecules triggering cellular proliferation in response to a variety of stimuli. Vascular ROS production has been mainly related to the activation of a family of enzymes termed NADPH oxidases. Originally, an NADPH oxidase consisting of at least four cytosolic factors (p40, p47, p67, Rac) and two membrane-bound subunits, p22 and gp91, has been described in neutrophils being responsible for the respiratory burst. Meanwhile, several NADPH oxidase variants have been identified, which express different isoforms of gp91, termed NOX1 to NOX5. Whereas a role of NOX1 and NOX4 in vascular ROS production has been described, the role of NOX5 in vascular cells is unclear. Several NOX5 isoforms have been identified in lymphocytes, spermatozoa and pancreatic cells, producing ROS in a calcium-dependent manner. We therefore aimed to identify and characterize NOX5 in the vasculature. Using a RT-PCR approach we cloned NOX5 as well as a NOX5 splice variant lacking exon 10 thus leading to a premature stop codon predicting a truncated protein in vascular smooth muscle cells. Both variants were also present in endothelial cells. In contrast to the previously described NOX5 isoforms, the vascular isoforms did not contain calcium-binding elements in their 5′end. Using two photon confocal microscopy we found intracellular, calcium-independent ROS production closely associated with NOX5 linked to a cyan fluorescent protein (CFP), which was 7-fold enhanced compared to the NOX5 variant or control vector. Both NOX5 and the NOX5 variant were thereby localized around the nucleus. Moreover, vascular cells overexpressing NOX5 showed enhanced proliferation. This response was not observed in cells expressing a NOX5 antisense vector or the NOX5 variant.

These findings show that in vascular cells a functional, but distinct NOX5 isoform is expressed which is able to induce ROS production and proliferation in a calcium-independent manner. Thus, NOX5-dependent ROS production may contribute to the proliferation of pulmonary vascular cells, whereas co-expression of an inactive NOX5 variant may have an inhibitory function thus limiting vascular remodeling processes.

P51

Assessment of association of variants and haplotypes of the remaining TBX1 gene with manifestation of congenital heart defects in monosomy 22q11.2 patients

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Objectives: TBX1 is a strong candidate gene for outflow tract heart defects in monosomy 22q11.2 patients. It is located within the DiGeorge/velocardiofacial syndrome region in 22q11.2. Although a common trait, the manifestation of congenital heart defects (CHD) in patients with this microdeletion is variable. We investigated, if mutations, variants or common haplotypes within the remaining TBX1 gene could modify the expression of the heart phenotype in patients with monosomy 22q11.2.
Methods: We sequenced the coding region of TBX1 in a total of 174 patients with 22q11.2 deletion, 123 of whom suffered from CHD (tetralogy of Fallot [51 patients], pulmonary atresia with ventricular septal defect (VSD) [22], VSD [17], interrupted aortic arch type B [16], truncus arteriosus communis [11], double aortic arch [2], double outlet right ventricle, cervical aortic arch, atrial septal defect and aortic valve aplasia with VSD [1 patient each]), while 51 did not have CHD. Our normal control group consisted of 96 healthy, dizygous individuals of Caucasian origin.
Results: Only in one patient with CHD, a 9 bp deletion mutation, which was neither detected in non-CHD patients nor in normal controls, was found. This mutation was inherited from the patient's father who also had a VSD but no 22q11.2 deletion and might therefore constitute a rare causative factor for CHD. The same might apply for a 321G-A (V107V) variation in one patient with pulmonary atresia-VSD, which was not detected in normal controls and which was the only variant to affect a conserved nucleotide. It might therefore also represent a relevant mutation, as synonymous changes can effect splicing. We detected 14 further sequence variants, 7 of which turned out to represent common single-nucleotide polymorphisms. None of these 14 variants affected a conserved nucleotide nor showed a significant difference in frequencies between 22q11.2 deletion patients with and without CHD or normal controls.
Conclusions: Our results exclude common TBX1 variants as modifiers of expression of congenital heart defects in 22q11.2 deletion

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patients. Even if the 9 bp deletion and V107V, respectively, are considered relevant contributors to the expression of CHD, functional studies are necessary to confirm the impact of these rare variants.

P52

NT-proBNP serum concentrations in the umbilical cord blood and in healthy neonates and children

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Objective: N-terminal brain natriuretic peptide (NT-proBNP) is a neurohormonal substance secreted mainly by the cardiac myocytes of the ventricles and to a less degree of the atria. The releasing mechanism is induced by an increased wallstress on the base of volume overload and/or pressure load of the ventricle. Its physiologic actions are prohibition of the sympathetic activity, vasodilatation, natriuresis, diureses and inhibition of the renin angiotensin system. Because of its high sensitivity and specifity in particular in relation to the left ventricular volume and pressure load BNP as well as NT-proBNP are well accepted markers of ventricular failure. It was the aim of the study to establish normal serum concentrations of NT-proBNP throughout childhood, in particular in the newborn age, by means of an immunoassay.
Methods: In a cross sectional study serum NT-proBNP concentration was measured by ElectroChemiLuminescenceImmunoAssay ('ECLIA' Roche) in the arterial and venous umbilical cord blood of 50 healthy term neonates and in 150 infants and children from birth to the age of 16 years.
Results: The concentration of NT-proBNP in the cord blood samples ranged from 280 to 3164 pg/ml; there was a marked increase on the day of delivery to levels about 2–3 times and a maximum on day 3 (621–8122 pg/ml). There followed a decrease during the 1st year of life after which the levels ranged at a constant niveau from 5–159 pg/ml up to the age of 16 years, which is comparable to that of healthy adults.
Conclusions: Within the family of the cardiac peptides NT-proBNP has been shown to represent a high sensitivity and specifity in the setting of ventricular dysfunction. The marked elevated serum concentrations during the change of perinatal circulation represent the ventricular volume and pressure load in the newborn period. The normal range of NT-proBNP levels in healthy subjects throughout childhood serve as a non-invasive marker for discrimination from pathologic conditions like acute myocarditis, hypertrophic or dilated cardiomyopathy, tachyarrhythmias and pulmonary hypertension.

P53

Decreased in vitro angiogenesis in patients after Kawasaki disease

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Kawasaki disease is a systemic vasculitis including coronary vascular systems. Giant coronary aneurysms may thrombose and lead to sudden death. Fatal cardiac events due to thrombotic occlusion occur mostly in the acute or subacute stage of illness. Angiogenesis is essential for survival as well as for vascualr repair; however, angiogenic capability of these patients remains largely unknown. We examined in vitro capillary tube-forming capability of human umbilical vein endothelial cells (HUVEC). Growth factor-reduced Matrigel was placed in the wells of plate and incubated at 37°C for 30 min to allow polymerization. HUVEC were treated with either patient or healthy control serum, plated onto Matrigel-coated wells, and incubated at 37°C for 18 hours. The tube formation was quantified by measurement of tube length. The gamma globulin and low dose aspirin did not affect the assay used. HUVEC formed extensive capillary-like tube structures in the presence of control serum, but partially failed to form them when treated with serum from patients who later developed coronary aneurysms. The maximum inhibition ($p < 0.01$) was obtained in the presence of patient serum at 1 to 6 months after aneurysm formation, which can be partially rescued by exogenous addition of growth factors. Furthermore, patient serum did not affect HUVEC proliferation and migration, suggesting that patient serum may affect the level of HUVEC differentiation. This impairment of in vitro angiogenesis has naturally resolved after 1 year of disease onset. These data suggest that the impairment of angiogenesis may be found in patients in the acute or subacute stage of Kawasaki disease. This may explain the higher rates of fatal cardiac events in the acute or subacute stage of Kawasaki disease.

P54

Deep hypothermia suppresses inducible nitric oxide synthase and inflammation in microglial cells

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Background: The pathomechanism underlaying the protective effect of deep hypothermia during cardiac surgery in children is unclear. Experimental settings of cardiac surgery have confirmed the involvement of astroglial rather than neuronal cells. Thus, changes in cellular signals including extracellular inflammatory cytokine and NO-metabolism were studied on lipopolysaccharide (LPS) stimulated BV2 microglial cells during dynamic temperature changes mimicking conditions of hypothermic cardiopulmonary bypass perfusion in vivo.
Methods: BV2 microglial cells were used in a specially designed incubator allowing dynamic continuous changes of temperature. The sequence of events was designed as follows: normothermia, rapid cooling (2.8°C/min), 60 min of stable deep hypothermia (17°C) followed by rewarming (3.0°C/min) and subsequent 24 h of normothermia (37°C). Extra cellular NO content (Griess Assay), IL-6 and TNF-a (ELISA) and intracellular iNOS-expression (Western Blots) were measured at 8 standardized time intervals.
Results: 188 cultures were studied in four independent experimental settings. The extra cellular concentration of NO release in the normthermic BV2 cell control-group increased after stimulation with LPS (1 μg/ml) from 0.5 μg/ml at baseline up to 40 μg/ml at the end of 24 h of normothermia. The experimental group under temperature changes provided significantly lower levels of NO (10 μg/ml) after 24 h of normothermia (37°C). Inflammatory cytokine release such as IL-6 and TNF-a were significantly suppressed under hypothermic temperature changes. Western Blot analysis demonstrated a markedly reduced time- and temperature dependent expression of iNOS under hypothermic conditions.
Conclusions: Hypothermia during cardiac surgery is neuroprotective, possibly through suppression of microglial activation and inflammation.

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P55

AMPKα2- but not AMPK α1 or adiponectin receptor mRNA is downregulated in neonatal cardiac myocytes cultured in high glucose medium compared to native neonatal cardiac tissue

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Objectives: Manipulation of myocardial energy metabolism appears to be a feasible therapeutic approach in chronic heart failure, ischemic heart disease and possibly also in children with congenital heart disease. Various metabolic drugs are under experimental and clinical investigation. To facilitate metabolic therapy, detailed knowledge of cardiac energy metabolism is necessary. The aim of this study was to investigate the expression patterns of key regulatory enzymes in the metabolic pathway under different physiological and artificial conditions with a focus on neonatal cardiac development.
Methods: mRNA expression of the catalytically active α-subunits (α1 and α2) of the AMP-activated protein kinase (AMPK) – a key sensor of cellular energy status – and of the peroxisome-activated receptor α (PPARα) – a nuclear receptor acting as a transcriptional activator for genes involved in fatty acid oxidation – were determined by real time PCR during neonatal development (days 1, 7 and 21) in native heart tissue as well as in cultured cardiomyocytes from rats. Additionally, mRNA-expression of a recently described possible receptor for adiponectin – an adipocyte derived hormone and AMPK activator – was measured.
Results: In cultured cardiomyocytes, mRNA-expression of the α1 subunit of AMPK was unchanged compared to native heart tissue. However, mRNA expression of the AMPK α2 subunit and PPARα was significantly reduced in cultured cardiomyocytes. There were no differences in mRNA expression profiles of both α-subunits of AMPK during neonatal cardiac development, whereas mRNA expression of PPARα increased twofold at day 7. Mimicking hypoxia by addition of desferrioxamine to cultured cardiomyocytes, a compound which stabilises the HIF-factor, did not change the expression patterns of the α-subunits nor of PPARα. The expression profiles of the possible adiponectin receptor remained unchanged in all cases.
Conclusions: Cultivation of cardiomyocytes in a glucose enriched medium results in a selective downregulation of α2-AMPK mRNA expression and PPARα, whereas = α1-AMPK is unchanged. This suggests a differential regulation of α2-AMPK expression. Cardiac expression of the adiponectin receptor is not influenced by developmental changes or energy substrate supply.

P56

CVGdb and d-matrix – an approach to store and analyse clinical and molecular phenotypes of patients with congenital heart diseases

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In order to match the wealth of molecular data, the phenotypic clinical data need to be obtained and recorded to a similar degree of precision than the molecular data. To achieve this goal, the development of detailed phenotype databases has recently been brought up to the forefront of the genome research. Approaches to the genetics of cardiovascular diseases are deeply depending on detailed phenotype information regarding the underlying aetiology; anatomical, morphological and hemodynamical abnormalities and the previous clinical record.

CVGdb
Our group aims on a long term to partially elucidate the genetic network involved in the cardiac developmental process. Therefore, we developed a dedicated phenotype scheme for congenital heart diseases (CHD) that is implemented in our CardioVascular Genetic database (CVGdb) based on Oracle 8i. This scheme reflects in principle the International Classification of CHD but is structured with regard to the different cardiac segments rising at certain developmental milestones. It is composed of three major parts: general information, structural cardiovascular phenotype and characterisation of the cardiac electrocardiogram. General information comprises demographic data as well as general clinical information. The cardiovascular part is subdivided in anatomical components and covers the anatomical, morphological and hemodynamical parameters for each segment. In addition, CVGdb holds all molecular data obtained from our patients analysed in ongoing SNP/mutation screening and cDNA Array approaches.

d-matrix
As the phenotype scheme consists of 150 different attributes and 400 values for each dataset, a major challenge of the project was the development of a graphical web-based front-end, allowing a dedicated user-specific display and analysis of the stored information, satisfying the association of clinical with molecular data. Based on these needs we developed "d-matrix", a data visualisation and analysis software with a display of three dimensions in form of colour-coded boxes and bars arranged as a matrix. Furthermore, we developed d-matrix to a stand-alone software, applicable to any database, independent of the data context.

P57

Improved recovery from hypoxia of the neonatally hypoxic adult rat heart following endothelin blockade

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We previously showed that recovery of cardiac contractile function from hypoxia and coronary vasodilation on reoxygenation of the adult isolated/perfused rat heart are impaired following chronic neonatal hypoxia (1). We have asked whether endothelin blockade can improve the recovery of cardiac function from hypoxia following neonatal hypoxia. The isolated hearts of 20 adult Sprague-Dawley rats (411 ± 9 g, 88 ± 1 d (SEM)) were studied. Ten animals experienced hypoxia neonatally (F_iO_2 = 0.12, days 1–10 of life, Neonatally Hypoxic) while 10 others did not (Control). The hearts were perfused in the Langendorff mode with a Krebs solution containing the non-specific endothelin blocker Tezosentan (10-6M). Left ventricular pressure (LVP), maximum rate of pressure increase (dP/dtmax) and total coronary flow (CorFlow) were monitored continuously. The hearts were perfused with: (I) oxygenated perfusate ($95\%O_2 + 5\%CO_2$) × 30 minutes, (II) hypoxic perfusate ($10\%O_2 + 5\%CO_2 + 85\%N_2$) × 30 minutes, (III) oxygenated perfusate × 30 minutes. The results were compared to our previous work utilizing this protocol without addition of Tezosentan to the perfusate (1). At baseline there was no difference between Neonatally Hypoxic and Control groups in LVP, dP/dtmax or CorFlow. Perfusion with hypoxic perfusate led to similar decreases in LVP as well as dP/dtmax and increases in CorFlow in both groups. The LVP and dP/dtmax at baseline and during hypoxic perfusion were not different from those previously obtained in the

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absence of endothelin block. After 15 minutes of reoxygenation LVP, dP/dtmax and CorFlow returned to >90% of the baseline level in both Control and Neonatally Hypoxic groups as previously seen in Control hearts not subjected to endothelin blockade (1). In contrast we previously observed in the absence of endothelin blockade that recovery of LVP and dP/dtmax in Neonatally Hypoxic hearts remained impaired after 15 minutes (65 ± 6%, 61 ± 7% of baseline) and 30 minutes of reoxygenation (72 ± 5%, 72 ± 5% of baseline) with CorFlow decreasing to significantly less than baseline values after 30 minutes of reoxygenation (3.6 ± 0.2 vs. 4.6 ± 0.2 ml/min/g) (1). Our findings suggest that the long-lasting cardiac vulnerability to hypoxic stress after chronic hypoxia neonatally is due to augmented endothelin mediated coronary vasoconstrictor influences.

Reference:

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P58

Cardiac arrhythmia and polycystic kidney diseases – a new phenotype linked to 7q22

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We report a kindred in which members in 4 successive generations suffered from paroxysmal ventricular tachycardia associated with polycystic kidney disease.

Clinical Features: All affected family members show the presence of both clinical features. The index person was admitted to the hospital for a syncopal attack without any conditions of a long QT syndrome, but a well-recorded polycystic kidney disease. Clinical work up showed severe polymorphic and polytopic ventricular extrasystoles. Her father died suddenly at age 30, under circumstances suggesting the presence of the same disorder. Autosomal dominant polycystic kidney diseases (ADPKD) is a frequent disorder with an incidence of 1 in 1000 but has not been reported in association with severe ventricular extrasystoles leading to tachycardia in the past.

Linkage Analysis: We could exclude both known genes for ADPKD on chromosome 16p13.3 and 4q21-q23 by microsatellite marker analysis and sequencing of PKD1. A Genome-wide linkage analysis using 385 microsatellite markers at a distance of 10cM identified two possible candidate regions, on chromosome 16 and 7, respectively. Further haplotype analysis could exclude the loci on chromosome 16 and additional marker supported linkage to 7q22 with the maximal multipoint lod score of 2.4. Mutation analysis of candidate genes is currently performed.

Summary: This is the first report of an autosomal dominant ventricular tachycardia associated with polycystic kidney diseases. A genome-wide microsatellite analysis shows linkage of this new phenotype to chromosome 7q22.

P59

Soluble adhesion molecule profile in pediatric patients in relation of aortic coarctation surgical repair

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Background: Increased blood pressure (BP) could lead to mechanical impair of vessel wall and endothelium especially and so contribute to initiation of atherosclerotic lesions. Higher concentrations of circulating adhesion molecules in patients with essential hypertension were described previously. The aim of the present study was to compare concentrations of these molecules in pediatric patients with aortic coarctation (AC) before and after successful surgical correction.

Patients and Methods: 17 children with AC were studied. Blood samples were drawn 1day before surgery and during follow-up long after surgery (346 ± 300 d) in order to exclude trauma and wound healing induced effects. Serum concentrations of soluble E-, P- and L-selectin, soluble intercellular adhesion molecule-1 (sICAM-1), platelet-cellular adhesion molecule-1 (sPECAM-1) were measured by ELISA.

Results: Both systolic and diastolic BP on arms decreased significantly after surgery (SBP/DBP: 126 ± 14/73 ± 13 mmHg vs. 103 ± 9/59 ± 8 mmHg, both $p < 0.001$). On legs, only systolic ($p = 0.003$) but not diastolic BP increased significantly (90 ± 10/59 ± 9 vs. 105 ± 12/56 ± 6). No differences were found in the concentrations of sE-, P- and L-selectin and sICAM-1 before and after surgical repair. In contrast, the serum concentration of sPECAM-1 decreased (59.6 ± 27.1 ng/ml vs. 42.8 ± 13.3 ng/ml, $p < 0.05$) after surgical correction.

Conclusions: Surprisingly, AC is not accompanied by the elevation of traditional markers of endothelial deterioration, like E-selectin. Elevated levels of sPECAM-1 (assumed as a mechanosensing molecule on the surface of the endothelial cells) before surgical correction indicate increased fluid shear stress and endothelial activation and could be specific for this type of hypertension.

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P60

Cyanotic hearts are less tolerant to standard cardiopulmonary bypass techniques

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Objectives: Standard perfusion protocols generally do not differentiate cyanotic versus acynotic hearts. We decided to evaluate the consequences of standard management on left ventricular function of chronically hypoxic versus normoxic hearts.

Methods: Male 5-week old Sprague-Dawley rats (230–250 g) were randomly divided into 2 groups (n = 7/group): normoxia (N) and chronic hypoxia (CH). N rats remained continuously exposed to room air (O_2 sat. = 21%) while CH rats were continuously housed in a normobaric hypoxic chamber (O_2 sat. = 10%). After 2 weeks all rats were sacrificed, hearts were isolated and connected to Langendorff system, perfused (15 ml/min) 30-min with hypoxic buffer (O_2 sat. = 10%), followed by 30-min reoxygenation (O_2 sat. = 100%), 30-min of global myocardial ischemia (no-flow) and 45-min of myocardial reperfusion (same condition as reoxygenation, full flow and O_2 sat. = 100%). This protocol was decided to mimick a conventional open heart procedure on a cyanotic heart, with full reoxygenation at the beginning of cardiopulmonary bypass, myocardial ischemia during aortic cross clamping needed for the intracardiac surgery, and reperfusion after intracardiac repair.

Results: During ischemia: the time to onset of myocardial contracture was the same in both groups, while maximal end-diastolic pressure (EDP) was significantly higher in N hearts (41.6 ± 4.2 mmHg) than in CH (29.4 ± 2.9 mmHg) ($P < 0.01$). End of the reperfusion: CH hearts showed an impaired systolic (Developed Pressure 39 ± 4 vs 81 ± 7 mmHg, $P < 0.001$; Developed Pressure × Heart Rate 7.7 ± 0.8 vs 17.5 ± 2.6, $P < 0.01$; +dP/dt

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1184 ± 128 vs 2148 ± 184, $P < 0.01$) and diastolic ($-dP/dt$ 751 ± 92 vs 1376 ± 129, $P < 0.01$; End Diastolic Pressure 47.2 ± 4.7 vs 29.2 ± 3.4 mmHg, $P < 0.05$) left ventricular function in comparison with N hearts.
Conclusions: Our results allow the following conclusions:

1. both systolic and diastolic left ventricular functions are more compromised in CH than in N hearts at the end of reperfusion, confirming the negative myocardial effects of chronic hypoxia;
2. a different technique of reoxygenation and reperfusion should be considered for chronically cyanotic hearts to reduce the negative effects of the ischemia/reperfusion needed for the intracardiac repair.

P61
Vascular endothelial growth factor in children after cardiac transplantation
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Objectives: Vascular endothelial growth factor (VEGF) is said to be expressed in allografts undergoing rejection as well as in patients with transplant vasculopathy (TVP). Recent studies suggest that the serum VEGF may play an important role in detecting rejection without knowing normal values of transplanted children. We determined normal values of serum VEGF after heart transplantation (HTx) in children. Special emphasis was placed on the pre- and early posttransplant course.
Patients and Methods: 220 VEGF levels were analysed in 60 heart transplanted patients between December 2001 and December 2003. After exclusion of blood samples during infections or rejections 123 analysis were left to generate "normal" values. From these remaining 50 patients 20 were male and 30 female (median age 11.8 years, range 78 days–27.5 years; median body weight 23.6 kg, range 4.1–95.0 kg; median time since HTx 400 days, range 1 day–12.8 years). In addition we determined serial serum VEGF levels of 5 children requiring support by ventricular assist device before cardiac transplantation. VEGF levels were analysed using an enzyme immunoassay (R&D Systems).
Results: Age or sex related differences of VEGF levels in children could not be found. In all patients VEGF dropped immediately after transplantation (day 0–3: mean 0 pg/ml) compared to preoperative values (mean 16.6 pg/ml). From day 4 after HTx onwards children showed increasing VEGF levels (mean 15.5 pg/ml) with peak around day 30 post-HTx (mean 57.3 pg/ml). VEGF concentrations decreased within the first year postoperatively (mean 14.4 pg/ml) reaching preoperative levels. Five and 10 years after HTx VEGF values were chronically elevated (mean 24.0 pg/ml and 23.27 pg/ml). There were no severe rejections. In children with biopsy proven mild rejections there was a tendency to elevated VEGF. In 5 previously resuscitated VAD children we found a continuous decrease during the unloading period on ventricular assist device. After HTx the course of VEGF did not differ from those without previous mechanical support.
Conclusions: We established normal values of VEGF concentration after pediatric HTX and found that they depend strongly on the time after HTX. In the future the study of more patients and events is necessary to prove whether VEGF is a reliable marker for the individual expression of TVP or acute rejection.

P62
Rac controls the expression of the hypoxia-inducible factor (HIF) target gene plasminogen activator inhibitor-1 (PAI-1) in response to thrombin in endothelial and smooth muscle cells
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Pulmonary hypertension is characterized by hypertrophy and enhanced proliferation of vascular cells, frequently associated with a prothrombotic state, which may play an important role in promoting vascular remodeling and thus the manifestation of dysfunction of the pulmonary vasculature. Elevated levels of the coagulation factor thrombin as well as of the inhibitor of fibrinolysis, plasminogen activator inhibitor-1 (PAI-1) have been found in pulmonary hypertension and may contribute to vascular remodeling. However, the mechanisms linking thrombin to the formation of PAI-1 are not completely understood. Since reactive oxygen species (ROS) have been shown to play an important role as signaling molecules in the vasculature, we investigated whether the GTPase Rac, which activates the ROS-generating NADPH oxidase, is involved in thrombin-regulated PAI-1 expression in pulmonary artery smooth muscle cells (PASMC) and microvascular endothelial cells (HMEC). Thrombin increased PAI-1 promoter activity, mRNA and protein levels as well as activity and protein expression of Rac. Active RacG12V or dominant-negative RacT17N increased or decreased, respectively, thrombin-stimulated ROS production as well as thrombin-induced PAI-1 promoter activity, mRNA and protein levels. The RacG12V-activated responses were prevented by antioxidants and the calcium chelator BAPTA-AM. Thrombin also activated the hypoxia-inducible transcription factor HIF-1, which regulates PAI-1 expression, in a Rac-dependent, redox- and calcium-sensitive manner. Finally, proliferation of vascular cells was increased by RacG12V and inhibited by RacT17N. These data show that Rac controls thrombin-induced PAI-1 expression via a redox-sensitive and calcium-dependent pathway, and is essentially involved in regulating the proliferative response of vascular cells. Thus, Rac may play an important role in promoting a prothrombotic state and vascular remodeling processes in pulmonary hypertension.

P63
Assessment of subacute inflammatory and proliferative response to native vessel stenting in a porcine model by local gene expression studies and histomorphometry
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Purpose: The aim of the study was to analyse inflammatory and proliferative response early after stent implantation into native arterial vessels by angiography, histomorphometry and local gene expression analysis using quantitative rt-PCR.
Methods: 8 German domestic pigs underwent stenting of the left coronary artery. Selective coronary angiography was performed after 14 days. Explanted coronary arteries were examined histomorphometrically after methacrylate-embedding. Snap-frozen samples were examined for local gene expression of TGF-β,

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TNF-α, GM-CSF, VEGF, PDGF and Fas Ligand by real-time quantitative rt-PCR normalized to the housekeeping gene GAPDH and compared to un-stented coronary arteries.
Results: All stented coronaries were patent with only little neointima formation. The median vessel diameter was 2.55 mm (range 2.43 mm–2.68 mm). Histopathology revealed little inflammatory response limited to the tissue surrounding the stent struts; luminal area ranged from 84 to 91%. Compared to unstented control arteries, no significant differences in local gene expression were detected for VEGF, PDGF, TGF-β, TNF-α and GM-CSF. Expression of Fas Ligand was upregulated as little as 1.7-fold ($p = 0.01$).
Conclusion: Stent implantation into native arterial vessels does not result in significant upregulation of investigated genes regulating vascular remodelling, inflammation or fibrogenesis. Whether upregulation of Fas Ligand as a marker gene of apoptosis is transient and biological significant requires further investigation.

Session 6: Fetal Cardiology

P64
Assessment of fetal atrio-ventricular (AV) conduction time by tissue Doppler and its correlation with conventional Doppler flow measurements

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Objectives: It is possible to identify the chronology of atrial and ventricular depolarization by indirect methods using mechanical or hemodynamic surrogates. Simultaneous recordings of blood flow velocities in the ascending aorta/SVC or left ventricular in/outflow are established methods for AV conduction time measurement with clear landmarks, but the measured AV events depend on loading conditions. Longitudinal myocardial tissue motion is less load-dependent and can be directly assessed by Tissue Doppler (TV). The objectives of this study are to establish normal value of AV conduction time measured by TV, and to correlate the findings with flow-velocity Doppler techniques.
Methods and Results: AV conduction time was assessed in 47 normal fetuses (gestational age range 15–38, median 20 weeks). Simultaneous Doppler flow tracings of left ventricular in/outflow tract as well as the aorta/SVC were obtained as described previously. 2-D guided TV images were obtained in a 4-chamber view (Vivid 7, 3.5- to 5-MHz transducers). The imaging angle was narrowed and the scale was optimized to obtain the highest frame rates. Frame rates ranged from 185 to 276 with a temporal resolution between 3.6 and 5.4 ms. At least 5 cine-loops of 4–10 cardiac cycles were stored as raw data and analysed off-line. TV curves were obtained from the right ventricular free wall at the level of the AV groove. AV conduction times were measured by the following methods: (1) In/outflow Doppler (interval between A wave onset and outflow onset); (2) Aorta/SVC Doppler (interval between A reversal wave onset in the SVC and onset of forward flow in aorta); and (3) TV (A wave signal onset to isovolumetric contraction wave onset). Normal AV conduction times by in/outflow and aorta/SVC Doppler were 115.8 ± 8.0 ms and 114.5 ± 7.6 ms respectively ($p = NS$). AV conduction time by TV was significantly shorter: 86.9 ± 8.4 ms ($p < 0.001$). There was a correlation between the 2 Doppler modalities ($r = 0.43, p < 0.01$) and in/outflow Doppler and TV ($r = 0.34, p < 0.05$). Intraobserver variability of measurement by TV was 2.0%, and interobserver variability was 8.8%.
Conclusions: Tissue Doppler allows reliable and reproducible measurements of AV conduction time of the normal fetus. TV is thought to be less dependent on ventricular loading and therefore may be more accurate in the presence of hemodynamic changes.

P65
Management of newborns with structural heart disease: impact of prenatal diagnosis

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Aim: The main objective was to assess the influence of prenatal diagnosis of structural heart disease on the cascade of measures leading to admittance to a heart centre and on morbidity and outcome.
Methods: Retrospective analysis of medical records of all newborns (admittance during the first 28 days of live) from 1998 to 2002 at a regional specialized pediatric cardiac centre.
Results: In the 5-year period 383 newborns were referred. 317 of them had congenital heart disease. An exact prenatal diagnosis was confirmed in 50 (15,8%). 81% of all patients with duct-dependent lesions had prostaglandin before arriving in the heart centre irrespective of the prenatal diagnosis. Prenatally diagnosed patients were less often ventilated mechanically (12% vs. 31%), they reached the heart centre earlier, had lower serum-urea levels, a tendency to lower and almost never excessive lactate levels. In contrast to the literature median pH values of patients with transposition and left heart obstruction were at least 7,32 at admission and there was no additional benefit of prenatal diagnosis. Newborns with left heart obstruction and prenatal diagnosis were operated 7 days earlier ($p < 0{,}001$), but hospital stay was not shortened in any of the analysed groups. There was no significant influence of prenatal diagnosis on early neurological outcome and on hospital mortality. In summary our data suggest a lower morbidity in newborn babies with prenatal diagnosis. Moreover we could identify 37 newborns who died from congenital heart disease in this region during the study period and never reached a heart centre. In most of them we failed to find out the correct morphological diagnosis and probably they have not been diagnosed prenatally. So we speculate that inclusion of these nonsurvivors into statistical analysis would have shown the benefit of prenatal diagnosis more significantly.
Conclusion: Correct prenatal diagnosis was established in 15,8% of all admitted newborns in a 5-year period which is comparable to the literature. We were able to confirm a headstart of patients prenatally diagnosed and a benefit of prenatal diagnosis on parameters of morbidity. So more efforts should be done to increase the number of selected prenatal echocardiographies.

P66
Long axis ventricular function in normal mid-gestation fetuses

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Objective: We have previously reported the feasibility of using angular M-mode echocardiography to assess long-axis function in the fetal heart. The aim of this study is to observe this in the fetal heart from 19–23 weeks of gestation.
Methods: This was a cross sectional prospective observational study. Only pregnancies thought to have a structurally normal heart were included in the study. Fetuses known to have had an increased Nuchal Translucency in the first trimester were excluded. The

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four-chamber view was obtained and angular M-Mode (Aloka SSD-5500 PHD) was applied either live or on a cine loop recording. The cursor line was placed from the cardiac apex to the tricuspid and mitral valve rings. M-mode tracings of valve movements towards and away from the apex throughout the cardiac cycle were obtained. Images were stored digitally and further analysis carried out off-line. Tricuspid and mitral valve movements over time were manually digitised from two to three cardiac cycles using a dedicated computer software. Total amplitude of valve excursion (VE), peak systolic (PSV) and peak diastolic velocities (PDV) and time to peak systolic (TPSV) and peak diastolic velocities (TPDV) were calculated for the right (RV) and left ventricle (LV) based on tricuspid and mitral valve movements, respectively.

Results: 18 fetuses from 19–23 weeks of gestation were included in the study. Tricuspid VE (mean = 4.5 mm; 95% CI of the mean: 4 to 5) was greater than mitral VE (mean = 3.4 mm; 95% CI of the mean: 3.1 to 3.8); p = 0.001. RV-PSV (mean = 3.3 cm/s; 95% CI of the mean: 3 to 3.7) was greater and LV-PSV (mean = 2.7 cm/s; 95% CI of the mean: 2.4 to 3); p = 0.01. RV-PDV (mean = 5 cm/s; 95% CI of the mean = 4.3 to 5.8) was also greater than LV-PDV (mean = 3.6 cm/s; 95% CI of the mean = 3.1 to 4.2); p = 0.001. Conversely, TPSV and TPDV were similar for RV and LV (TPSV, p = 0.43 and TPDV, p = 0.7).

Conclusion: In mid-gestation, the greater amplitude of tricuspid movement and greater RV systolic and diastolic velocities may be related to lower RV afterload. Similar RV and LV time to reach PSV and PDV suggest that despite differences in indices of systolic and diastolic function, timing of right and left ventricular shortening and lengthening are coordinate.

P67
Late recurrence of tachycardia in the cases with prenatally detected supraventricular tachycardia

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Supraventricular tachycardia (SVT) is the most common type of fetal tachyarrhythmia. Although pre- and postnatal treatment and short-term outcome have been reported, long term outcome is still unclear. Early disappearance and late recurrence of SVT was reported for the neonate and infant with WPW syndrome whose initial SVT began at age 0 to 2 months. Hence, the purpose of this study is to elucidate long term outcome of SVT presented before birth.

Patients and Method: Fetal SVT was defined as fetal tachycardia with the ventricular rate more than 200 bpm with 1:1 atrioventricular conduction. All 6 cases with fetal SVT between 1982 to 2001 were included. Postnatal follow-up period was 2 to 18 years (mean 8.3 years).

Result: Perinatal course: Fetal PSVT was diagnosed at 20 to 40 weeks of gestation (mean 30). Of the 6, 5 underwent prenatal antiarrhythmic treatment, and all were effective (all 3 cases with fetal hydrops were resolved in utero). These cases were delivered at 35 to 40 weeks of gestation. After birth, 4 cases revealed atrioventricualr reentry tachycardia (AVRT), and all 4 had refractory AVRT with the requirement of 2 or more medication until 39 to 97 days after birth. Postnatal ECG in 3 cases revealed delta wave in neonatal period. However, once the episode of tachycardia was disappeared, AVRT had been rarely recurrent and preventive antiarrhythmic drugs had been able to be discontinued until 9 months after birth. Late recurrence: Of the 6, 2 had recurrent AVRT. For the first female case with no delta wave in neonatal period, AVRT was reappeared at 10 years after birth. Her ECG revealed delta wave at that time. Her AVRT was frequently recurrent, and she underwent ablation therapy at 14 years old. For the second male case, AVRT was also reappeared at 10 years after birth. His ECG showed delta wave at neonatal period. During follow-up, ECG revealed two forms of delta wave suggested two accessory connections. He is currently followed with no medication, since the episode of AVRT is not frequent.

Conclusions: Early disappearance and late recurrence was also found for the cases with fetal SVT. This recurrence may not be predicted by the presence of the delta wave by the ECG at neonatal period. Hence, close long-term follow-up is required for the cases with AVRT started in fetal period.

P68
Foetal increased nuchal translucency: is this associated with foetal cardiac dysfunction?

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Introduction: An increased nuchal translucency (NT) appears in several fetal abnormalities. A common morphological denominator explaining the complete spectrum of malformations in which it is found, its cause and its transient nature remains unknown. Tempory cardiac failure has been proposed.

Aim: Objective measurement of cardiac performance at 11–14 weeks gestation in fetuses with an increased and normal NT.

Methods: Prospective pilot study of 29 fetuses. An increased NT was defined as ⩾2,5 mm at 11 weeks, ⩾2,7 mm at 12 weeks and ⩾2,9 mm at 13 weeks gestation. Fetal echocardiograms were performed (mostly transabdominal) measuring: great vessel diameter, peak velocity (pkV) and the ejection flow velocity time integral (FVI), E/A ratio over atrioventricular valves and heart rate (HR). The great vessel cross-sectional area (CSA) was calculated ($CSA = \pi(D/2)^2$). Systolic cardiac function expressed as stroke volume (SV), SV (ml) = FVI × CSA and cardiac output (CO), CO (ml/min) = SV × HR. Diastolic function expessed as E/A ratios. Where indicated chromosomal analysis was performed.

Results: 10 normal NT (mean NT 1,8 ± 0,4), 12 increased NT (mean NT 3,4 ± 0,4) and 7 increased NT with chromosomal abnormalities (mean NT 3,7 ± 0,9). The chromosomal abnormalities were 1 trisomy X, 3 trisomy 18 and 3 trisomy 21.

		LV SV	LV CO	RV SV	RV CO	TV E/A	MV E/A
Normal NT	Mean	0,225 (±0,11)	35,7 (±15,6)	0,132 (±0,05)	21,3 (±5,2)	0,554 (±0,08)	0,530 (±0,1)
	Median	0,19	29,3	0,144	24,6	0,568	0,518
Increased NT	Mean	0,185 (±0,13)	28,2 (±18,2)	0,114 (±0,06)	17,7 (±9,2)	0,577 (±0,09)	0,565 (±0,14)
	Median	0,159	24,4	0,113	16,2	0,557	0,533
Chrom AbN	Mean	0,102 (±0,05)	16,3 (±7,5)	0,096 (±0,06)	15,7 (±9,5)	0,558 (±0,07)	0,531 (±0,06)
	Median	0,118	19,0	0,091	15,4	0,553	0,533

Conclusions:

1. No relationship between increased NT and diastolic cardiac dysfunction.
2. SV and CO diminishes with increasing NT measurements suggesting systolic dysfunction in these fetuses. This is exaggerated in the presence of chromosomal abnormalities.

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3. The RV SV is greater than LV SV at gestational age 11–14 weeks.
4. Direct measurement of cardiac function is possible in 11–14 week fetuses.

Limitations: Small study size, accuracy of measurements.

P69

Evaluation of a new technology using 3-dimensional ultrasound and 3-dimensional Color- and Power-Doppler in fetuses with and without congenital heart defects

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Objectives: The technology of fetal 3-dimensional echocardiography and 3-dimensional Color- and Power-Doppler seems to get more and more used. To clarify the question if 3-dimensional evaluation of fetuses with and without congenital heart defects is improving current diagnostic tools the presented pilot trial has been undertaken.
Material and Methods: We investigated 25 singleton fetuses between 17 and 35 weeks of gestation. In 8 cases no heart or other morphologic defect was seen. In 18 cases complex or simple congenital heart defects were present. Using the Voluson 730 Expert Machine (GE, Germany) a standardized sonografic evaluation was intended including: biometry, Pulsed Doppler of umbilical artery and middle cerebral artery, 3-dimensional volumetry of brain, lungs and heart, and 3-dimensional analysis of all cardiac valves. The left- and right-ventricular mass were calculated, and 3-dimensional Colour- and Power-Doppler reconstruction of the pulmonary and placental vessels was also performed on each patient.
Results: In general volumetry of the fetal heart and other organic systems could be performed with increasing feasibility paralleling advancing gestational age. Practicability of 3-dimensional evaluation of the cardiac valves was only given in approx. 25% of the cases. During pregnancy a steady increase in left and right ventricular mass could be demonstrated. 3-dimensional Colour- and Power-3D-Doppler allowed visualisation of central and peripheral pulmonary as well as placental vessels in most cases.
Conclusions: Using the new 3-dimensional techniques in prenatal sonography and echocardiography more insights and improved diagnostic features are given. In individual cases there may be great benefit in preterm diagnostic knowledge, so that postnatal procedure can be elaborated together with pediatric cardiologists, obstetricians, and pediatric cardiac surgeons.

P70

Anatomical considerations in Ebstein anomaly in fetuses and its influence on neonatal treatment

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The aim of the study was to evaluate anatomic and hemodynamic spectrum of fetuses with Ebstein anomaly (Eb) and changes which occurred in neonates. Eb was suspected in 13, and proved in 12 fetuses. In one excluded case abnormal musculature of the RV was the cause of fetal TR, without anatomical changes of TV. It resolved spontaneously during 5 months of life, the child has normal heart. Time of fetal diagnosis was 16–37 weeks, mean 29 weeks. Karyotype was evaluated in 9 fetuses. Trisomy 21 was in 1, dysbalanced translocation in 1 – this pregnancy was terminated. Obstetric USG findings were suspicious of Eb in all cases: enlarged RA in all, grossly enlarged heart (Ha/Ca $>=$ 0,5) in 10. In 1 fetus Norrie syndrome was suspected. In echocardiography: TV appearance was suggested of Eb in all cases. Holosystolic TR was in all fetuses, with Vmax varied from very low in fetuses with functional pulmonary atresia (PAtr), to high in fetuses with forward flow or artretic pulmonary valve. Forward flow in PA was in 3 fetuses, PA was atretic in 3, functional PAtr was in 5. Nonimmune hydrops was in 2 fetuses, 1 of whom died in utero, 1 in the neonatal period before the operation. In 1 fetus with functional PAtr all signs resolved spontaneously during the first month of life, the child is in good condition with mild type of Eb. 2 children did not require operation. 1 neonate with genetic Norrie syndrome died just after delivery. Four neonates were operated on in two different cardiac surgery departments. Just one with severe Eb with functional PAtr survived, in whom precise perinatal treatment was planned before delivery. Modified Starnes procedure was performed. The boy is 2 years old now after hemi-Fontan operation with good general condition. 4 children survived: 3 without surgery and 1 after emergency neonatal treatment. We conclude that:

1. Spectrum of prenatal Eb is wide and signs can change after delivery due to lowering pulmonary vascular resistance in the neonatal period.
2. Detailed anatomy of a TV can be difficult to evaluate in fetuses.
3. Holosytolic TR can lead to severe fetal heart failure causing fetal hydrops.
4. Functional PAtr complicated fetal Eb is real neonatal emergency and close cooperation between fetal cardiologist and cardiac surgeon is needed to improve outcome.
5. Chromosomal and genetic disorders can coexist with Ebstein syndrome.

P71

Cardiovascular score in fetuses with complete heart block – diagnosis, management, outcome

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Objectives: To find prognostic signs in fetal echocardiography to predict outcome in fetuses with CHB.
Material and Methods: In 17 fetuses with CHB cardiovascular score (CVS) was used to evaluate fetal circulation. Normal was classified as 10 points. 6 fetuses had CHB without heart defect (CHD), 5 of them had positive anti-Ro antibodies. CHB and CHD were in 11: left atrial isomerism (LAI) in 8, dextrocardia/l-TGA in 2, AVSD/Down syndrome in 1. First exam was performed between 10 and 34 weeks of pregnancy (WOP), median 23 $\pm$ 5.
Results: CVS in 6 fetuses without CHD was between 5 and 9, initial HR varied from 48 to 80. 2 women terminated pregnancy. Dexamethason and salbutamol was administered in 4, resulted in lowering degree of CHB from 3rd to 2nd in 1; increasing FHR in 2. There were signs of lupus erythemathosus in 1 neonate in whom CHB did not respond to oral β-mimetics. Pacemaker was implanted in the 4th month. In 8 fetuses with LAI, CVS varied from 1 to 6. In 6 with nonnimmune hydrops (NIH) CVS was below 4 and 5 of them died in-utero. β-mimetics were administered in 5, without

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effect. 2 fetuses without NIH were delivered vaginally. In 1 pacemaker was implanted but he died 2 months later. In 1 case CHB was complicated by atrial flutter. The boy died in the 5th day of life. Condition of a fetus with AVSD and Down syndrome deteriorated in spite of β-mimetics. CVS was 1. She died in the 3rd day of life. 2 fetuses with l-TGA had higher CVS (6 and 7). β-mimetics "speeded-up" their HR. In spite of this, neonatal condition was severe in both cases, and both died – one after pacemaker implantation, the second before the operation. All fetuses with CHB and CHD died in utero or after delivery. All of them had signs of fetal circulatory failure, and those with NIH had CVS less than 4. Children who survived had CVS above 7.
Conclusions:

1. CHB without CHD was due to positive anti-Ro antibodies, could be successfully treated in-utero, outcome was good.
2. Neonates with CHB without CHD did not require pacemaker during neonatal period, but careful cardiac evaluation was needed.
3. CHB with CHD, according to our experience, was lethal condition, what was necessary information when counseling parents, planning transplacental treatment and way of delivery.
4. CVS is easy and useful prognostic factor to predict outcome of fetuses with CHB.

P72
Totally anomalous pulmonary venous drainage in the fetus revisited
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Totally anomalous pulmonary venous drainage (TAPVD), as an isolated lesion, is a rare form of congenital cardiac malformation occurring in approximately 1/20,000 live-births. It can be a difficult diagnosis to make echocardiographically in a symptomatic infant and more so in fetal life, in the absence of clinical suspicion. Therefore, the connection of at least one pulmonary vein to the left atrium must be demonstrated by cross-sectional imaging in every fetus, and confirmed using colour flow mapping. However, where image quality is limited, this may be challenging. In a previous paper, it was suggested that most cases of TAPVD would demonstrate ventricular and great artery disproportion, with right-sided dilatation relative to the left. The explanation for this was increased venous return to the right side and decreased to the left, when the veins all drained to the coronary sinus or to a supracardiac vein. The only case examined in fetal life of TAPVD draining below the diaphragm did not show disproportion, presumably because the addition to venous return in the inferior vena cava was directed across the foramen ovale to the left ventricle. However, we have recently detected two cases of TAPVD, which appeared to drain to the SVC, but there was absolutely no disproportion at repeated scans. A possible explanation for this is that there was a large secundum atrial septal communication allowing a greater than normal right to left shunt, but this was not evident on direct examination of the foramenal defect. Another suggested aid to the diagnosis of TAPVD has been to use pulsed Doppler as a confirmation of a normal pattern of pulmonary venous blood flow. However, although this was abnormal in one of our cases, where the ascending vein was obstructed, it was normal in the other unobstructed case. In conclusion, there are no consistent clues to the diagnosis of isolated TAPVD in the fetus and the confident demonstration of at least one pulmonary vein with flow into the left atrium is mandatory in order to exclude this malformation.

Session 7: Interventional Cardiology

P73
Work-up of cardiovascular implants after explantation for biocompatibility screening
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Background: Interest in information on biocompatibility of implants after interventional implantation is increasing. We discuss methods for biocompatibility screening of cardiovascular implants.
Discussion of Methods and Obtainable Results: Use of standard histology after embedding in paraffin is limited since metallic implants have to be removed during work-up with disruption of the specimen. Alternatively tissue blocs containing an implant can be embedded in methylmetacrylate and processed by sectioning with a diamond cutter and grinding, thus leaving the implant in situ and saving the tissue/implant interface for detection of local inflammation. New methacrylate resins and embedding techniques allow for specific immunohistochemical staining of the specimens. Another important aspect of evaluation should be the progress of organisation of thrombotic material which often surrounds the implant immediately after implantation. Scanning electron microscopy (SEM) allows evaluation of endothelialisation of the vascular surface of the implant. Tissue blocs submitted for SEM should be fixed in glutaraldehyde 2,5%, while the parts for embedding in paraffin or methylmetacrylate should be placed in formaline or alcohol (ethanol/methanol 1:1).

Illustrating the use of these technologies we demonstrate findings in >100 tissue specimen (human and animal). Early endothelialisation of the vascular surface is seen after implantation of septal occluders, stents, or coils. Implant-host reactions range from mild to moderate (metal MP 314 N, Nitinol, polyvinylalcohol) to severe (Dacron®, Gore Tex®). Charaterisation of tissue surrounding the implant can be demonstrated by means of immunohistochemical stainings.
Conclusions: With an optimal work-up of cardiovascular implants, ingrowth and endothelialisation as well as inflammatory reactions in the surrounding tissue can be characterised. These information serve as valuable basis for optimisation of biocompatibility by implant modification.

P74
The new low profile, high pressure NuMED COEfficient™ balloon catheter: usefulness in the treatment of complex CHD in infants & small children
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Objectives: Treatment of resistant vascular obstructions associated with CHD requires high pressure balloon angioplasty (BA) and/or stent implantation. Vascular access & vessel size in infants & small children may limit therapeutic transcatheter options. We report the use of the NuMED COEfficient™ Balloon Catheter (COE) in the treatment of complex CHD in this patient population.
Methods: CATHETER SPECIFICATIONS: balloon diameters: 4–12 mm, shaft size: 3.5 Fr, guidewire: 0.018#, rated burst pressure: 7–13 atm, introducer sheath required: 4–7 Fr. STUDY POPULATION: From 9/02 through 12/03, 310 interventional caths were performed. In 24 patients (8%), a COE was used. Age ranged from 2 wks to 4½ yrs, with 60% = 6 mos, while weight ranged from 3.2 to 15.2 Kg. Expanded balloons ranged from

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4–12 mm using 8–16 atm & requiring 4–6 Fr sheaths. 26 lesions were treated: BA performed for resistant native CoA (3), recurrent CoA (6), LPA stenosis (4) & RPA stenosis (2), cavopulmonary obstruction in single ventricle patients (2), & dysplastic pulmonary valve stenosis (DPVS) (5). Stents mounted on the COE were implanted for LPA stenosis (2), while (1) required intraoperative delivery of 2 stents to reconnect an isolated LPA during a "Hybrid" procedure off CPB, with (1) infant undergoing redilation of an SVC stent.
Results: BA was successful in all vascular obstructions, while the 5 infants with DPVS also had effective treatment, avoiding surgery. All stents were deployed and/or redilated successfully without residual obstruction, stent migration or malposition. There was no vascular compromise.
Conclusions:

1. The low profile, high pressure COE is effective in the transcatheter therapy of complex CHD in infants & small children.
2. Resistant lesions are especially suitable for this catheter.
3. Age & size should not delay transcatheter therapy.

P75
Is interventional treatment of pulmonary atresia intact septum (PA-IS) a low-risk approach?
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Interventional treatment is considered the elective approach to favourable forms of PA-IS. However this method is still burdened by a not negligible risk. The aim of our study was to investigate the causes of death in children undergoing interventional treatment of with PA-IS.

From January 1991 to January 2003, 54 new borns with PA-IS underwent initial interventional treatment. There were 8 hospital deaths and 1 late death (16.7%).

All patients with a fatal outcome had tripartite right ventricle (RV) and tricuspid z value >-2; 4(44%) had tricuspid valve dysplasia with moderate to severe regurgitation. Six patients underwent urgent surgery, consisting into B/T shunt in 5 cases and B/T shunt plus pulmonary valvotomy in one case. Reasons for surgery were persistent duct dependent circulation in 4 and reversed flow from the patent ductus to pulmonary trunk (PT), RV and right atrium in 2. There were 3 periprocedural deaths (5.5%): 2 were due to infundibular perforation, 1 to refractory supraventricular tachycardia. Six postoperative deaths occurred, due to complete heart block (day 2), massive pulmonary embolism (day 9), sepsis (day 2), late shunt occlusion (11 months). Patients operated for reversed flow through pulmonary trunk and right cavities died; the first one in the operating room after duct ligation, the second one 1 day after B/T shunt. In conclusion, early mortality remains elevated in patients undergoing interventional treatment of PA-IS. Reversed flow trough the pulmonary trunk and RV can be fatal. Patients with incompetent dysplastic tricuspid valve should probably undergo primary surgical repair.

P76
Partial anomalous pulmonary venous drainage of the upper left pulmonary veins – catheter-interventional treatment is sometimes possible
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Objectives: Partial anomalous pulmonary venous drainage (PAPVD) of the upper left pulmonary veins (ULPV) is a rare congenital anomaly leading to left to right shunt. Embryogenesis is thought to consist of nondevelopment of the connection between ULPV and left atrium and the persistence of a primitive "vertical" vein between ULPV and the left brachiocephalic vein. The lesion must not be confused with a persistent left superior vena cava. Standard treatment is surgical redirection of the ULPV to the left atrium.
Methods: Retrospective observational report.
Results: Six cases with PAPVD of the ULPV (0.4%) were identified out of 1500 patients who underwent cardiac catheterizations for congenital heart disease performed from 01/2000 to 12/2003. Age ranged from 1.5 to 35 years (median 7.5 years). Four patients had additional cardiac defects (small ventricular septal defect, aortic coarctation, tetralogy of Fallot, Mb. Ebstein). In three of the six patients (50%), there was an additional connection to the left atrium, which could be identified by contrast injection into the vertical vein after superior balloon occlusion. All three of them had larger left to right shunts (Qp:Qs $>$ 2:1) than expected from PAPVD alone. Surgical ligation of the vertical vein was performed in the first, transcatheter device closure of the superior part of the vertical vein led to successful correction in the other two of these patients. Surgical redirection by anastomosis of the LUPV to the left atrial appendage was performed in the other three patients.
Conclusions: PAPVD of the ULPV is extremely rare. An additional connection to the left atrium might easily escape detection. Although a rate of 50% in this small series of six is probably completely random, such an additional vessel should be looked for, because interventional correction might then be possible. We were not able to find descriptions of this special, intervention-friendly type of lesion in the literature.

P77
Mid-term effects of stent implantation for aortic reaorctation on systemic hypertension and endothelial dysfunction
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Primary stent implantation is a diffuse technique to treat aortic recoarctation. However, the effects of stent implantation on hypertensive profile and vascular dysfunction have not been extensively investigated. From 1/1/99 to 1/1/2003, 16 patients (median age and weight 17 years and 56 Kg) underwent direct stenting of aortic recoarctation. Indication to interventional treatment were: clinical gradient $>$ 20 mmHg in 14, systemic hypertension at rest in 5, severe systemic hypertension on effort in 11. Five patients received anti-hypertensive treatment. All patients had vascular dysfunction, consisting in increased carotid endothelium thickness, decreased vasodilation after glyceryl trinitrate administration, diminished elasticity and compliance. Immediately after stent implantation median gradient through the isthmus diminished from 32 to 5 mmHg and morphological result was invariably good. At a median follow-up of 24 months 13 patients underwent effort test and vascular echography. One patient had a persistent systemic hypertension at rest, 8 had mild and 5 severe systemic hypertension on effort; 3 received an anti-hypertensive treatment. Vascular echography invariably showed persistent increased endotheliad thickness and diminished elasticity, compliance and vasodilation. In conclusion primary stenting for aortic recoarctation has excellent immediate results. However, systemic hypertension at rest and on effort is reversed in a minority of patients. In addition, all patients continue to show vascular dysfunction at mid-term follow-up.

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Long term studies are needed to investigate possible normalisation of vascular reactivity. Systematic medical treatment of these patients could be taken into consideration.

P78

Transcatheter closure of a patent arterial duct in small children

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Introduction: Percutaneous closure is a method of choice in Patent Ductus Arteriosus (PDA) treatment in all cases but premature infant with large duct. In infants PDA can close spontaneously and therefore the elective interventional catheterization is usually performed after twelve months of age, except cases with large PDA causing congestive heart failure. The aim of the study was to present the treatment results in small children treated in one centre.
Material and Methods: Between 1996 and 2003 catheter closure of PDA was performed in 252 patients, 61 of them were younger than 24 months of age (mean age 13,8 range 6–24 mths). Body weight ranged between 6 and 17 kg (mean 11 kg). Coils (C) were employed in 52 cases, Amplatzer Duct Occluder (ADO) in 9 (from 1998). Mean PDA diameter in group C was 1.9 mm (range: 1.2 to 3.2 mm). In ADO group mean PDA diameter was 3.3 mm (range 2.8–4.6 mm). Before the "ADO era" two coils (via artery or via artery and vein access) were employed.
Results: In all 61 pts interventional catheterization was performed in a standard fashion. In 6 pts with PDA diameter bigger than 2.5 mm simultaneous implantation of two coils was performed. In group C in 7/52 (13.5%) pts residual shunting was noted. In 4 of them another coil was added in order to achieve definitive closure. In group C in 8 pts heparin infusion during the first day after procedure was used because of temporary lack of femoral pulse. In three cases in group C coil migration to pulmonary artery (2 pts) and aorta (1 pt) occured. All embolized coils were snared and in 1 pt another C was successfully implanted during the same procedure. In one infant after 8-PDA-5 C implantation "jo-jo phenomenon" happened. This coil was also retrieved. In one child in C group 6 mmHg gradient in left pulmonary artery was noted, which resolved with time. In ADO group all PDA were closed without residual shunting after 24 h. There were no procedure related complications.
Conclusions: Catheter PDA closure is an interesting and effective alternative to surgical ligation in small children. In PDA diameter $>$ 2.5 mm Amplatzer Duct Occluder is a method of choice. Smaller PDA may be closed safely with coils.

P79

Transcatheter closure of coronary artery fistulas

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Eight patients with a coronary artery fistula underwent percutaneous transcatheter embolization. Four patients were female and four female; the age range was 5 to 41 years [median 15]. In all patients, the fistula was an isolated congenital anomaly; in one, it was a residual shunt after previous surgical closure. The fistula arose from branches of the left (n = 3) and right (n = 5) coronary arteries and drained to the right ventricle (n = 2), right atrium (n = 4) and pulmonary artery (n = 2). Different embolization techniques were used to occlude the coronary fistulas. Both retrograde and antegrade approach was used for device placement. The embolization materials included an Amplatzer ASD Occluder (n = 1), Amplatzer ASD DuctOccluder (n = 1), embolization with Pfm coils (n = 6; single coil in 3 patient, multiple coils in 3 patients). In one patient a coil embolized into the right atrium and was removed in the same session. There were no other procedural problems or associated complications in any other patient. Follow-up investigation by Doppler ultrasound or coronary angiography 3 months after showed that permanent occlusion was achieved in six patients. In 2 patients a residual shunt remained after implantation of a single coil. The residual shunt could be successfully closed using additional coils. Transcatheter embolization can be considered the treatment of choice for coronary artery fistulas.

P80

Intracardiac perforation with left ventricular to right atrial shunt after fracture of a right sided arm of an atrial septal occluder

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Interventional closure of atrial septal defects is a general accepted alternative to surgery. Different types of devices have been developed with different configurations and closing mechanisms. As there is no ideal device there are potential risks for complications.

We report on a 17 year old girl who underwent an interventional closure of a defect within the oval fossa 3,5 years ago. The right ventricle was enlarged, QP/QS ratio was 2:1. The stretched diameter of the oval defect was 18 to 12 mm. We implanted a 40 mm Starflex device. The defect was completely closed, repeat investigations with transthroracic echocardiography (TTE) and fluoroscopy were normal. On the last outpatient appointment a shunt between left ventricle and right atrium as was seen in TTE and confirmed by transoesophageal echo. Fluoroscopy now showed a fracture of one of the right sided arms. The device was removed surgically. Opening the right atrium the broken leg could be seen in the right atrial cavity pointing towards the membranous septum. It was not covered by endothelium or patch material.

Arm fracture are described after implantation of Starflex/Cardioseal devices depending on the size, mostly in the biggest 40 mm device. We describe a case of late complication after arm fracture with perforation of the membranous septum leading to a Gerbode defect. Late follow-up in large Cardioseal/Starflex are mandatory.

P81

Stent implantation for aortic coarctation in a 1500 g premature triplet through the left carotid artery

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Objectives: Therapeutic cardiac catheterization in native coarctation is an effective approach in patients with left ventricular failure. After they recover, surgery can be performed safely. We report

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on a patient with delayed diagnosis of aortic coarctation, cardiac resuscitation, where stent implantation was a life saving catheter intervention.

Patient: A 1470 g premature triplet suffered from mild respiratory distress syndrome after birth. On the 18th day of life he developed tachypnoe, bradycardia and cyanosis. Cardiopulmonary resuscitation was necessary and echocardiography assessment of heart function confirmed aortic coarctation and absence of an arterial duct.

Methods: With continuous infusion of catecholamines, controlled mechanical ventilation he underwent heart catheterization with balloon dilatation of a severe native coarctation to a final balloon diameter of 4 mm. Despite improved abdominal flow he developed renal failure and moreover recoarctation within 24 hours. Because of multiorgan failure, surgery was not possible he had repeat recatheterization. There was no further arterial access from the femoral vessels, so we decided to canulate the left carotid artery. After placement of a 4F introducer sheath, the hypoplastic aortic arch with recoarctation was visualized angiographically. A coronary stent 9 × 4 mm (Tecnic, Sorin/Biomedical) was advanced in the stenotic region and implanted to a final diameter of 4 mm. Arterial pressure in the ascending aorta dropped from 115/52 mmHg to 79/39 mmHg the pressure gradient went down from 75 to 15 mmHg.

Clinical Course: Even after effective treatment of the coarctation persistent renal insufficiency required peritoneal dialysis for 7 days. He developed congestion of the bowel but no necrotizing enterocolitis. Heart function improved and neurological diagnostics revealed no cerebral damage or EEG abnormalities. On medication with captopril and atenolol he now has moderate arterial hypertension. Until now there is no recoarctation, surgical removal of the stent will be performed when recoarctation develops.

Conclusions: In premature newborns with aortic coarctation, stent implantation can be performed safely with vascular access from the carotid artery. The intervention might be life saving when surgical correction is obsolete.

P82

Percutaneous balloon aortic valvotomy in childhood

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We assessed the effectiveness of balloon valvuloplasty in 34 children who were admitted for aortic valve balloon dilatation over 7 years (97–03) in two institutions (Stuttgart, Zagreb) with prevalence of male (28/6; $p < 0.01$), age at dilatation 35.61 ± 55.39 months (mean ± SD, min 1 day, max 14.2 years), BW 13.1 ± 15.9 kg (min 2640 gr, max 57 kg). Fifteen of them (47%) had no clinical symptoms, 12 (35.3%) were dyspnoeic, and 6 (17.6%) in severe heart failure. According to ECHO, ventricular function was normal in 6 (17.6%), moderately limited in 17 (50%) and severely impaired in 11 (32.4%) pts. Immediately after valvuloplasty the mean systolic pressure gradient across the aortic valve decreased from 70.62 ± 20.78 (max 120, min 45 mmHg) to 20.03 ± 13.7 (max 65, min 0 mmHg; $p < 0.01$) (cath. measurement). The balloon/aortic valve ring ratio was 0.85 ± 0.09 (max 1.00, min 0.60). The degree of aortic regurgitation immediately after catheterisation did not significantly increase; only 1 patient developed moderate aortic regurgitation which was treated with surgical valve reconstruction on the same day. Long term results were studied in 30 patients 1–72 months (mean 18.53 ± 19.69) after valvuloplasty and revealed continuously increasing residual aortic valve gradient 31.35 ± 12.01 (max 50, min 15 mmHg), still beeing significantly lower ($p < 0.001$) than before valvuloplasty. On following aortic regurgitation increased to grade 3 in 3 (10%) and to grade 2 in 7 (23.3%) pts. Eight required reintervention, one of them balloon valvuloplasty only, 4 balloon valvuloplasty and surgery and 3 surgery only. Among 7 pts. requiring surgery, three were operated during a period of 27–77 months and four within one month after the dilatation. One patient died one week after dilatation, redilatation and surgery due to fibroelastosis (HHLS like heart), all other survived without complications. The overall actuarial survival rate after 7 years was 96.6%. The freedom of any reintervention was 76%, 73% and 61%, and for surgical reintervention 88%, 85% and 79% at 2, 4 and 7 years. In the subgroup of pts. having criteria for critical aortic stenosis no difference was found in comparison to other patients. Percutaneous balloon valvotomy provides effective interventional method in the treatment of infants and children with aortic valve stenosis, including critical aortic stenosis.

P83

Balloon occlusion test of atrial septal defect – its diagnostic value

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Background: Temporary balloon occlusion (TBO) of atrial septal defect (ASD) during cardiac catheterization may imitate the situation after planned surgical or interventional procedure and therefore may be helpful in decision making. TBO of ASD were employed in 3 patients (pts) with different clinical problems.

Material and Methods: Pt No 1 (age 20/12, weight 8 kg) dgn: pulmonary artery atresia with intact ventricular septum and tricuspid insufficiency after previous surgical right ventricle outflow tract reconstruction and B-T shunt with cyanosis (Sat O_2 75%) caused by right to left shunt through ASD. Diagnostic aim of TBO, if closure of ASD would not provoke right heart failure. TBO performed with OBW calibration balloon with simultaneous RV pressure measurements.

Pt No 2 (age 6/12, weight 3.4 kg) dgn: premature infant with borderline left ventricle hypoplasia, coarctation of aorta (after surgical repair in neonatal period and successful balloon angioplasty in 3/12) with congestive heart failure and ventilator dependant respiratory insufficiency caused by left to right (L-R) shunt through ASD II. Diagnostic aim of TBO, if ASD closure would not provoke left heart failure. TBO performed with "wedge" catheter with simultaneous LA pressure measurements.

Pt No 3: age 65, weight 75 kg dgn. ASD with L-R shunt and mitral insufficiency (+++). Diagnostic aim of TBO, if ASD closure would not provoke pulmonary oedema. TBO performed with OBW calibration balloon with simultaneous LA pressure measurement.

Results: In all pts the result of TBO was positive.

Pt No 1: ASD II was closed with 12 mm Amplatzer Septal Occluder (ASO). Sat O_2 rise to 96%, and body weight gain 3.5 kg during 15 months follow up.

Pt No 2: Surgical closure of ASD II (after failure of interventional closure) was performed: weaning from ventilator and decrease of congestive heart failure symptoms were achieved.

Pt No 3: ASD II was closed with 30 mm ASO without any complications.

Conclusions: In certain clinical situations temporary occlusion balloon test of ASD may be an attractive diagnostic tool, helpful in

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decision making when surgical or interventional treatment modalities are of concern.

P84

Residual Potts anastomosis: first case of definitive percutaneous occlusion

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Potts anastomosis was used as a palliative systemico-pulmonary shunt in the 60s and 70s for cyanotic congenital heart disease (CHD). Apart the risk of acquired pulmonary hypertension, its major inconvenient was the difficulties of its surgical control during the complete repair of the CHD. So, many patients are now living with a permeable Potts anastomosis not occluded during the complete repair.

A 30-year-old patient had Potts anastomosis for atypical tetralogy of Fallot in his first year of life. He underwent complete repair at the age of 7, but despite a surgical attempt, the anastomosis could not be closed efficiently. Twenty years later, he developed left heart overload with pleural effusion and atrial flutter. A first attempt of occlusion with a PDA Amplatzer device failed because the oval shape of the defect (13 × 10 mm) and the relative small size of the left pulmonary artery (15 mm), assessed by further CT scan. The definitive procedure was performed under general anesthesia. After puncture of the left humeral artery, a pigtail catheter was pushed in the pre-isthmic aorta in order to control arterial pressure and to repeat angiographies. Through a puncture of the right femoral artery, a extrastiff guidewire was introduced in the ascending aorta. After dilatation of the cutaneous orifice, a covered stent (Medtronic Talente – length: 90 mm, diameter: 24 to 28 mm) was introduced percutaneously and deployed under iterative angiographic control. Despite correct deployment, a small residual shunt remained. A second inflation was performed with a 25 mm low pressure balloon: the shunt definitively disappeared. The percutaneous access was closed with a Perclose device without any complication. Six months later a CT-scan showed the stent in normal position without residual shunt. The left pulmonary artery was not restrictive and the patient was dramatically improved without any left ventricular overload nor signs of cardiac failure. Covered stents can be used only in adult patients, but patients with residual Potts anastomosis are now all adults. Despite the large diameter of the device (22F), a percutaneous access is possible. This new utilization of covered stents offers an elegant solution to the delicate problem of residual Potts anastomosis after surgical correction of cyanotic CHD.

P85

Immediate and early results of transcatheter closure of patent ductus arteriosus using Amplatzer Duct Occluder

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Objectives: To report the immediate and early results, safety and efficacy of Amplatzer Duct Occluder (ADO) in transcatheter occlusion of patent ductus arteriosus.
Material and Methods: From January 2002 to December 2003, 28 patients (26 girls and 2 boys) underwent transcatheter closure of patent ductus arteriosus at the median age of 5.9 yrs and the mean age of 5.8 yrs (range 14 months to 12 yrs) SD 3.3. The median weight of 14 kg and the mean weight of 14.2 kg (range 6.0 to 24 kg) SD 4.7. A lateral view aortogram was performed to determine the morphology of the ductus and to select the size of the device. Follow up evaluation was made by chest-X-ray and echocardiogram at 24 hours, 1 month and 6 months after the device implantation.
Results: 16 patients immediately achieved complete closure. On color Doppler echo within 24 hours, 23 pts (82%) revealed a complete closure, 1 pt (3.5%) showed smoky residual shunts and 4 pts (14%)showed very small residual shunt. After 1 month and 6 months only 1 patient revealed small residual shunt which was not significant. The median diameter of the duct was 3.4 mm. The mean diameter was 3.8 mm (range from 2.0 mm to 8.0 mm) SD 1.6. The device size that were implanted ranging from 4 to 10 mm with the median of 6.0 mm and the mean of 6.7 (SD 1.6).The mean flouroscopic time was 14.1 minutes (range 5.6 to 21 minutes) SD 3.7 mm. The mean procedure time was 78.6 minutes (range 30 to 122 min) SD 25.5. Until recently there was no complications detected in all ptients.
Conclusions: Transcatheter closure of PDA using Amplatzer Duct Occluder (ADO) in children is safe and effective procedure and revealed satisfactory results. It should be a reasonable alternative to surgery.

P86

Systemic hypertension associated to aortic coarctation improves after percutaneous angioplasty

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Background: Systemic hypertension characterize aortic coarctation (AC). Percutaneous transluminal angioplasty (PTA), is a valid therapeutic option for recurrent or native AC. We report our Institutional experience of this procedure and mid-term follow up. Material and
Methods: From June 2002 to February 2003, 8 consecutive patients with AC underwent PTA. Three children, mean age 14 ± 7 months, with recurrent AC following aortoplasty with end-to-end anastomosis technique, were treated by PTA alone. Five adult patients, mean age 32 ± 9 years, with native AC received direct stenting; four had systemic hypertension treated with multiple drugs, one had enlarged ascending aorta. All received diagnosis of AC during cardiovascular standard examination (CSE) and transthoracic echocardiography (TTE). All patients underwent TTE, treadmill test (TT), 24 hours Holter blood pressure monitoring (HBPM), magnetic resonance study before the procedure. The interventional procedures were performed in the standard fashion under general anaesthesia. In adults 13 F Mullins sheath was used and routinely removed surgically. Follow-up was performed with CSE and TTE in adults in addition HBPM, and angio-CT scans were performed.
Results: All the procedures were uneventful. The invasive transisthmus mean gradient in the babies was 56 ± 49 mmHg, the balloon mean diameter was 8 ± 2 mm, final gradient was 0. In adults the mean gradient was 47 ± 12 mmHg; one PS 5014 and four PS 4014 were used, obtaining a mean final diameter of 19 ± 3 mm, the final gradient was 1 ± 2 mmHg. In the patient with the residual gradient (5 mmHg) the stent was not fully expanded on

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purpose. One patient had paradoxical hypertension after the procedure treated with propranolol e.v. At follow up all babies are doing well, the one with HBP normalized the blood pressure in 6 months. All adults were discharged in medical therapy for hypertension and aspirin for 6 months. The TT performed at three months showed normal pattern of the BP; all except one showed a normal 24 hours blood pressure level at the HBPM in therapy washout. All CT scan showed normal stent position and absence of aneurism.
Conclusions: PTA in recurrent AC and HBP can improve systemic hypertension in babies. Native AC in adults can be efficaciously treated with PTA and direct stenting; after the procedure the systemic hypertension improves at the mid-term follow up.

P87
Retrograde use of the Starflex® device
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Background: The Starflex® septal occluder is usually deployed through a long transvenous sheath for closure of ASDs, PFOs and less commonly VSDs. We describe two patients with residual (functional) left to right shunting after surgical palliation and illustrate the successful use of the double umbrella device in these situations using a retrograde transarterial approach.
Patients: Patient 1 was a 15-year-old boy with d-TGA and a large muscular VSD. He was transferred at age 3 years after balloon atrioseptostomy and hemodynamically ineffective pulmonary artery banding. Since systemic pressure in the subpulmonary ventricle due to increased pulmonary vascular resistence was found and the child became progressively cyanotic a palliative Mustard operation without VSD closure was performed. However, after the operation and with further decrease in VSD size the pressure in the subpulmonary ventricle droped to half-systemic with a Qp:Qs of 1,6:1. After balloon sizing (streched diameter 17 mm) and test occlusion, the VSD was closed using a 33 mm Starflex® device through a 10F Mullins long sheath introduced retrogradely through the femoral artery. Patient 2 was a 37-year-old woman with teralogy of Fallot. She was palliated with a 4 mm Waterston-Cooley anastomosis in her first year of life. During subsequent corrective surgery the Waterston-Cooley anastomosis was ligated but found to have recanalised years later. Cardiac catheterization documented a step-up in oxygen saturation from 74% in the right atrium and ventricle to 89% in the right pulmonary artery. The shunt was closed using a 17 mm Starflex® device introduced through a 9F Mullins long sheath via the femoral artery.
Results: There were no complications during retrograde device closure or during follow-up in the two patients reported. Patient 1 showed mild, patient 2 trivial residual shunting through the porous polyester-fabric of the Starflex umbrella immediately after device deployment. No residual shunt was detected in both patient by colour doppler at 3 months follow-up and the percutaneous arterial entry site did not exhibit any sequelae (normal pulses, blood pressures, doppler recordings).
Conclusions: Effective and safe closure of left to right shunting can be achieved by retrograde use of the Starflex® device in selected elder children and grown-ups with congenital heart disease (VSD, aorto-pulm. shunt).

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Evaluation of postnatal myocardial cells proliferation in the pulmonary ventricle submitted to pressure overload: an experimental study
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Objective: When the timing for neonatal arterial switch operation to treat transposition of the great arteries is lost, a pulmonary artery banding (PAB) is required to prepare the left ventricle. Controversy exists regarding the existence and extension of myocyte hyperplasia after the neonatal period in response to the pressure overload imposed by the PAB. This study is aimed at analyzing experimentally the cell proliferation rate in pulmonary ventricles submitted to different types of PAB (continuous and intermittent).
Methods: Twenty healthy 60-days-old goats (beyond the neonatal period) were divided in three groups: control (n = 6, no surgical procedure), continuous stimulation (n = 7, submitted to PAB by surgical implantation of a external device, inflated continuously and progressively for five days), and intermittent stimulation (n = 7, PAB, stimulated intermittently – 12 hours/day, for five days).

At the end of the experiment the goats were sacrificed, the hearts excised and 5 μm thick histological sections from the right ventricle (RV), left ventricle (LV) and ventricular septum were submitted to imunohistochemistry with the monoclonal antibody Ki-67 to identify cell proliferation. Labeled cardiomyocytes and interstitial/vessel cells were quantified microscopically (number/high power field).
Results: The number of proliferating RV cardiomyocytes in the continuous stimulation group was significantly higher when compared to the respective LV and to both ventricles of the controls ($p < 0.05$). When comparing the intermittent stimulation group with the controls, no difference was detected in the rate of proliferating RV cardiomyocytes. Numbers of proliferating RV interstitial/vessel cells were significantly higher in both stimulated groups when compared to the respective LVs and to the controls ($p < 0.05$).
Conclusion: The continuous pressure overload imposed to the RV increased significantly the proliferation of cardiomyocytes and interstitial/vessel cells. The intermittent stimulation in the conditions of the present study was not capable of enhancing the proliferation of cardiomyocytes; however, it caused interstitial/vessel cells hyperplasia. Our study demonstrated that cardiomyocytes are able to reenter the cellular cycle even after they become differentiated, but this process is not so easy as it is for cells belonging to the interstitium and vessels.

P89
One stage correction for aortic arch obstruction and VSD
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Objectives: To evaluate the current outcome and the re-operation rate after one-staged correction strategy for IAA + VSD and CoA + VSD adopted since 11/1999.
Methods: From 11/99 until 01/04 a total of 29 consecutive patients underwent definitive repair of aortic arch obstruction + VSD.

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There were 15 patients in the coarctation (CoA) group and 14 patients in the interrupted aortic arch (IAA) groups. Mean age at operation was 12 days (3–92 d), mean weight 3.1 kg (2.1–7.3), 12 patients were under 3.0 kg. Two of IAA were diagnosed as type A, 10 type B and 2 type C. Associated anomalies in IAA group included a large ventricular septal defect in all, d-TGA in 1, APW in 2 and PTA in two patients. Three patients in IAA and 2 in CoA group demonstrated left ventricular outflow tract obstruction (LVOTO). In all patients a selective brain and coronary perfusion during extended aortic arch anastomosis was used. Mean follow up consists of 2.5 ± 0.8 years.

Results: There was no early, no late death, no neurologic complications postoperatively. Selective brain and coronary perfusion time was 34 ± 13 min. Twenty patients required secondary sternal closure at mean 1.7 day postoperatively. Four patients from IAA group developed aortic arch re-stenosis. Balloon dilatation was done successfully in three, one patients got additionally aortic arch stent. One patient with d-TGA underwent RVOT reconstruction 7 months after the initial repair. All 29 patients are asymptomatic.

Conclusions: One-stage complete correction is feasible with good surgical and functional outcome in all newborns with aortic arch obstruction and even with very complex additional pathologies. Selective brain and coronary perfusion during the aortic arch reconstruction reduces the neurological complications and reduced the morbidity. IAA type B is a risk factor for development of recurrent aortic arch obstruction.

P90

Truncal block rotation – a new option in double outlet right ventricle or transposition of the great arteries/ ventricular septal defect and left ventricular outflow tract stenosis

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Background: The classical treatment option in DORV or TGA/ VSD and left ventricular outflow tract (pulmonary or subpulmonary) obstruction is the Rastelli procedure. A long intraventricular tunnel bearing the risk of subaortic stenosis and a homograft conduit from the right ventricle to the pulmonary artery are potential problems of this procedure. The aortic translocation (Nikaidoh) is an alternative method reported in some series, coming closer to a more anatomic repair. However,tension on the right coronary artery, affording its anterior translocation, and the lack of a pulmonary valve and a separate backwall of the pulmonary artery are disadvantages of this operation.

Method: A 5 yr old boy with DORV, transposition of the great arteries, subpulmonary VSD and subpulmonary stenosis was referred to our unit after 5 palliative procedures which resulted in a common atrium, right classic Glenn and a stenotic pulmonary artery on the left side. We performed a take-down of the Glenn and a complete repair using the principle of truncal block rotation without a homograft valve. Both great arteries were divided, right and left coronay artery were excised and mobilized. The complete truncal block (aortic and pulmonary root en block) was excised including the conal septum, rotated by 180° and reimplanted. So the original aortic valve is positioned above the left ventricle. The VSD was closed and the coronaries reimplanted. Aorta and aortic root were reanastomosed. The pulmonary arteries were reconnected and reanastomosed to the pulmonary root anterior to the aorta. The original pulmonary valve, now located over the right ventricle, was bicuspid and was mobilized by a commissurotomy.

Results: The postoperative course was uneventful. He was on the ventilator for 8 hours, transferred to the normal ward on day 3 and discharged on day 13.There is no significant gradient across the pulmonary valve and no residual shunt.

Conclusions: In consent with other authors (Yamagishi et al., Haas et al.) we have demonstrated that the principle of truncal block rotation has the potential of complete anatomic repair in DORV or TGA/VSD and LVOT obstruction, especially when the LVOT stenosis is predominantly subvalvar.

P91

Surgical repermeabilization of occluded pulmonary arteries in patients with congenital heart disease

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This study was undertaken to determine outcomes and best strategies for treatment of occluded pulmonary branches in patients with congenital heart disease.

Between 1998 and 2002, occlusion of a previously patent pulmonary branch was established in 23 patients. Data were obtained retrospectively. Diagnosis were: pulmonary atresia and ventricular septal defect in 11, tetralogy of Fallot in 6, others forms of pulmonary stenosis or atresia in 6. Median age and weight at diagnosis were 9 years (range 6 days–43 years) and 24 Kg (range 2.6–60). Fourteen patients had had a previous surgery. The occluded pulmonary branch was visualized at angiography by wedge injection or injection in collateral circulation. Left pulmonary branch was occluded in 20 patients, right pulmonary branch in 3. Criteria for repermeabilization were: estimated duration of occlusion <6 months and ratio occluded/controlateral branch >0.2.

Twelve patients fulfilled these criteria and underwent pulmonary branch reconstruction at a mean interval of 2 months (range 6 days–6 months) from evidence of occlusion. Six patients had pericardial patch reconstruction, 3 termino-terminal anastomosis, 2 thrombectomy, 1 Blalock-Taussig shunt. There was 1 late death. At a median follow-up of 4 years (2 months–5 years), all patients underwent cardiac catheterization: in 8 patients the reconstructed branch was patent, in 3 re-occluded. Hypoplasia of the occluded branch was reversed in 6 patients.

Our data show that, in selected patients, reconstruction of an occluded pulmonary branch can restore pulmonary vascularisation and reverse branch hypoplasia. Strict surveillance is mandatory to prevent pulmonary branch loss.

P92

Long-term follow up after post surgical diaphragmatic paralysis

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Objectives: Phrenic nerve injury resulting in diaphragmatic paralysis (DP) is a rare complication after cardiac surgical procedures in children. However, DP is characterized by an increased postoperative morbidity including extended need for artifical ventilation, increased incidence of lung infections and a longer hospital stay. Some children can only be weaned from the respirator after

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diaphragmatic plication. Little is known about lung function and cardiopulmonary exercise capacity in children with DP in long-term follow up.
Methods: During a period of 10 years (1990–2000) 63 (median age 7 month; range 2 days–17 years) out of 3649 children undergoing cardiac surgery for congenital heart defects showed postoperative DP (prevalence 1.7%). Nearly half of the children with DP developed long-term respiratory failure and required diaphragmatic plication. The patients who required diaphracmatic plication were significantly younger (median age 6 months vs. 4 years). We examined lung function and exercise capacity in children with DP in the postoperative follow up in comparison to a control group (CG) of children undergoing cardiac surgery without DP.
Results: There were no significant differences in age, height or body weight between the DP and control group. Lung function analyses in DP patients showed reduced lung function with lower values for vital capacity (VC), forced exspiratory volume (FEV1) compared to controls. Exercise testing in the DP group showed a lower level of cardiopulmonary exercise capacity compared to controls. There was no significant difference in exercise capacity or maximal oxygen uptake between children with and without diaphragmatic plication.

	DP (n = 16)	CG (n = 90)	p
Max. exercise capacity (W/kg) (median/quartiles)	2.0 [1.9/2.5]	2.4 [1.8/2.9]	NS
$VO2_{max}$ (ml/kg/min) (median/quartiles)	23.3 [21.4/29.7]	30.7 [24.4/37.6]	0.006

Conclusions: Postoperative diaphragmatic paralysis is a rare complication after cardiac surgery in children. Children with DP show, in addition to the prolonged initial hospital stay, an increased incidence of lung infections and a longer duration of ventilation. These children also present reduced lung function in the long-term follow up. Diaphragmatic plication has no negative influence on the long-term outcome concerning lung function and exercise capacity.

P93

Revision of atriopulmonary Fontan to total cavo-pulmonary connection: short term results

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Objectives: Revision of atriopulmonary Fontan to Total Cavopulmonary Connection is carried out for haemodynamic and electrophysiological indications. We present our experience of converting patients to total cavopulmonary connection (TCPC) with and without arrhythmia surgery.
Methods: Between 1997–2001 fourteen patients (mean age 20.5 ± 7.9 years) underwent conversion surgery 13 ± 3.7 years after atriopulmonary Fontan. The indication was persistant arrhythmia, obstruction to the pathways or poor functional ability. The preoperative NYHA functional class was II in 2, III in 6 and IV in 6. Four underwent intracardiac lateral tunnel whilst 10 received extracardiac TCPC with right atrial cryoablation.
Results: No mortality occured. One developed conduit thrombosis in the immediate postoperative period requiring immediate replacement, another required redo surgery for presumed endocarditis. Average hospitalisation was 18.5 ± 9 days; chest drains were removed on day 6 ± 6. At follow-up (mean 22 months, range 1–46 months) late atrial arrhythmias had recurred in 2/4 patients with intracardiac TCPC (without ablation) and 1/10 with extracardiac TCPC with ablation. All patients are in NYHA class I or II.
Conclusions: Fontan conversion with or without arrhythmia surgery can be achieved with low mortality and an improvement in NYHA class. Medium term results suggest that concomitant arrhythmia surgery reduces the incidence of late arrhythmias.

P94

Somatic growth and neurodevelopmental risk factors in children 5 to 8 years after anatomical correction of transposition of the great arteries (TGA)

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Objectives: Transposition of the great arteries (TGA) is congenital, potentially lethal heart disease. Arterial switch operation (ASO) restore normal anatomical and hemodynamic state of the circulatory system. In our institution ASO is the therapy of choice for patients with TGA since 1991. Between July 1991 and July 2002 over 350 children underwent ASO. We try to identify potential risk factors influencing late outcome after ASO, especially somatic growth and neurodevelopmental disturbances.
Methods: The group consists of 100 unselected children, in the age 5 to 8 years, after ASO, performed in our institution from July 1991 to July 1996. There were three groups of patients: I – simple TGA, II – TGA + VSD, III – TGA + AAA. Parameters of somatic development (weight, heigh) and neurodevelopmental status (IQ, harmony of development, visual-motor coordination) were evaluated. A multivariable analysis was applied to study the impact of risk factors on somatic and neurodevelopmental outcome parameters. We assumed the following risk factors: 1. low birth weight (<3000 g), 2. low Apgar score (<7 points), 3. severe circulatory perioperative insufficiency: IV and V in modified by Kirklin NYHA classification, 4. age at operation: over 7 days (group I and III), over 14 days (group II), 5. aortic cross clamping time over 80 min, 6. peri- and postoperative complications.
Results: In our study group low birth weight was statistically significant related to low weight (p = 0,028). Children with birth weight 3000–4000 g had four times higher chance to have normal weight in the age 5 to 8 years, than children with birth weight lower than 3000 g. In a group 7 to 8 years old children intelligence (based on IQ assessment) was statistically significant (p = 0,045)correlated with birth weight. In a group 5 to 8 years old Apgar (p = 0,059) and complications (p = 0,058) were in a border of statistical significance. Children with Apgar score above 7 points had five times higher chance to obtain IQ in normal range than children with Apggar score below 7. In this group of children, where complications appeared, risk of IQ below normal range for age was four times higher.
Conclusions: The most important for good somatic growth and neurodevelopmental outcome of children, 5 to 8 years after anatomical correction of TGA, is their birht weight over 3000 g and good condition at the moment of birth.

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Post-operative chylothorax contributes important morbidity but not mortality after cardiovascular surgery in children

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We sought to determine the incidence, risk factors, clinical spectrum and impact on outcomes of chylothorax in children undergoing cardiovascular surgery. Medical records were reviewed, and factors associated with risk of chylothorax and the amount and duration of drainage were sought. From 9/2000 to 12/2002, there were 48 cases of chylothorax associated with 1,329 surgeries, for an incidence of 3.6% (95% CI, 2.6% to 4.6%). Cases were not significantly younger at surgery ($p = 0.47$), but had smaller body surface area ($p = 0.02$). Mortality was similar ($p = 1.00$), but cases had longer post-operative hospital stays (mean 28 vs. 15 days; $p = 0.003$). There were significant differences in incidence between surgeons. For cases, the median age at surgery was 5.3 months (range, 1 day to 17.8 years). Downs syndrome was noted in 6 patients and 6 additional patients were diagnosed with other syndromes. Surgery was by median sternotomy in 98%. Diagnosis of chylothorax was made at a median of 6 days (range, 1 to 50 days) after surgery, with bilateral chest tube drainage in 55% and associated pericardial drainage in 15%. Fluid analysis showed the presence of chylomicrons in 98%, with median triglyceride level of 1.38 mmol/L and white cell count of 1,753/mL. Nutritional management strategies included change in formula or diet in 85%, with 10 patients taken off enteral feeds and 12 given parenteral nutrition. Medical management included diuretics in 90% and octreotide in 13%. Surgical management included thoracic duct ligation in 4 and pleurodesis in 1 patient. During management, 54% required reinsertion of chest tubes. There were 3 deaths, none attributable to chylothorax. Other morbidity was high, with an episode of infection in 46%, respiratory complications in 46% and thrombosis in 35%. Median time from surgery to final removal of chest tubes in survivors was 16 days (range, 5 to 315 days). The only independent factor associated with increased duration of drainage was cavopulmonary shunt surgery ($p = 0.003$). Of note, the incidence of chylothorax with cavopulmonary shunt surgery was 7%. Chylothorax is an important complication after cardiovascular surgery in children, and contributes to considerable morbidity and increased length of hospital stay, particularly in smaller patients and those undergoing cavopulmonary shunt surgery.

P96

Surgical closure of secundum ASD during infancy: clinical considerations, dilemma, and outcome

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Background: Atrial septal defects are usually asymptomatic and are closed surgically or by a catheter-implanted device in preschool age. When symptoms are seen in infancy they are usually due to combined clinical situations and management at this age is debated. Aim of study: To report our experience with a large group of infants undergoing surgical closure of Secundum ASD in order to clarify their clinical course, considerations, and outcome.

Methods and Results: Surgical and medical records of children younger than 12 months of age undergoing surgical closure of Secundum ASD during a 20 year period (1983–2003) were reviewed retrospectively. Over this period, 50 infants underwent surgery in two medical centers. There were 27 (54%) male patients. The mean age at surgery was 220 ± 84 days (mean ± SD). Primary diagnosis was isolated ASD in 39 patients, ASD with small insignificant PDA in eight patients, and ASD with anomalous drainage of one pulmonary vein in three patients. Congestive heart failure and failure-to-thrive were noted in the majority (34/50, 68%) of the patients. Associated extra-cardiac medical conditions were common. Genetic abnormalities were seen in 18 (36%) patients, prematurity and chronic lung disease in 12 (24%) and omphalocele in three (6%) patients. Follow-up period was 56.3 ± 57.4 (mean ± SD) months and outcome was favorable regarding weight gain and cessation of symptoms in 32 (64%) patients.

Conclusion: Symptomatic ASD during infancy is rare and usually associated with extra-cardiac conditions. Although management at this age is debated, we have found that surgical closure is of benefit in the majority of patients.

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Successful treatment of chronic pleural effusion following extra-cardiac Fontan by intravenous Cyklokapron®

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Objectives: Report successful treatment of chronic pleural effusion after extra-cardiac Fontan with Gore-tex conduit.

Method: A 6 year old boy with Ivermark syndrome, mesocardia, asplenia, complete atrio-ventricular septal defect, pulmonary atresia and persistent left superior vena cava, underwent left and right cavo-pulmonary bidirectional connection at 8 months of age. At 6 year, modified Fontan was completed with extra cardiac Gore-tex tube graft interposition between inferior vena cava and right pulmonary artery. After 3 months pleural effusion persisted despite strict chylothorax diet, hydric restriction and diuretics. A cardiac catheterization showed anatomically and hemodynamically good Fontan procedure. Leaking Gore-tex graft due to increased fibrinolysis was the suspected diagnosis. Pleural effusion and blood analysis confirmed increased fibrinolysis and leakage of low molecular weight proteins across the Gore-Tex graft. Intravenous administration of Cyklokapron® (tranexanic acid) (25 mg/kg every 9 hours × 3) was initiated for 24 hours. The fibrinogen deposition on the walls of the graft was successfully monitored by pre and post therapy nuclear scan with fibrinogen antibody marked with technicium 99 (FBDTc 99).

Results: Pleural effusion disappeared and did not recur since 4 months; the diuretics were weaned and normal diet reinstored. At the last blood test, fibrinogen level was normal 3.9 g/l along with Ddimers (0.32 μg/ml).

Conclusions: As described previously in modified Blalock-Taussig shunt (high blood flow Gore-tex shunt), Gore-tex leakage may be due to absent fibrinogen deposition secondary to unusual fibrinolysis. This hypothesis should also be considered in the context of low flow of a post Fontan patient. Intra-venous Cyklokapron® is an effective therapy which can be monitored with innovative nuclear markers. Finally an other alternative to Gore-tex material should be considered in extra cardiac Fontan.

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Thoracotomy scars have a more negative impact than sternotomy scars

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Objectives: To evaluate patient attitudes sternotomy and thoracotomy scars following surgery for congenital heart disease.
Methods: Two hundred and ninety nine patients were randomly selected from population of adults with congenital heart disease. They underwent structured interview by a trained nurse.
Results: Two hundred and one patients (101 female) had a scar from cardiothoracic surgery. One hundred and five (54 female) had undergone sternotomy, 36 (17 female) thoracotomy and 60 (30 female) had both scars. Comparable proportions of each group reported that they did not like or hated their scar (23/105 (22%) sternotomy, 9/36 (25%) thoracotomy, 17/60 (28%) both scars). Significantly more patients stated that they where embarrassed by and/or their choice of clothing was affected by a thoracotomy scar (20/36, 56%) than those with a sternotomy scar (36/105, 34%), $p = 0.04$. A sternotomy scar alone is also less likely to cause embarrassment and/or affect choice of clothing than having both scars (36/105 (34%) vs 34/60 (57%), $p = 0.008$). Contrary to previous reports a significant number of males (40/100, 40%) were concerned by the impact of their scar on daily life. Neither age at surgery (range 0–30 years, median 7 years) nor age at interview (range 17–60, median 31 years) significantly affected attitudes to the scar.
Conclusions: Surprisingly adults who have undergone surgery for congenital heart disease are more likely to have a negative attitude to a thoracotomy than a sternotomy scar. A sternotomy scar also has significantly less negative impact than having both scars. Patients who will eventually need a sternotomy for correction or definitive palliation should also have their initial palliation (such as a shunt) carried out through a sternotomy if surgical risk is unaffected. Before a change in surgical approach is considered based on patient preferences, the acceptability and psychological impact of the different scars following surgery needs formal study.

P99

Cleft mitral valve: new morphologic findings and surgical results

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Background: Cleft mitral valve (MV) is a relatively uncommon lesion morphologically different from an atrioventricular (AV) canal defect. The left AV junction is normal with a posterior leaflet comparable in size to the dimension seen in a normal MV and normally positioned left ventricular papillary muscles. Controversy exist with the constant presence of chordal attachments from the cleft to the ventricular septum and the anterior position of the cleft.
Methods and Results: A retrospective review identified 11 patients operated upon with cleft MV, normal ventriculo-arterial connections and 2 well-developed ventricles. The median age at surgery was 5,5 years (1,3 to 10,6 years). Nine patients had a significant mitral regurgitation (MR) and 2 had subaortic obstruction. In 4 cases with MR, intraoperative examination revealed no chordal attachments of the cleft to the ventricular septum. In all the patients, the cleft was directed toward the aortic root. Mitral valve was repaired in all the cases. A direct and complete suture of the cleft was accomplished in 8 patients. In 2 cases with moderate to severe MR, the suture of the cleft was impossible because of retraction of both parts of the anterior MV leaflet. Augmentation of the anterior leaflet with a pericardial patch was accomplished in one case whereas the other patient had an Alfieri-type procedure. A mitral annuloplasty was associated in 3 cases. In 2 cases with subaortic stenosis, it was necessary to resect chordal attachments to the ventricular septum. During the median follow-up period of 2,5 years (0 to 7,8 years), one patient was reoperated for mitral stenosis associated with aortic valve stenosis. Follow-up echocardiography demonstrated no significant MR nor subaortic obstruction in all cases and a moderate MV stenosis in 2 patients.
Conclusions: Intraoperative examination demonstrated the anterior direction of the cleft MV in all the patients and the absence of chordal attachments to the ventricular septum in several cases with MR. Surgical repair of the cleft always seems possible. However, with long standing MR and older age at repair the direct suture of the cleft may not be possible or effective. Surgical repair should then be advocated in younger children even without symptoms.

P100

Congenital coronary artery fistulas in children and adults: diagnosis and surgical management of seventeen patients

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Objectives: Coronary artery fistula (CAF) is a very rare heart defect, but it is one of the most common coronary malformations and is being diagnosed with increasing frequency with widespread use of selective coronary arteriography.
Methods: A retrospective study has been performed during 10 years on all patients who were diagnosed with coronary artery fistula from June 1990 to December 2000.
Results: Seventeen consecutive patients (mean age 31 years, ranging between 1 and 65 years) 11 male and 6 female were diagnosed during ten years. Thirteen of the patients were treated surgically and 4 patients were treated medically and are still under observation. Eight (47%) patients were below 20 years of age. Age at the time of diagnosis and surgery was between 6.5 months to 60 years (mean 27.25 years). In 13 patients (76.5%) the origin of fistula was the left coronary artery (LCA), in one patient (5.9%) was the right coronary artery (RCA), in one patient (5.9%) was the ascending aorta directly and two (11.8%) presented with fistulas originating from previous bupass grafts. The RCA fistula terminated into the right ventricle. Among fistulas from LCA terminations were localized in the main pulmonary artery (6 patients), right ventricle (4 patients), Right atrium (2 patients, and into the coronary sinus (1 patient). Associated pathologic findings were aneurysm of fistula in eight patients, coronary artery disease in 2 patients, and mitral valve stenosis and regurgitation in 2 patients due to rheumatic heart diease. Surgical closure of the fistulas were successful in all operated patients. Neither hospital mortality nor severe complications occurred in this group of patients. The mean follow up interval was 47.5 months (ranging between 5.5 and 84 months).
Conclusions: In the presence of symptoms of congestive heart failure, significant left to right shunt and arrhythmias, elective closure

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of coronary artery fistula is generally accepted; wheras the indication is more controversial in asymptomatic patients.

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Preoperative symptomatic status does not predict early postoperative pulmonary hypertensive complications in complete atrioventricular canal

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Based on the assumption, that symptoms of heart failure in atrioventricular canal (CAVC) were due to increased pulmonary blood flow, the hypothesis was tested whether symptomatic infants, when compared to oligosymptomatic infants would have less pulmonary hypertensive (PH) complications in the early postoperative period.

A retrospective study was performed covering an 8-year period ending in May 03. Patients with CAVC (with or without trisomy 21) and age at surgery <1 year were grouped according to the presence of preoperative signs of heart failure as assessed by clinical criteria, feeding behaviour and need for medication. Postoperative pulmonary artery pressure (PAP) was invasively measured and expressed as percentage of the also invasively measured arterial pressure. In both groups, PAP-levels were compared as was the occurrence of PH-cisis, definded as sudden increase in PAP of >10 mmHg.

Of 64 patients, 22 were assigned to the symptomatic group, 42 were considered to be oligosymptomatic. The 2 groups did not differ with regard to defect size (mean of 14 mm in each group), severity of tricuspid or mitral regurgitation, ventricular dimensions, perioperative variables such as bypass time and aortic cross clamp time. Symptomatic patients were slightly younger at surgery (mean of 14 weeks versus 17 weeks). The 2 groups did not differ statistically with regard to early postoperative PAP-levels (31% versus 27% of simultaneous arterial pressure), final PAP-level (prior to removal of PA-catheter), number of episodes of PH-crisis/patient (mean of 15 in each group), number of patients in each group with PH-crisis episodes (80% versus 90% respectively). There was no difference between the groups with regard to duration of intubation, need for inotropic support, lenght of intensive care stay (6.6 days in asymptomatic versus 7.7 days in symptomatic infants), hospital stay and overall outcome.

Conclusion: Early postoperative PAP-levels were not different in symptomatic infants with preoperative high pulmonary blood flow when compared to preoperatively oligosymptomatic infants.

P102

Median sternotomy for reoperation of the distal aortic arch in post-coarctectomy patients

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Introduction: This study reports our experience using a median sternotomy approach for redo-surgery in adolescent and adult post-coarctectomy patients with complex residual aortic gradients and/or aortic arch aneurysms.
Methods and Results: Thirteen post-coarctectomy patients (8 males, 5 females, aged 14 to 38 years) underwent reoperation for recoarctation, hypoplastic aortic arch and/or aortic arch aneurysm at our clinics between December 1994 and December 2003. Initial coarctation repair was performed by end-to-end anastomosis in 6 patients, patch aortoplasty in 3, subclavian flap aortoplasty in 3 and tube interposition in 1. Mean age at reoperation was 26 ± 9.1 years. All patients were hypertensive before reoperation and were on antihypertensive medication. Patients were reoperated for recoarctation and/or hypoplastic aortic arch in 7 patients, distal aortic arch aneurysms in 3 and a combination of the two in 3. A median sternotomy was used in all patients. Mean cardiopulmonary bypass time was 195 ± 53 minutes. Reoperation was performed by tube graft interposition in 7 patients, patch aortoplasty in 5 and resection and end-to-end anastomosis in 1. Aortic cross clamping was used in 11 procedures (median 71 minutes, range 32–203 minutes). Total circulatory arrest was necessary in 5 procedures for a median of 29 minutes (range 20–48 minutes). In 8 patients circulatory arrest with selective antegrade cerebral perfusion was used (median 53 minutes, range 22–83 minutes). There were no deaths. Four patients developed postoperative hoarseness due to recurrent laryngeal nerve paralysis. Patients were followed for a mean duration of 3.0 years. Six patients remained hypertensive after reoperation. All patients underwent postoperative echocardiography and/or magnetic resonance imaging of the aorta. In all patients satisfying results were obtained: no significant residual aortic gradients or aortic aneurysms were found.
Conclusion: Median sternotomy can achieve extensive mobilization of the thoracic aorta. This approach allows repair of hypoplasia of the transverse aortic arch, distal aortic arch aneurysms and coexisting cardiac anomalies and can be performed with low morbidity and mortality.

P103

Rastelli operation for TGA, VSD and PS: a follow up study

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Objective: This retrospective follow up study aimed to describe the results of the Rastelli operation for transposition of the great arteries, ventricular septal defect and pulmonary stenosis (TGA, VSD, PS) and to assess short-term as well as long-term morbidity- and mortality-outcomes.
Methods: 23 consecutive patients were selected from paediatric and adult cardiology-databases from our institution in the period May 1973–November 2002. Survival curves and event-free periods were obtained by using the Kaplan-Meier method.
Results: There were 20 males and 3 females in the studygroup. Median age at operation was 3 years. One patient was lost to follow up, yielding a 96% complete follow up (median follow up 10,6 years, max. follow up 29,6 years). There were 4 hospital deaths (17%:95% CL = 2–33%), all before 1997 and 4 late deaths (22%:95% CL = 10–90%) mostly due to fatal rhythm disturbances (ventricular fibrillation). Reoperation was needed after a mean period of 3 years. In 11/19 patients a conduitstenosis was diagnosed, three times a residual VSD. 2 patients needed pacemaker implantation because of arrhythmias. Overall survival after the Rastelli operation was 83%, 77%, 71%, 62% and 51% at 1 month, 5, 10, 15 and 20 years respectively. Freedom from reoperation or death was 52%, 15% and 15% at 5, 10 and 15 years respectively.
Conclusion: Although short-term outcome after Rastelli operation is reasonable to good, late outcome in comparison with arterial switch operation (ASO) for (forms of) TGA is disappointing. ASO is the procedure of choice for all patients with all variants of TGA, unless a PS is not amenable to relief and requires a Rastelli operation as an alternative.

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P104

Surgical repair of congenital mitral valve insufficiency in infants and children, long term results

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Background: Mitral valve replacement in children with congenital mitral valve insufficiency is associated with a non-negligible complication rate. Conservative mitral valvar repair has therefore become the preferred treatment modality. Immediate results have been promising, while only few long-term studies have been reported.
Material and Methods: Between 1994 and 2001, 31 pediatric patients (9 boys and 22 girls) with an average age 6.0 ± 4.5 years (2 months to 14 years) with congenital mitral valve insufficiency underwent mitral valve repair, including mitral annuloplasty ring. Ten patients (32%) had simultaneous additional surgical procedures. The majority of patients were followed by serial echocardiography examinations every 6 to 12 months.
Results: There was 1 hospital death, 3.1%. Due to a failed mitral valve repair mitral valve replacement was performed on the 5th postoperative day, but the valve thrombosed one month later. One death, for unknown reasons, occurred during the follow up, 13 months after the mitral valve repair. During follow up 2 patients (6.7%) required mitral valve replacement at 5 days and at 7 years after the mitral valve repair. The remaining patients have shown good mitral function without or with only minimal mitral regurgitation. Actuarial survival at 5 years was 93.4% and freedom from reoperation at 5 years was 96.8%
Conclusions: Mitral valve repair in children with congenital mitral valve insufficiency has excellent immediate results with low operative risk and satisfactory long-term results and should be the preferred surgical treatment modality.

P105

Use of Dexmedetomidine after congenital cardiac surgery

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Aim: The aim of this study was to assess the efficacy and safety of Dexmedetomidine as a sedative and analgesic agent during the postoperative course of children undergoing cardiac surgery.
Material and Methods: Eighteen patients in the cardiac ICU, 7 females and 11 males, aged 2 to 20 years (mean 11.4) were included. Operations performed: 4 Heart transplants, 3 ASD, 3 Coarctation, 2 RV to PA conduits, 1 VSD, 1 TOF, 1 Aortic valve replacement, 1 subaortic membrane, 1 supra-pulmonic stenosis and 1 Ross operation. Dexmedetomidine was started post-extubation in 16 patients (range 1–4 hrs, mean 1.2) and during mechanical ventilation in 2 (1 and 12 hrs pre-extubation). Blood pressure (BP), heart rate (HR), respiratory rate (RR) and ABGs were recorded hourly. Sedation was evaluated with a 0 to 3 score scale (0 – non sedated, calm, 1 – mild, 2 – moderate, 3 – severe sedation) and pain was assessed with the FLACC and Numeric Visual Analog Scales (0–10 score).
Results: Starting Dexmedetomidine dose was 0.1 to 0.3 mcg/kg/h (mean 0.18). The maintenance dose ranged between 0.1 and 0.7 mcg/kg/h (mean 0.3) and infusion continued for 9 to 26 hrs (mean 17.2). In 5 (3 Coarctation and 2 RV to PA conduits) Dexmedetomidine was also used for controlled hypotension (Baseline mean SBP 128 mmHg, during Dexmedetomidine mean SBP 100 mmHg). Overall the mean changes in BP, HR and RR were not clinically significant. The lowest mean SBP occurred 5 hrs after starting the infusion (−5.4% from baseline) and correlated with higher dose. In 1 patient Dexmedetomidine was held for 1 hour because of a 15% drop in the SBP. No respiratory depression was noted throughout, and no reintubation occurred. Desirable sedation was achieved in most patients with an average score of 1.45 (mild). One patient had severe somnolence lasting 2 hrs at 0.5–0.7 mcg/kg/min. Pain level ranged from 0–8 with an average score of 1.64 (mild). Six patients required each one time Fentanyl or Morphine for pain. Table 1 shows rest of hemodynamic comparisons.

	Baseline (mean)	Max. on Dexmedetimidine (mean)	Min. on Dexmedetomidine (mean)	Mean change %
SBP (mmHg)	86–129 (113)	105–154 (125)	69–108 (92)	−3.7
DBP (mmHg)	42–69 (56)	52–78 (66)	32–62 (48)	−0.7
MBP (mmHg)	63–94 (76)	75–98 (86)	44–76 (64)	−4.0
HR (bpm)	65–150 (112)	90–152 (117)	66–121 (95)	−5.5
RR (bpm)	10–34 (24)	21– 64 (32)	10–26 (18)	+1.2

Hemodynamic comparison before and during Dexmedetomidine.

Conclusions: Dexmedetomidine is a new sedative agent that can be used during the postoperative course of pediatric patients undergoing cardiac surgery. It provides an effective means of sedation and analgesia both in intubated and non-intubated patients, without significant hemodynamic or respiratory side effects. Furthermore it can provide controlled hypotension in a subset of patients.

Session 9: Cardiac Imaging

P106

Biventricular diastolic response to dobutamine stress in young patients with pulmonary regurgitation (PR) after repair for tetralogy of Fallot (TOF) using Cardiac Magnetic Resonance Imaging (CMR)

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Introduction: Biventricular systolic and diastolic dysfunction are known problems after repair of TOF. CMR combined with physical stress has been used to study both. Assessment of diastolic changes using physical stress has not been successful. Pharmacological stress with an inotropic drug may be a good alternative. With PR diastolic filling of the RV ccurs through 2 valve orifices. CMR provides a unique tool to quantify valvular flow. Post-processing of data allows 2 flow measurements to be combined into 1 ventricular filling curve.
Methods: 23 patients operated for TOF (mean age 16,9 ± 4,7 y, mean age at repair 1,0 ± 0,6 y) underwent CMR at rest, and during low dose dobutamine stress (max. dose 7,5 γ). Flow measurements

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were performed using a PVC MR technique. Time-volume flow curves were derived from these data. RV time-volume change curves were reconstructed by summation of tricuspid and pulmonary flow, assuming changes in RV volume are equal to the sum of flow entering and leaving these two valve planes. The same principles were used for the LV. Indices of diastolic function were derived from the ventricular time-volume change curve.
Results: Diastolic function indices at rest and during stress for RV: peak early filling rate (ml/s) (PeFR) 461 ± 157 vs 520 ± 175 ($p < 0,01$), peak atrial FR (ml/s) 250 ± 145 vs 365 ± 177 ($p < 0,01$), time to PeFR (ms) 107 ± 23 vs 92 ± 21 ($p < 0,05$), decelleration time (ms) 222 ± 57 vs 271 ± 87 ($p < 0,05$). For the LV: PeFR 419 ± 118 vs 493 ± 129 ($p < 0,01$), PaFR 127 ± 56 vs 183 ± 98 ($p < 0,01$) and ttPeFR 127 ± 20 vs 113 ± 25 ($p < 0,05$). No changes in ttPaFR, atrial filling fraction (FF) and early FF (filling during first 1/3 of diastole). Time-volume change curves showed enddiastolic forward flow (EDFF) in the pulmonary artery in over 70% of patients (at rest 6,2% ± 5,9 versus 6,1% ± 5,0 with stress (n.s.)).
Conclusions: CMR offers a unique tool to study ventricular filling characteristics in patients with PR. A pharmacological stress protocol restricts motion artefacts and allows diastolic biventricular functional measurements. RV and LV diastolic function are abnormal after TOF. Changes with stress are compatible with abnormal ventricular relaxation. In over 70% of patients EDFF was present, confirming RV diastolic restrictive function.

P107
Ultrasonic left and right ventricular strain rate and strain indices: are they affected by percutaneous closure of an atrial septal defect?

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Background: Ultrasonic strain rate and strain are new indices which reflect regional myocardial function. However their load-dependency needs to be further assessed. Aim: to evaluate the influence of percutaneous closure of an atrial septal defect (ASD) on left and right ventricular ultrasonic deformation indices 'strain rate and strain'.
Methods: 12 patients (age 2,8–19,9 yrs) who underwent percutaneous closure of an ASD (Amplatzer device n = 10, Helex device n = 2) were studied. Atrial left to right shunt was significant in all the patients (mean Qp/Qs = 2,1 ± 0,5/1). A Colour Doppler Myocardial Imaging (CDMI) study was performed before (pre) and 24 hours after (post) ASD closure. Peak systolic velocity (Vel), peak systolic strain rate (SR) and systolic strain were measured and high frame rate (>120 frames/sec) acquisition was used. For longitudinal function assessment data were obtained from the base and the midsegments of the right ventricular (RV) free wall and the interventricular septum, for radial function from the left ventricular posterior wall (LVPW). The results were compared before and after ASD closure. The parameters were also compared with those from 30 age-matched normal controls.
Results: The results are shown in the Table. In the patients with ASD the RV peak systolic Vel were significantly higher, but returned to nomal values after ASD closure. RV peak systolic SR and systolic strain were not different compared to normals and remained unchanged after ASD closure. In the septum there is a significant decrease in peak systolic Vel and systolic strain, whereas peak systolic SR values remain unchanged. For the radial LV function higher peak systolic Vel were recorded in the ASD patients which returned to normal values after ASD closure. Radial systolic strain and peak systolic SR was normal and did not change after ASD closure.

	RV base	Septum base	LVPW
Peak syst Vel ASD pre cm/sec	12 ± 2⋆	5 ± 1	4 ± 0.8⋆
Peak syst Vel ASD post cm/sec	9 ± 4#	3 ± 1#	3 ± 0.1#
Peak syst Vel normals cm/sec	8 ± 1.8	4.8 ± 0.8	3.7 ± 0.8
Peak syst SR ASD pre/sec	3.1 ± 2	2.3 ± 0.7	3.8 ± 1.5
Peak syst SR ASD post/sec	3.0 ± 1.9	2.4 ± 1.9	3.9 ± 0.9
Peak syst SR normals/sec	2.4 ± 0.7	1.9 ± 0.7	4 ± 0.9
Syst strain ASD pre%	39 ± 17	29 ± 7⋆	54 ± 12
Syst strain ASD post%	42 ± 8	22 ± 7#	57 ± 9
Syst strain normals%	36 ± 10	24 ± 6	59 ± 11

⋆$p < 0.05$ ASD pre vs normals; #$p < 0.05$ ASD post vs ASD pre. Vel = velocity, SR = strain rate, LVPW = left ventricular posterior wall.

Conclusion: In patients with ASD, RV peak systolic myocardial velocities are significantly higher but normalized after percutaneous ASD closure. In contrast systolic strain rate and strain are not different from normal and no significant changes could be found after ASD closure. These findings suggest that the deformation parameters peak systolic strain rate as well as systolic strain are relatively load independent indices of ventricular function within the clinical range.

P108
Myocardial performance index (MPI) in right ventricular function assessment in children with hypoplastic left heart syndrome (HLHS) prior and after Norwood procedure

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Background: Right ventricle (RV) is a functionally single ventricle in children with HLHS. The assessment of RV function is difficult because of its complicated geometry. Myocardial performance index (MPI) allows for quantitative assessment of RV regardless of its shape because it is independent of geometric assumptions. MPI is calculated as a ratio of isovolumic time intervals to ventricular ejection time derived from the Doppler curves.
Aims: The aim of this study was to estimate usefulness of RV-MPI for RV function assessment in children with HLHS prior and after Norwood procedure.
Material and Methods: RV-MPI was calculated from Doppler tracings of RV inflow and outflow. Echocardiograms of 110 patients with HLHS (78–70.9% boys and 32–29.1% girls) were reviewed preoperatively (mean age of 5.3 ± 3.97 days). Twenty five age and weigh-matched newborns served as controls. We compared preoperative RV-MPI in 54 children who survived Norwood procedure with RV-MPI of 56 children who died during first 30 days after operation. In survivors group RV-MPI was also calculated postoperatively and before the second stage of surgical treatment (BCPA) (mean age of 5.3 ± 0.98 months).

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Results: There was significant difference ($p < 0.001$) between RV-MPI for children with HLHS comparing with controls (0.519 ± 0.17 vs. 0.3 ± 0.075). There was no significant difference in preoperative RV-MPI in patients who survived Norwood procedure comparing with no-survivors (0.519 ± 0.158 vs. 0.519 ± 0.182). There was no significant difference in RV-MPI prior and after Norwood procedure (0.519 ± 0.15 vs. 0.501 ± 0.154). RV-MPI calculated before the second stage of surgical treatment was significantly ($p < 0.001$) higher comparing with early postoperative RV-MPI (0.501 ± 0.154 vs. 0.63 ± 0.148).
Conclusions:

1. RV-MPI is significantly higher in patients with HLHS comparing with healthy neonates.
2. Preoperative RV-MPI seems not to be a risk factor of Norwood procedure.
3. Norwood procedure seems to have no impact on right ventricular performance.
4. RV-MPI in children with HLHS increases in time suggesting RV dysfunction and the early need for reduction of volume load during the second stage of surgical treatment (BCPA).
5. MPI is a useful and objective method of assessing and following RV function that is independent of ventricular geometry.

P109

Value of non-invasive tissue Doppler derived Tei-index in detection of coronary allograft vasculopathy in pediatric heart transplant recipients

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Introduction: Coronary allograft vasculopathy (CAV) remains a main factor limiting long-term survival after pediatric heart transplantation (HTX). The diagnosis of CAV is still based on serial coronary angiography and histopathological studies of biopsies sampled from the right ventricle. Since CAV is expected to be associated with altered global myocardial systolic and diastolic performance, evaluation of the global and regional myocardial contractile function by Tissue Doppler Imaging (TDI) may provide information on chronic coronary vascular disease. In this study, we evaluated the diagnostic value of the tissue derived parameters in detection of CAV in relation to invasive assessment of coronary angiography.
Method: Out of 132 pediatric heart transplants recipients in the German Heart Centre Berlin 25 pediatric patients with a median age of 13 years and median period of 5.98 years after heart transplantation, received selective coronary angiography to determine the degree of coronary artery disease (CAV), according to criteria of Gao et al. (1988). TDI was performed one day before cardiac catheterization and systolic and diastolic myocardial wall motions were analyzed in 4 chamber view. The left ventricular (LV) Tei-index (summation of the isovolumetric contraction and relaxation time divided by the ejection time) was calculated from the TDI-spectral waves and color M-Mode.
Results: Seven patients had CAV (28%). The development of CAV correlated significantly with the post transplant period ($p < 0.01$). The Tei-index among patients with CAV was significantly higher in comparison to patients without CAV ($p < 0.01$), reflecting altered global function. The significantly higher Tei-index among the TVP group was attributed to significant prolongation of the isovolumetric contraction time ($p < 0.001$), mainly indicating deterioration of the LV systolic function.
Conclusions: The non invasive tissue Doppler (TDI) derived Tei-index may identify pediatric heart transplant recipients with coronary allograft vasculopathy (CAV) and may help to develop successful early preventive and therapeutic strategies, that may influence the course of this disease.

P110

The influence of pulmonary regurgitation on ultrasonic strain rate and strain indices in patients after surgical repair of tetralogy of Fallot

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Aim: To assess the influence of pulmonary regurgitation (PR) after repair of tetralogy of Fallot (TOF) on right ventricular function using ultrasonic derived deformation indices: strain rate and strain.
Methods: 40 patients with various degrees of PR after repair of TOF were studied (age 11.1 ± 3.3 yrs). Patients with significant RVOT obstruction (PIG $\geq$ 25 mmHg), branch pulmonary artery stenosis or tricuspid regurgitation were excluded. Pulmonary regurgitant fraction (PRF) was quantified using MRI velocity mapping (PRF = pulmonary regurgitant flow/total forward pulmonary flow). Color Doppler myocardial imaging (CDMI) data were obtained at high frame rates ($\geq$150 frames/sec). Longitudinal systolic strain rate and strain were calculated for the basal, mid and apical segments of the right ventricular free wall. These deformation indices were compared between a patient group with severe (PRF $\geq$ 30%, n = 22) vs mild to moderate (PRF $\leq$ 30%, n = 18) PR. To assess the influence of the duration of PR, systolic strain rate/strain indices were compared between an older ($\geq$10 yrs, n = 9) and a younger ($\leq$10 yrs, n = 13) patient group with severe PR.
Results: In patients with severe PR (PRF 42 ± 7%) systolic deformation indices were significantly lower compared to patients with mild to moderate (PRF 19 ± 8%) PR (strain rate 1.29 ± 0.23 vs 1.59 ± 0.45/sec, $p = 0.02$; strain 17 ± 4.3 vs 24.6 ± 7%, $p \leq 0.001$). In patients with severe PR, systolic strain rate is significantly lower in the older compared to the younger age group (1.12 ± 0.14 vs 1.42 ± 0.2/sec, $p = 0.001$). Systolic strain tended to be lower in the older patients but the difference was not significant.
Conclusion: Systolic strain rate and strain of the right ventricle is inversely correlated with increasing PR. Longstanding PR leads to further deterioration of systolic strain rate. These quantitative indices of myocardial deformation are a promising tool to monitor right ventricular function during follow-up of postoperative TOF patients.

P111

Computer based training. A two dimensional echocardiography simulator

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Echocardiography requires visual-perceptive (interpretation of the echocardiographic images) and sensorimotor (steering of the ultrasound probe) skills, both of which cannot be taught sufficiently

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to beginners by lectures or textbooks. New learning media like high-end computer simulators might overcome this problem. We have designed a Java-based simulator that addresses these problems.

1. On a standard computer screen a side-by-side presentation of a three-dimensional (3D) virtual reality (VR) scene on the right and a two-dimensional echocardiographic (2DE) view on the left side is given. The VR scene consists of a 3D heart model and an animated ultrasound (US) probe with scan plane. The 2DE view is calculated from real 3D echocardiographic data sets that are registered with the heart model to achieve spatial and temporal congruence. The displayed 2DE view is defined and controlled by the orientation of the virtual scan plane. This side-by-side presentation supports the beginner to get a visual explanation of the spatial relationship between a certain probe position and corresponding 2DE view. Since the 2DE views are derived from a 3D echocardiographic data set in real time they are not restricted by pre-recorded views but the beginner can interactively explore the heart by manipulating the virtual US probe.
2. To train the hand-eye-coordination one has to simulate the US probe and link it to the virtual scene. Therefore we equipped a plastic dummy transducer with a 3D electromagnetic position sensor and placed it on a plastic torso. Through a calibration the spatial relationship between this dummy transducer and the dummy torso is the same as the relationship between the virtual US probe and the virtual heart on the computer screen. Any manipulations of the dummy transducer are transferred in real-time to the application showing the 2DE views that would be achieved if a "real" patient is scanned. Thus, all components of a 2DE examination are simulated. The advantage of the simulator compared to "just watching" an expert performing a 2DE examination is that the beginner does not need a verbal explanation of the relationship between US probe and resulting 2DE view but gets a visual presentation of these complex spatial relationships. He is able to experience and understand the results of his own probe manipulations.

P112

Congenital pulmonary and systemic venous anomalies: MRI and MRA evaluation

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Purpose: To evaluate the diagnostic value of MRI and Gadolinium (Gd)-enhanced three-dimensional (3D) magnetic resonance angiography (MRA) in patients with congenital anomalies of the pulmonary (PV) and systemic (SV) veins.
Material and Methods: The study retrospectively analyzed 51 patients (age 1 month to 70 years; average 14 years) with congenital anomalies of the pulmonary and systemic veins, who underwent complete MRI examination including Fast spin-echo, cine and velocity-encoded sequences as well as Gd-enhanced 3D MRA with multiple reconstruction techniques. The findings were correlated with echocardiography and cardiac catheterization available data.
Results: 64 congenital anomalies of the pulmonary and systemic veins were found in 51 patients. The SV anomalies (n = 48) consisted of the following: bilateral superior vena (n = 7), left superior vena cava (n = 22), retroaortic innominate vein (n = 6), interrupted inferior vena cava (IVC) with azygos continuation (n = 9), left sided IVC (n = 3), anomalous drainage of the IVC to the left atrium (n = 1). The PV anomalies (n = 16) consisted of the following: total anomalous pulmonary vein connection (n = 6), partial anomalous pulmonary vein connection (n = 9), and levoatrial cardinal vein (n = 1). Among the SV anomalies, only 16 out of 28 cases with data available were detected by echocardiography and 7 out of 19 cases with data available were detected by catheterization. All retroaortic innominate veins, associated in all cases with tetralogy of Fallot where missed by both methods. Among the PV anomalies, 13 out of 15 cases with data available were detected by echocardiography and 7 out of 7 cases with data available were detected by catheterization.
Conclusions: MRI is an accurate noninvasive tool for the diagnosis of congenital anomalies of the pulmonary and systemic veins. Gd-enhanced 3D MRA, with the help of multiple reconstructions is particularly useful.

P113

Identification of hypertrophic cardiomyopathy by magnetic resonance imaging in the absence of echocardiographic diagnosis

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Background: The two-dimensional echocardiogram (ECHO) has been the standard, noninvasive diagnostic test for the clinical diagnosis of hypertrophic cardiomyopathy (HCM) and also has an important role in risk stratification. A direct relation between magnitude of wall thickness and risk for sudden death has been demonstrated. We hypothesized that MRI would be more powerful than echo in establishing the diagnosis and measuring the extent of hypertrophy in HCM.
Methods: Forty-nine patients (age: 34 ± 16) suspected of (or known to have) HCM were imaged by both ECHO and MRI. With cine imaging 10–15, short axis slices (5–8 mm) were aquired to assess LV wall thickness. Standard LV cross-sectional views were obtained by Echo and compared to MRI. Maximum wall thickness was measured in 8 anatomic segments (anterior and posterior ventricular septum; anterolateral and posterior free wall) in both the distal and proximal LV; a total of 392 segments were assessed in 49 pts. Wall thickness measurements were made in a blinded fashion with ECHO and MRI.
Results: In 3 of 49 pts (6%) with a family history of HCM, Echo was judged normal without LV hypertrophy in any segment. However, MRI showed otherwise undetected hypertrophy in the anterolateral free wall (17 and 20 mm; twins age 14 and 19 mm; age 43) resulting in the phenotypic diagnosis of HCM for the first time and triggering implantation of cardioverter-defibrillator for primary prevention of sudden death in 2 of them. MRI more commonly showed the greatest LV thickness than did Echo (26 vs 16 pts; $p < 0.05$). Also, MRI identified LVH (undetected by echo) in 29 of 392 segments (7%) in 16 pts (33%). In 3 pts, echo was technically suboptimal and MRI confirmed the diagnosis of HCM (2 pts) or excluded it (1 pt).
Conclusions: MRI was advantageous over ECHO to: (1) definitively identify regions of LV hypertrophy not recognized by ECHO, and therefore was solely responsible for the identification of the phenotype in 6% of pts; (2) enhance assessment of the magnitude of LV hypertrophy for the purpose of risk stratification and provide a diagnostic alternative for pts with technically inadequate Echo.

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P114

Quantitative analysis of the the right atrial performance in patients after surgical repair of tetralogy of Fallot by means of Tissue Doppler Imaging (TDI) and Magnetic Resonance Tomography (MRT)

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Background: Atrial function is a crucial determinant of ventricular filling. Data on right atrial (RA) function in TOF patients is sparse and there is no data about the interaction between RA and right ventricular function among this group of patients.
Objectives: We aimed firstly to assess the right atrial (RA) performance in patients after corrective surgery of tetralogy of Fallot (TOF) and secondly to clarify the relation between the RA pump function and right ventricular (RV) systolic function using tissue Doppler Imaging (TDI) and magnetic resonance tomography (MRT).
Material and Methods: Fifty asymptomatic patients following corrective surgery of TOF and 30 age matched normal subjects were included in this study. RA maximal area (Amax), area at onset of atrial kick (Aa), minimal area (Amin) were measured by 2-D echocardiography and the RA active emptying area fraction [(Aa − Amin)/Aa × 100%] was applied to evaluate the atrial pump function. TDI derived right atrial strain rate systolic (Ssr), early diastolic (Esr) and late diastolic (Asr) peaks were measured in the middle of atrial lateral wall. Magnetic resonance tomography was used to assess the RV ejection fraction (RV EF).
Results: Compared to controls TOF patients had significantly reduced right atrial active emptying area fraction ($p = 0.005$) and right atrial Ssr ($p = 0.001$), Esr ($p = 0.038$), Asr ($p = 0.002$). TOF patients with RV ejection fraction less than 50% shows a significantly higher late diastolic RA strain rate peak when compared to patients with RV EF over 50%.
Conclusions: Compared to normal controls patients following corrective surgery of TOF have a reduced right atrial performance. Among this group of patients, however, the right atrial pump function may be relatively enhanced to compensate right ventricular dysfunction.

P115

The prevalance of interatrial septal aneurysms in newborns and their natural courses

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Background: Limited numbers of studies had been described about the atrial septal aneurysm (ASA) in children. Therefore, the incidence of ASA does not well known in children. The aim of this study: 1–to evaluate the incidence of ASA in newborn, 2–to define the natural course of this lesion, 3–to find the associated other cardiac defects.
Methods and Results: In the period from May 2002 to April 2003, 1076 neonates were examined to evaluate ASA with echocardiographic study between postnatal 24th and 72th hours. When the ratio of the interatrial aneurysmatic excursion length from the interatrial septum to the width of the involved atrium is 25% or greater it was accepted as ASA. The echocardiographic examination of the case with ASA was repeated with three month intervals beginning from the first month. 1072 newborns were included in this study. While septal aneurysmatic excursion into the atria was seen in 101 newborns, 81 of them (7.6%) had ASA. The prevalance of ASA was slightly higher among preterm newborns than terms (respectively 11.1% and 7.1%). All ASA were moving (56.8% into the left, 3.7% into the right and 39.5% into the both atriums) and all were spherical in shape. The mean excursion length was 5.3 ± 1.1 mm and mean protrusion ratio was 37.3 ± 9.2%. Although 59 (72.8%) of 81 newborns with ASA had interatrial shunt, 27.2% had no shunt. Follow-up examination was completed in 65 of 81 newborns and the lesion was disappeared in all cases. ASA was straightened in 69.2% at first month, 24.9% at third month and 6.2% at sixth month. There were still ASA in 2 cases on the ninth month. Twelve newborns with ASA had associated other cardiac lesions; 6 ASD, 4 VSD, 1 minor coronary artery anomaly and 1 ALCAPA syndrome.
Conclusions: The prevalance of ASA in newborns is higher than the older children and adults (7.6%) and higher in preterms than in terms. Our results indicate that ASA is a benign lesion in newborn period and it straightens mostly at first and then at third months. We found that excursion length, protrusion ratio, linear length of IAS, base length do not influence the disappearing time of ASA.

P116

Tricuspid annular velocities and time intervals in normal infants and children

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Noninvasive quantification of right ventricular (RV) function in children is very difficult due to complex shape, respiratory-variable filling, age and heart rate dependence.
Objectives: The objective of this study was to analyze tricuspid annular motion and age influence in a cohort of normal children using Doppler Tissue Imaging (DTI) technique.
Methods: Study group included 88 normal children. To better characterize age-related changes in tricuspid annular motion, the study population was divided into 2 groups: those aged less than 1 year (infant group, n = 18), and those aged more than 1 year (older group, n = 70). Pulsed DTI tracing of lateral tricuspid annular motion were recorded from an apical window using 4-chamber view. Tricuspid DTI measurements included: early peak diastolic annular velocity (Ea), peak diastolic annular velocity with atrial contraction (Aa), peak systolic annular velocity (Sa), isovolumic contraction (IVC) and isovolumic relaxation (IVR) time intervals. Two parameters were calculated: Ea/Aa ratio and Tei index as a parameter of global RV function (IVC + IVR/systolic time).
Results: Main results are presented in this Table:

Variable	Study group (n = 88)	Age < 1 year (n = 18)	Age > 1 year (n = 70)	p Value
Age (months)	97.4 ± 67.0	3.5 ± 2.9	123.5 ± 50.5	$p < 0.001$
Heart rate (beats/min)	99.3 ± 33.1	156.1 ± 11.1	86.3 ± 18.7	$p < 0.001$
Ea (cm/sec)	17.6 ± 3.6	16.7 ± 2.9	17.8 ± 3.8	NS
Aa (cm/sec)	11.3 ± 2.8	11.9 ± 2.3	11.2 ± 2.9	NS
Ea/Aa	1.7 ± 0.5	1.5 ± 0.5	1.7 ± 0.5	NS
Sa (cm/sec)	15.1 ± 2.0	13.2 ± 3.1	16.3 ± 2.0	$p < 0.05$
IVC (ms)	40 ± 30	30 ± 20	50 ± 30	$p < 0.005$
IVR (ms)	30 ± 10	30 ± 20	30 ± 10	NS
Tei index	27 ± 12	29 ± 9	27 ± 13	NS

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Conclusion: Systolic tricuspid annular velocity and isovolumic contraction time varied with age of children and appeared to distinguish RV function in infants when compared with older children. Diastolic annular velocities and RV myocardial performance index (Tei index) were not significantly age related. Doppler Tissue Imaging is a promising and useful technique for the assessment of RV function, because most parameters are not strongly age dependent.

P117

Tei-index as a parameter to detect acute rejection in pediatric transplant patients

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Introduction: Systolic and diastolic myocardial performance characteristics may be altered during allograft rejection in pediatric heart transplant patients. Noninvasive diagnostic tools for the detection of allograft rejection are limited. We examined the clinical utility of the Tei index (calculated as a ratio of isovolumetric contraction time and isovolumetric relaxation time to the ejection time), to detect acute graft rejection in pediatric heart transplant recipients.
Method: Among 21 out of 132 pediatric heart transplant patients at the Deutsches Herzzentrum Berlin serial measurements of Tei-index by standardised Doppler-echocardiography were performed. During this time acute biopsy-proven moderate rejection with a grading 3A according to ISHLT was diagnosed in six patients. In this patients Tei-index was assessed daily by echocardiography during treatment of rejection, and after recovery from rejection. Parallel a group of 15 patients without graft-rejection (ISHLT grade $<$1A) served as a control group and Tei-index was measured serially during the same time period.
Results: In all patients with rejection Tei-index was significantly increased compared to baseline-values (0.48 vs. 0.34, $p = 0.018$), and Tei-index decreased to nearly baseline values after treatment. In the control group there were no significant difference in Tei-index during the serial measurements.
Conclusions: These results indicate that serial measurement of Tei-index can point out early changes of diastolic and systolic performance related to cardiac allograft rejection and subsequent treatment.

P118

Right pulmonary artery left atrial tunnel presenting with brain abscess: a case report

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Brain abscess is one of the most serious complications of pulmonary arterio-venous connections. Most of these connections are located intrapulmonary. Pulmonary artery-left atrial connection is an unusual form of right to left shunting lesions. Herein, we report a girl with right pulmonary artery left atrial connection presenting with brain abcess.
Case Report: A 16-year-old cyanotic girl was referred to our neurosurgery department because of her brain abscess. By history she has been cyanotic since her early infancy but has not been evaluated as cardiac patient. Physical examination revealed marked central cyanosis and clubbing. Heart sounds were normal. A 12-lead ECG showed mild left atrial hypertrophy. A 2-D echocardiography and color Doppler studies were normal. In a contrast echocardiography early filling of the left atrium with contrast material was seen. A cardiac catheterization revealed normal right atrial, right ventricular, pulmonary artery, left atrial, left ventricular, and aortic pressures. Oxygen saturations were as follows: 49–51% in the right atrium, the right ventricle and the pulmonary artery, 77–79 in the left atrium, the left ventricle and the aorta, and 97% in the pulmonary vein. The Qp/Qs was calculated as 0.62/1. In a pulmonary artery angiogram, a large tunnel between right pulmonary artery and left atrium was observed. When a NIH catheter was pushed into the tunnel, it was easily passed to the left atrium and then to the left ventricle. We performed left ventricle, and left atrium angiograms, and also angiograms while the catheter was located in the tunnel.

After surgical drainage of the abscess and four weeks of intravenous antibiotic therapy, the patient underwent an operation for her cardiac anomaly.

The tunnel between the right pulmonary artery and the left atrium was closed with simple ligation and transfixion techniques. The patient became acyanotic with arterial oxygen saturations of 98% after surgery and remained asymptomatic during the six months of follow-up since the operation.

P119

Assessment of left and right ventricular functions in children with sickle cell anemia using the Doppler myocardial performance index

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Patients with sickle cell anemia have some cardiac abnormalities secondary to chronic volume overload. Previously, systolic and diastolic functions have been widely evaluated separately in this disease. However, the myocardial performance index (MPI), is a relatively new index which is defined as the ratio of the summation of isovolumic contraction and relaxation time to ejection time. Although, the MPI is useful for the assessment of global cardiac function in adults and children, its use in the pediatric patients with sickle cell anemia is limited. The purpose of this study was to evaluate left and right ventricular function by using the Doppler MPI in children with sickle cell anemia. We examined 32 patients (age: 10.4 $\pm$ 4.2, range 5–21 years) with sickle cell anemia and 30 (age: 9.9 $\pm$ 3.7, range 5–20 years) healthy children. The entire echocardiography and Doppler examination was completed for each subjects. MPI was calculated from the Doppler tracings. Compared with controls, left ventricle end-diastolic diameter (42.7 $\pm$ 5.2 mm vs 38.9 $\pm$ 5.1 mm, $p = 0.006$) and end-systolic diameter (24.8 $\pm$ 3.9 mm vs 22.2 $\pm$ 3.2 mm, $p = 0.006$), early diastolic mitral flow velocity (1.03 $\pm$ 0.15 m/s vs 0.94 $\pm$ 0.15 m/s, $p = 0.025$), late diastolic mitral flow velocity (0.66 $\pm$ 0.14 m/s vs 0.58 $\pm$ 0.10 m/s, $p = 0.014$), early diastolic tricuspid flow velocity (0.86 $\pm$ 0.28 m/s vs 0.75 $\pm$ 0.12 m/s, $p = 0.044$) and late diastolic tricuspid flow velocity (0.65 $\pm$ 0.15 m/s vs 0.53 $\pm$ 0.09, $p = 0.000$) were significantly increased in the patients with sickle cell anemia. On the other hand, the left ventricle ejection fraction and shortening fraction did not differ between the two groups. Both left ventricle MPI and right ventricle MPI were significantly higher in patients than those in normal children (0.319 $\pm$ 0.109 vs 0.175 $\pm$ 0.053, $p = 0.000$ and 0.223 $\pm$ 0.058 vs 0.171 $\pm$ 0.047, $p = 0.000$ respectively). Although, the left ventricle ejection fractions were in the normal ranges in patients and controls, MPI was higher in the patients with sickle cell anemia. These findings suggest that MPI is more sensitive parameter for assesing the ventricle functions.

We conclude that the Doppler index of myocardial performance may be a useful noninvasive and sensitive tool for assessing the left and right ventricle functions in patients with sickle cell anemia.

P120

Impaired longitudinal myocardial deformation in patients late after anthracycline treatment. A Doppler myocardial study

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Objectives: Cardiac toxicity remains as a major limitation for the use Anthracyclines (Ant) when treating malignancies. Currently no good method for early detection of cardiac dysfunction or a good predictive parameter is available. Strain (S) and Strain Rate (SR) imaging is a new technique that allows the quantification of regional myocardial function. We aimed to assess the effect of Ant on regional cardiac function using S and SR in patients who had previously received these chemotherapeutic agents.
Methods: We studied 50 patients, mean age: 13.10 ± 3.5 years, who had received Ant for different malignancies before the evaluation, mean: 3.5 years (range: 1.2–10.2 years). Conventional echocardiographic indices (grey scale M mode, 2D, blood pool Doppler) and Colour Doppler Myocardial Imaging were recorded. We measured and calculated radial Myocardial Velocities (Vel), S and SR in the mid infero-lateral wall using a short axis view. Longitudinal Vel, S and SR were calculated for the basal, mid and apical segments of the septum and lateral wall from apical 4-chamber view. We compared the obtained values with 33 age-matched normals using an unpaired t test.
Results: There was no significant difference between the patient group and the normals for the fractional shortening (36% ± 6 vs. 35.9% ± 3.9), the left ventricular end diastolic diameter (45.3 mm ± 4.5 vs. 43.0 mm ± 3.7), the A/E ratio (2.1 ± 0.7 vs. 2.3 ± 0.3) or the heart rate (76.8 ± 14.4 vs. 72.6 ± 12.3). There was also no difference in the radial function neither for peak systolic Vel (3.8 cm/s ± 1.0 vs. 3.7 cm/s ± 0.6), peak systolic SR ($3.7/s^{-1}$ ± 1.0 vs. $4.1/s^{-1}$ ± 1.1) nor peak systolic S (53.7% ± 15.2% vs. 59.3% ± 10.0). For longitudinal function there was a significant difference in the six analysed segments when measuring peak systolic longitudinal S, as well as in five of the segments when measuring peak systolic longitudinal SR. Peak Systolic Vel failed to show any change in any of these segments.

	Peak systolic velocity (cm/s)		Peak systolic strain rate (s^{-1})		Peak systolic strain (%)	
	Anthracyclines	Normals	Anthracyclines	Normals	Anthracyclines	Normals
Basal septal	4.7 ± 1.2	4.9 ± 0.9	−1.7 ± 0.4⋆	−2.0 ± 0.7	−18.9 ± 6.3#	−23.3 ± 5.4
Mid septal	3.7 ± 1.0	3.8 ± 0.9	−1.6 ± 0.7⋆	−2.0 ± 0.6	−18.9 ± 5.0#	−25.0 ± 6.5
Apico septal	2.6 ± 0.9	2.2 ± 1.0	−1.6 ± 0.4⋆	−1.9 ± 0.5	−20.2 ± 7.5#	−26.2 ± 5.6
Basal lateral	8.0 ± 2.1	7.5 ± 1.9	−1.9 ± 0.5⋆	−2.4 ± 1.1	−21.2 ± 9.2⋆	−27.7 ± 11.8
Mid lateral	7.3 ± 2.7	6.7 ± 1.8	−1.8 ± 0.9⋆	−2.3 ± 1.1	−20.5 ± 11.5#	−28.5 ± 9.3
Apico lateral	5.9 ± 2.0	5.1 ± 2.5	−2.0 ± 0.7	−2.2 ± 0.9	−21.8 ± 8.8#	−27.0 ± 7.5

Values are expressed as mean ± SD, ⋆$p < 0.05$ vs. normals, #$p < 0.01$ vs. normals.

Conclusions: By measuring deformation indices, it can be shown that longitudinal, but not radial, function appears to be affected in patients late after Ant treatment. This could be the reason why conventional echocardiography, which measures the radial fractional shortening or ejection fraction, fails to show dysfunction. The predictive value of this finding needs to be further determined.

P121

Coronary flow reserve after reimplantation of coronary arteries – intracoronary measurements

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The fate of the coronary arteries after reimplantation is important for the clinical course in some patients. We started to perform selective coronary angiograms and flow reserve measurements.
Patients and Methods: We examined 11 patients, 8 after arterial switch operation, Ross-procedure (1), Kawasaki-syndrome (1) and 1 patient with reimplantation of the left coronary artery in Bland-White-Garland-S. The age was between 5 and 16 years. Via the femoral artery selective right and left intubation of the coronary arteries was performed. We were more successful with mulipurpose catheters for the right and Amplatz for the left. After selective contrast injection a 0.014″ doppler-wire was introduced and adenosine was given. The baseline and peak flow was measured, the flow reserve calculated.
Results: We found excellent postoperative anatomic results, no stenosis. The coronary flow reserve was between 2,9 and 4,1.
Conclusions: The coronary flow reserve (CFR) was normal compared with results in literature. Hamaoka (Circulation 1995) reported data in children with angiographic normal coronary arteries, but after Kawasaki-syndrome. Oskarsson (Circulation 2002) examined 11 patients after ASO, and found normal CFR values between 3,0 and 4,8.

P122

Tissue doppler imaging in adolescents with anorexia nervosa

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Introduction: Tissue Doppler imaging (TDI), which can record motion velocity of ventricular wall, is a recently developed technique and is expected to provide important information about hemodynamic abnormalities in children with various heart diseases. Anorexia nervosa (AN) in adolescents can be associated with cardiac changes, such as thinning of the LV wall, reduction of the LV mass index (LVMI), pericardial effusion, mitral valve prolapse and ECG abnormalities.
Purpose: We used TDI to examine the hemodynamic abnormalities along the longitudinal, radial and transverse axes in the acute state of adolescents with anorexia nervosa (AN).
Patients and Methods: 17 adolescent patients with AN (14.32 ± 2.25 y) BMI (15.5 ± 2.28 kg/m^2) with self-induced starvation were referred. None of them had clinical symptoms of cardiovascular disease and all were studied in the acute state of the disease. In the study we included controls (13.95 ± 2.88 y) BMI (19.7 ± 2.41 kg/m^2). None of the controls had a family history of cardiac diseases or cardiotoxic medication. Patients and controls underwent a 2-D-echocardiography and tissue Doppler imaging (Vivid 7).

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Results:

	Strain rate (1/s)			Strain (%)		
	Anorectics	Controls	p value	Anorectics	Controls	p value
SAX	4.081 ± 0.992	3.755 ± 0.970	0.230	0.647 ± 0.137	0.572 ± 0.118	0.031
LAX	3.962 ± 0.796	3.755 ± 0.970	0.437	0.650 ± 0.164	0.572 ± 0.118	0.038
Septal	−2.893 ± 1.656	−1.833 ± 0.584	<0.001	−0.281 ± 0.131	−0.245 ± 0.0062	0.034
Lateral	−2.969 ± 1.941	−2.156 ± 0.982	0.004	−0.304 ± 0.186	−0.258 ± 0.092	0.080
RV	−3.762 ± 1.387	−2.642 ± 0.999	<0.001	−0.478 ± 0.183	−0.391 ± 0.146	0.004

Conclusions: We can conclude that strain rate values are significantly higher ($p < 0.01$) in the anorectic group in the longitudinal direction in the septal, left lateral and right ventricle.This is probably the result of the thinning of the myocardial wall.

P123

Regional myocardial functions in rheumatic mitral regurgitation by Doppler tissue imaging

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To investigate the regional myocardial functions of children with rheumatic mitral regurgitation (MR) by Doppler tissue imaging (DTI), left ventricular (LV) regional functions were analyzed with early diastolic (peak E), late diastolic (peak A) and systolic (peak S) myocardial velocities in the longitudinal axis from apical view (lateral and medial mitral annular, basal and middle segments of interventricular septum (IVS) and posterior wall of LV (LVPW)) with pulsed DTI and with conventional echocardiographic examinations in 19 children with rheumatic MR (Group 1: age: 12.5 ± 2.0 years) and 21 healthy children (Group 2: age: 13.1 ± 3.1 years). All children underwent detailed conventional echocardiographic study. M-mode, 2-D measurements, and pulsed wave inflow velocities of LV were recorded in all.
Results: Compared with Group 2, patients in Group 1 showed; (1) higher LV end-diastolic diameter on M-Mode echocardiographic examination (47.7 ± 6.6 mm in Group 1 and 43.7 ± 3.3 mm in Group 2, $p = 0.019$), (2) longer mitral acceleration time on conventional pulsed wave echocardiographic examination (89.2 ± 13.8 msec in Group 1 and 80.5 ± 12.9 msec in Group 2, $p = 0.047$), (3) higher peak A and S velocities of basal and middle segments of IVS and LVPW on DTI examination (Table), (4) lower E/A ratio of both conventional LV inflow velocity measurements (1.6 ± 0.4 in Group 1 and 1.8 ± 0.3 in Group 2, $p = 0.037$) and DTI measurements from lateral mitral annular site (2.0 ± 0.6 in Group 1 and 2.6 ± 0.9 in Group 2, $p = 0.022$). We also found that E/A ratio of LV inflow velocities was positively correlated with E/A ratio of DTI in healthy controls ($r = 0.575$, $p = 0.008$). However, this positive correlation was lost in children with rheumatic MR ($r = 0.213$, $p = 0.382$).

Myocardial velocities of IVS and LVPW with DTI in children with rheumatic MR and in controls.

Myocardial velocities (cm/sec)	Group 1 (n = 19)	Group 2 (n = 21)	p
Peak A of basal IVS	10.5 ± 4.4	8.2 ± 1.9	0.036
Peak S of basal IVS	11.2 ± 2.1	9.7 ± 1.9	0.022
Peak A of middle IVS	8.4 ± 3.4	6.8 ± 1.9	0.070
Peak S of middle IVS	9.7 ± 2.4	10.5 ± 4.4	0.013
Peak A of basal LVPW	10.9 ± 2.3	8.7 ± 2.3	0.010
Peak S of basal LVPW	12.6 ± 3.4	10.1 ± 2.6	0.024
Peak A of middle LVPW	9.2 ± 2.1	7.4 ± 2.4	0.025
Peak S of middle LVPW	11.4 ± 3.3	8.9 ± 2.2	0.021

DTI: Doppler tissue imaging, MR: mitral regurgitation, IVS: interventricular septum, LVPW: posterior wall of left ventricle.

Conclusions: We conclude that increased left ventricular volume due to MR results increased left ventricular myocardial velocities of both IVS and LVPW during systole (peak S) and late diastole (peak A). In these patients the ratio of early diastolic (peak E) and late diastolic (peak A) mitral velocities are increased on both conventional and DTI echocardiographic examinations. The loss of correlation between E/A ratio of LV inflow velocities and E/A ratio of DTI in patients with rheumatic MR suggests that myocardial velocities relatively independent from compensatory alterations in LV filling due to increased preload.

P124

The prevalance of interatrial septal openings in newborns and their natural courses

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Even if interatrial shunts in the newborn are frequently encountered, little is known about their natural course and knowns are various. To establish the prevalance of interatrial septal openings (ISO) diagnosed in the first days of life and their spontaneous closure frequency, 1072 neonates born in the same center were examined by echocardiography and the newborns with an interatrial shunt confirmed by color Doppler echocardiography were invated to follow-up examinations for one year.
Methods: In the cases whose interatrial shunt was confirmed with color Doppler echocardiography, the diameter of IASO and the length of IAS were measured with two-dimensional echocardiography in the subcostal view. The septal ratio of IASO diameter was calculated. Kaplan-Meier curve was obtained to predict age of spontaneous closure.
Results: The prevalance of IASO was 78.6% in newborns. Sex and gestational age did not have any effect on the prevalance. The follow-up examinations were completed in 464 cases. An overall rate of spontaneous closure was 96.1%. Frequency and timing of closure were inversly correlated to IASO diameter: closure occured in 99.3% (303 of 305) of IASO in group 1 (diameter <3 mm), 94.8% of IASO (127 of 134) in group 2 (diameter 3 to 5 mm), 68.4% of IASO (13 of 19) in group 3 (diameter 5 to 7 mm) and 50.0% of IASO (3 of 6) in group 4 (diameter ⩾7 mm). Spontaneous closure did not occur in each 2 cases with IASO diameter ⩾8 mm in group 4 during the 12 months follow-up period. In addition, the time of IASO closure was positively correlated with the septal ratio of IASO diameter ($r = 0.118$, $p = 0.014$).
Conclusions: These results suggest that the rate of the spontaneous closure is very high for IASO with the diameter of <5 mm. The frequency of spontaneous closure is decreased in the cases with IASO diameter ⩾7 mm. The follow-up examinations should be in hort intervals in such cases. There was a positive correlations between the initial size and spontaneous closure time. With this report we also want to emphasize that the septal ratio of IASO diameter is an important parameter affecting the time of spontaneous closure.

P125

Visualization of peripheral pulmonary artery stenosis using high resolution multidetector computertomography

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Background: Exact knowledge of the pulmonary vasculature is the basis for adequate treatment in many cases of complex congenital heart disease. So far the gold standard for precise description of the pulmonary vasculature is cardiac catheterization. A case report of a child with peripheral pulmonary artery stenosis with pulmonary atresia and ventricular septal defect (Fallot type) and with multiple collateral arteries to both lungs demonstrates the facilities of high resolution multidetector computertomography (MDCT).
Methods: In a ventilator dependent child (3 years 9 months) the chest x-ray showed hyperperfusion of the right upper and middle pulmonary lobes and hypoperfusion of the right lower pulmonary lobe. After injection of 17 ml of contrast agent (Imeron 500®) within 10 seconds nearly isotropic voxels (0,5 × 0,5 × 0,6 mm) were obtained of the lower neck, thorax and upper abdomen using MDCT (Somatom Sensation®, 16 detectors). The pulmonary vasculature was visualized as multiplanar reformatted images and after segmentation as a surface rendered model. A stenosis in the right lower lobe branch of the pulmonary artery was detected. It was balloon dilated successfully in the catheterization laboratory. After intervention perfusion of the right lower pulmonary lobe increased and hyperperfusion of the right upper and middle pulmonary lobes decreased significantly. The child was weaned of the ventilator.
Conclusion: High resolution three-dimensional visualization of the pulmonary vasculature in children is possible with MDCT. These three dimensional data sets may be very helpful for diagnosis and planning of interventional procedures in the catheterization laboratory.

P126

Comparison of the tissue Doppler derived left ventricular Tei Index to that obtained by pulse Doppler in patients with congenital heart disease

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Background: The PW Doppler derived Tei-index is an useful parameter to assess the global left ventricular function without the need of any geometrical assumption. Measuring this index as described by Tei et al., however, it has two limitations. One is that the time interval between the end and onset of mitral inflow and ejection time are measured sequentially, i.e. not in the same cardiac cycle. The second limitation is one can not determine whether the altered global function is mainly due to systolic, diastolic or combined dysfunction. Tissue Doppler imaging (TDI) is a promising method to quantify regional wall motion. A recent study has demonstrated that the major cardiac events, i.e. opening and closure of the mitral and aortic valve, can be accurately determined from TDI stored squences.
Objectives: To compare the left ventricular Tei-index measured by TDI to that obtained by pulsed Doppler (PW) in patients with congenital and aquired heart disease.
Methods: In fourty consecutive patients with different congenital heart disease the LV PW Doppler derived Tei-index was assessed on-line as previously described. TDI derived M-mode and the unprocessed velocity trace from the basal septum were used to identify the opening and closure of the mitral and aortic valve in one cardiac cycle respectively. The TDI Tei-index was calculated off-line according to the same equation applied for PW Doppler derived Tei index (isovolumetric relaxation time + isovolumetric contraction time)/ejection time.
Results: A highly significant correlation was observed between the Tei index values determined by PW Doppler and TDI ($r = 0.92$, $p < 0.001$). The mean difference between the TDI-Tei- index and the pulsed Doppler Tei-index was 0.002 ± 0.037. Interobserver variability for the measurement TDI derived Tei index was $5 \pm 3\%$.
Conclusions: TDI Tei-index can be used to assess the global left ventricular function in patients with congenital heart disease. In contrast to PW Doppler derived Tei index, TDI derived Tei index obtained from the same cardiac cycle may help to differniate systolic from diastolic dysfunction via providing specific information on the isovolumetric intervals.

P127

Tissue Doppler derived strain rate for the assessment of atrial and ventricular function in patients with tricuspid atresia following Fontan operation

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Background: Assessment of systolic and diastolic atrial and ventricular function in children and adult with morphologically and functionally univentricular heart is difficult using the conventional echocardiography method. Tissue Doppler derived (TDI) strain rate (SR) can effectively quantify local myocardial deformation independent of the overall heart motion.
Objectives: The aim of this study was first to evaluate the regional myocardial function in patients with tricuspid atresia after palliative extracardiac Fontan operation and secondly to assess the atrial pump function among this group of patients.
Design and Patients: We studied 14 patients with a median age of 10.1 years (range 4 to 29 years) with primary diagnosis of tricuspid valve atresie after a median period of 4.3 (range 2 to 12) years after palliative extracardiac Fontan operation and compared to normal collective of children (20). In a standard apical longitudinal view myocardial velocity at the basal segment of the anterior right and posterior left ventricular wall was examined. From the myocardial velocity curve the systolic (ST), early diastolic (ET) and atrial contraction myocardial motion (AT) were measured. TDI derived atrial strain rate (SR) systolic (SSR), early diastolic (ESR) and late diastolic (ASR) peaks were measured in the middle of free atrial lateral wall. Atrial maximal area (Amax), area at onset of atrial kick (Aa), minimal area (Amin) were measured by 2-D echocardiography and the atrial active emptying area fraction [AEAF = (Aa − Amin)/Aa × 100%] was applied to evaluate the atrial pump function.
Results: In comparison to controls, postoperative Fontan patients had a significantly reduced annular anterior right ST ($p < 0.01$), and ET ($p < 0.01$) wall motion. In contrast the posterior left myocardial wall velocities (ST, ET, AT) at the posterior left wall did not show significantly differences when compared to controls. The atrial active emptying area fraction ($p < 0.05$) and ASR ($p < 0.05$) were significantly reduced in patients with tricuspid valve atresia in comparison healthy persons.
Conclusions: Abnormal myocardial ventricular and atrial wall motions are detectable in children and adults with univentricular heart after palliative cardiac procedures using TDI. The value of these parameters needs, however further validation, using other comparative methods.

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P128

Echocardiographic differentiation of atrioventricular septal defects from inlet ventricular septal defect and mitral clefts, the difficult made easy

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Although inlet ventricular septal defect (VSD) and isolated mitral valve clefts (IMC) are anatomically distinct from atrioventricular septal defect (AVSD), echocardiographic differentiation is often difficult. We established two echocardiographic measurements that were previously validated morphologically: the left ventricle inlet/outlet ratio (I/O ratio) and the percentage of the left atrioventricular valve guarded by the posterior (mural) leaflet (PLPL). We performed these measurements in 148 patients. The patients were divided into 3 groups; group 1: Normal (n = 100), group 2: AVSD (n = 34) and group 3: isolated inlet VSD with or without MVC (n = 14).
Results: For group l the I/O ratio was 1.03 ± 0.072. For group 2 the I/O ratio was significantly lower than normal (0.82 ± 0.062), p = 0.000 (CI: 0.180–0.237). For group 3 the I/O ratio was low compared to normal, 0.958 ± 0.07, p = 0.000 (CI: 0.034–0.116) but still higher than the AVSD group, p = 0.000 (CI 0.175–0.091). The PLPL for group 1 was 53.22 ± 2.38. For group 2 it was 48.30 ± 2.711, p = 0.000 (CI: 3.42–6.40). This percentage for group 3 was normal (53.92 ± 1.96).
Conclusion: This is the first study to set the normal echocardiographic values for left ventricle inlet/outlet (I/O) ratio and the percentage of the left atrioventricular valve guarded by the posterior leaflet (PLPL). These measurements can reliably differentiate the two pathologies. Although the I/O ratio in patients with inlet VSD with or without MVC can be abnormally low it is still not reaching the AVSD range.

P129

The use of contrast-enhanced MR angiography for planning interventional catheterization

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Background: Interventional catheterization is being increasingly used for relief of residual lesions in congenital heart disease. Exact anatomical imaging is crucial in the planning of an intervention. This can be provided non-invasively and without radiation by contrast-enhanced MR angiography (CEMRA).
Aim: To evaluate the accuracy of the measurements of the vessels obtained by CEMRA in comparison to those obtained by conventional X-ray angiography (CXA).
Methods: Retrospective blinded measurement of the diameters of aorta and pulmonary arteries on the CEMRA and CXA images, in the same locations. Comparison of the results by Pearson correlation and by calculating the limits of agreement.
Results: Twenty-one children with congenital heart disease, mean age 5.6 ± 5.2 years, weight 21.1 ± 18.4 kg, underwent CEMRA and catheterization for assessment or treatment of a residual lesion. The time interval between the CEMRA and the CXA examination was 2.6 ± 2.3 months. A total of 98 measurements, 37 of the aorta and 61 of the pulmonary arteries were performed on the images obtained by each technique. The correlation between CEMRA and CXA measurements was excellent, r = 0.97, p < 0.0001. The mean difference between the two techniques was 0.018 ± 1.1 mm; the limits of agreement were −2.14 and +2.18 mm. Similar agreement was found for measures of the aorta (r = 0.97, mean difference 0.20 ± 1.08 mm) and of the pulmonary arteries (r = 0.97, mean difference 0.048 ± 0.89 mm).
Conclusions: CEMRA provide accurate quantitative anatomical information, which highly agrees with CXA data, and can therefore be used for planning interventional catheterization.

P130

Blood flow quantification in the great arteries by phase-contrast magnetic resonance imaging combined with sensitivity encoding (SENSE)

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Purpose: To evaluate the feasibility of rapid free-breathing phase-contrast MRI (PC-MRI) at different in-plane resolutions combined with sensitivity encoding (SENSE) for flow quantification in the great arteries in healthy adult volunteers.
Methods: In 13 volunteers (mean age 33.0 ± 7.4 years), blood flow rate in the pulmonary artery (Qp), ascending aorta (Qs) and flow ratio Qp/Qs were determined by PC-MRI with SENSE reduction-factor 2 and 3 (SF-2, SF-3). Additionally, we used PC-MRI with higher spatial in-plane resolution (1.6 × 2.1 mm vs. 2.3 × 3.1 mm) with/without SF-3. Results were compared with standard (= reference) PC-MRI.
Results: Reduction of signal averages and application of SENSE accelerated flow measurements by a factor of 3.8 (5.5) using PC-MRI with SF-2 (SF-3), compared with standard PC-MRI: Scan time was 36 seconds (SF-2) and 25 seconds (SF-3) at average heart rate of 69/min., respectively. Mean Qp/Qs by reference PC-MRI was 1.03 ± 0.07 (range, 0.89–1.16), and 1.08 ± 0.11 (range, 0.86–1.24) by PC-MRI + SF-3. For blood flow rate through the pulmonary artery and aorta, and for Qp/Qs ratio, we found differences of −3.3% to +4.4%, lower limits of agreement (mean − 2SD) between −14% and −21%, and upper limits of agreement (mean + 2SD) between +8% and +29%, demonstrating good agreement with standard PC-MRI (Bland-Altman analysis). PC-MRI at higher in-plane resolution both with/without SENSE slightly underestimated aortic and pulmonary flows (mean difference 5.9–6.8%, p < 0.05).
Conclusions: In healthy adults, flow quantification in the great arteries by PC-MRI can be accelerated by a factor of >3.8 when using SENSE. Cardiac shunting is unlikely to occur at Qp/Qs ratio >0.85 and <1,25. High in-plane resolution was not advantageous.

P131

Quantitative analysis of paradoxical interventricular septal motion following corrective surgery of tetralogy of Fallot

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Objectives: To quantify paradoxical interventricular septal motion (PSM) among asymptomatic TOF patients following corrective surgery of tetralogy of Fallot (TOF).

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Design and Patients: Twenty asymptomatic TOF patients without severe pulmonary regurgitation and 20 age matched normal subjects were examined. The PSM was quantified using the 2-D echocardiography derived paradox index. Tissue Doppler derived strain rate was used to assess the longitudinal and radial systolic function of the interventricular septum (IVS). The Tissue Doppler imaging derived Tei index was used to assess the global LV ventricular function.
Results: Compared to the control group the paradox index was significantly higher ($p = 0.001$), while the regional IVS longitudinal ($p = 0.03$) and radial ($p = 0.01$) systolic strain rate peaks were significantly reduced in TOF patients. Among the TOF patients the paradox index correlated inversely with the IVS radial peak systolic strain rate ($r = -0.64$, $p = 0.004$) and positively with QRS duration ($r = 0.50$, $p = 0.02$). The LV Tei-index correlated significantly with the paradox index ($r = 0.71$, $p = 0.001$), QRS duration ($r = 0.79$, $p = 0.001$) and with the septal radial systolic strain rate peak ($r = 0.59$, $p = 0.004$).
Conclusions: Paradoxical septal motion in asymptomatic postoperative TOF patients without significant right ventricular volume overload is related to electrical delay and reduced septal systolic function. The reduced LV systolic function among this group of patients is mainly secondary to diminished regional septal systolic function and the paradoxical septal motion.

P132

Cardiac magnetic resonance and echocardiography for the evaluation of congenital aortic stenosis. A correlation study

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Objective: In the last 20 years Echocardiography (Echo) has become the most standard technique to evaluate patients with Aortic Stenosis (AS). Echo has some limitations, first, a good image quality is not always feasible; secondly the peak velocity may be underestimated due to the angle dependency of the Doppler technique; third, the valve area measured by planimetry mostly requires a transesophageal approach. The use of Cardiac Magnetic Resonance (CMR) is gaining more and more acceptance, given it can thoroughly, accurately and reproducibly assess different cardiac disorders. We aimed to compare the correlation between the Echo and CMR measurements in a group of patients with AS.
Methods: We studied 16 patients with moderate to severe congenital AS, mean age: 12.8 ± 2.8 years. The presence of more than mild aortic regurgitation was considered exclusion criteria. The Echo evaluation included grey scale M-mode, 2D and blood pool continuous and pulsed Doppler imaging employing a GE Vingmed System V, Horten, Norway, equipped with a 3.5 MHz probe. The CMR studies were performed using a 1.5 T Philips Intera CV unit. Cine CMR images, with ECG gating, were acquired using balanced fast field echo sequences, true fisp echo cine images and flow velocity mapping images. We measured, with both methods, the left ventricular mass, stoke volume, end diastolic and systolic volumes, peak gradient. The aortic valve area was measured using the continuity equation with Echo and by planimetry with CMR. A linear regression analysis was performed to describe the correlation between the two techniques.
Results: Results are shown in Table. Regardless of the higher values measured with Echo; there was an excellent correlation between the two techniques for the left ventricular mass. Also strong correlations were noted for the stroke volume, the end diastolic and systolic volumes and the peak gradient. The measurement of the aortic valve area yielded a weak agreement between the two methods.

	Echocardiography	Cardiac magnetic resonance	Correlation (r)
LV mass (gr)	223.3 ± 84.3	123.2 ± 54.3	0.96
Stroke volume (ml)	76,7 ± 31.0	84.2 ± 33.7	0.75
End diastolic volume (ml)	95.4 ± 29.0	108.5 ± 41.1	0.86
End systolic volume (ml)	30.0 ± 7.6	24.3 ± 11.6	0.83
Peak aortic gradient (mmHg)	56.3 ± 17.4	91.5 ± 57.0	0.79
Aortic valve area (cm^2)	1.0 ± 0.4	1.6 ± 0.4	0.44

Conclusions: In an AS population, the different parameters measured by CMR correlated well with the ones calculated by Echo; nevertheless some differences were perceived for the aortic valve area calculation. These data suggest that CMR could be an alternative to Echo for the evaluation of patients with AS.

Session 10: GUCH (CHD in Adults)

P133

Recovery kinetics of oxygen uptake is prolonged in adults with Mustard/Senning repair for transposition of the great arteries

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Background: Adults with a previous Mustard/Senning (M/S) repair for transposition of the great arteries have a reduced exercise ability and a reduced peak oxygen uptake (VO_2), but the ability to recover from exercise has never been investigated in this population.
Methods: 40 consecutive patients who underwent M/S repair in infancy (mean age at surgery 13 ± 4 months) followed-up at our outpatient clinic underwent a maximal cardiopulmonary exercise test with breath-by-breath measurement of expired gases at an age of 19.5 ± 11.3 years. For comparison, an age, sex and weight matched group consisting of 168 healthy individuals was built. The constant decay of VO_2, CO_2 production (VCO_2), minute ventilation (VE), and heart rate (HR), expressed as the first degree slope of a single linear relation were calculated for the first minute of recovery.
Results: When compared to the control group, M/S patients had reduced vital capacity ($p < 0.0001$), forced expired volume in the 1st second ($p < 0.0001$), peak VO_2 ($p < 0.0001$), peak O_2 pulse ($p < 0.0001$), and a higher VE/VCO_2-slope during exercise ($p < 0.0001$). Peak heart rate was not significantly different between the two groups ($p = 0.078$). M/S patients also showed, during recovery, a prolonged VO_2-slope ($p < 0.0001$), VCO_2-slope ($p < 0.0001$) and VE-slope ($p < 0.0001$) when compared to control subjects, whereas heart rate slope was not different ($p = 0.27$).

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Conclusions: A limited cardiopulmonary reserve in patients underwent M/S repair for transposition of the great arteries appears to affect not only maximal exercise responses but also the recovery phase.

P134

Tissue Doppler and strain echocardiography reflects systolic dysfunction in patients with systemic right ventricle

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Background: In certain congenital heart defects, the morphologic right ventricle (RV) supports the systemic circulation. RV failure is common and develops gradually in these patients. Accurate evaluation of RV function and early detection of RV systolic failure would greatly assist clinical management. Tissue Doppler and strain echocardiography (TDSE) are two novel echocardiographic techniques that are able to quantify ventricular function. We tested whether TDSE is feasible and reflects RV reduced systolic function in systemic RV.
Methods: Conventional echocardiography and TDSE were performed on consecutive patients (n = 10) with systemic RV due to congenitally corrected transposition of great arteries (ccTGA), reporting to the adult congenital heart disease clinic and on healthy controls (n = 12). Standard echocardiographic cross sections were used for conventional echocardiography and a right index of myocardial performance (RIMP), also known as Tei-index, was calculated to express ventricular function. For TDSE, narrow sector and high frame rate imaging of the RV free wall, in both patients and controls, was performed in the apical 4-chamber view. Longitudinal tissue velocities, strain rate and strain of the RV free wall were calculated. Statistical analysis was performed and expressed as median and range.
Results: Median age of our patient group was 42 years (26, 64); median age of our control group was 63.5 years (36, 72). Median RIMP was higher in patients than in controls (0.72; 0.54, 1.36 vs. 0.40; 0.19, 0.86; p = 0.001). Median visually estimated ejection fraction (EF) was reduced in patients with a systemic RV (40%; 25, 50). RV tissue displacement (TD), peak systolic strain rate (PSSR) and peak systolic strain (PSS) were significantly lower in patients with systemic RV compared to controls (Table).

Results of the analysis of the systolic values (i.e. tissue displacement (TD), peak systolic strain rate (PSSR) and peak systolic strain (PSS)).

	Tissue displacement (mm)	Peak systolic strain rate (s^{-1})	Peak systolic strain (%)
ccTGA (n = 10)	5.7 (3.3, 7.9)	−1.11 (−0.72, −1.66)	−13.1 (−5.4, −21.5)
Controls (n = 12)	12.7 (8.6, 27)	−1.7 (−0.55, −3.2)	−23.9 (−13.5, −32.7)
p-value	≤0.0001	0.01	0.0004

Conclusion: TDSE is feasible in patients with systemic RV and is able to accurately reflect reduced systolic function in a systemic RV. TDSE may be a useful quantitative technique to follow RV function in patients with systemic RV. RIMP gives an accurate indication of ventricular performance, however not on systolic function alone.

P135

Heart rate turbulence – a new prognostic parameter for patients with congenital cardiac disease? – first results of a prospective study

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Background: Heart failure and sudden death are the most common causes of death in adults with congenital cardiac disease (CCD). Therefore the identification of patients at risk for sudden death is of major interest. Although heart rate turbulence (HRT) is a strong predictor for mortality in chronic heart failure and after myocardial infarction, there are no data available regarding the predictive value of HRT for patients with CCD. The objective of the current study was to assess the prognostic value of HRT for risk stratification in adults with CCD.
Patients and Methods: 43 patients (median age 29 years; 27 male) with operated and unoperated CCD were included. HRT-parameters, Turbulence Onset (TO) and Turbulence Slope (TS), were calculated from a Holter-ECG. Clinical functional class (FC), serum levels of BNP and the occurrence of sudden death in the follow up period were documented.
Results: 33 patients had normal TO- and TS-values or no utilizable ventricular ectopic beats during holter monitoring. HRT was abnormal in 10 adults. Four of them had pathological values for TO and TS. TO was abnormal in another four patients, and two additional patients had abnormal TS. All four patients with abnormal TO and TS died within the clinical follow-up period. All of them were in FC IV and had highly elevated levels of BNP (>1300 mg/dl).
Conclusions: HRT, most probably an autonomous baro-reflex, is the physiological response of the sinus node to premature ventricular contractions. If the autonomic control system is impaired, this reaction is either weakened or entirely missing. The present study verifies for the first time that HRT – alone or in combination with other parameters (FC; BNP) – may be a suitable predictor for prognosis and mortality, not only in chronic heart failure and after myocardial infarction, but also in pts with operated or unoperated with congenital cardiac disease.

P136

Progression of congenital aortic stenosis at adult age

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Purpose: Very little information is available about the progression rate of congenital aortic stenosis in adult patients. This information is mandatory to define the optimal freqency of follow-up.
Methods: Retrospectively we studied all patients with congenital valvular aortic stenosis with serial echocardiography. Patients who

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only had 1 echo-study were excluded from the study. The study period was from the first echo-study until last follow-up or valve replacement.
Results: Eighty-two patients (56 men) were studied. The mean age at the beginning of the study-period was 17.9 (range 3–40) and at the end of the study 29.6 (range 17–55) years. Thirty-seven of the patients had 1 or more interventions: 15 surgical valvotomy, 17 balloon angioplasty and 22 valve replacement (9 artificial valve, 8 Ross-procedure and 5 homograft). The mean follow-up period was 11.7 year and a mean of 8 (range 2–17) echo-studies was performed per patient. The mean maximal velocity over the aortic valve at the start of the study was 3.2 (range 1.2–5.3) m/s and the mean progression per year was 0.05 (range 0–1.5) m/s. In the patients with at least 1 intervention the mean progression was 0.10 m/s per year. In the patients without intervention the mean maximal velocity over the aortic valve at last follow-up was 3.6 (range 2.0–5.2) m/s and in the patients with at least 1 intervention 4.6 (range 2.5–6.7) m/s. No patient showed a progression of more than 1.5 m/s/year. Left ventricular posterior wall thickness progressed from 9.9 mm at the start of the study to11.5 mm at the end of the study and in patients with intervention it progressed from 9.8 mm to 12.7 mm. The ascending aorta diameter progressed from 28.9 mm at the beginning of the study to 39.7 mm.
Conclusion: The rate of progression of congenital aortic stenosis in adult patients is very low and the progression seems lower than in adults with degenerative aortic valve disease. In most patients a control visit once in 3 years seems sufficient. Although there was very little progression in the gradient over the aortic valve, the increase of left ventricular wall thickness was striking. Furthermore a remarkable progression in ascending aorta dimension was found.

P137
How safe is pregnancy following Mustard procedure?
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Background: Mustard procedure was performed between 1966 and the early 1990s for infants with Transposition of the Great Arteries. Many of these patients are now of child bearing age.
Objective: To assess maternal outcome of pregnancy following Mustard procedure.
Method: Case note review of the 18 women followed up at our institution after Mustard procedure.
Results: Seven women had a total of eight pregnancies that they intended to carry to term. The mean age at conception was 24.5 years (range 19–31 years). Five women (six pregnancies) were in NYHA class 1 and had good ventricular function before conception. All six babies were born without neonatal complication. No change in patient's exercise tolerance was reported in the year following each delivery, however two had deterioration in cardiac function on echo. Subsequently one went on to require cardiac transplant 11 years following delivery (31 years following the Mustard procedure). Two women were in NYHA 2 at conception. One was 31 years of age and had poor systemic ventricular function. She was admitted 18 weeks pregnant requiring intra-venous inotropes. She died 25 weeks pregnant following a VF arrest. The second patient (28 years old) presented 38 weeks pregnant with poor cardiac function. In view of deteriorating maternal condition caesarean section was performed. Following delivery she remains in heart failure despite maximal therapy and is currently on the active cardiac transplant list.
Conclusion: Pregnancy is extremely high risk for women in NYHA 2, or worse exercise limitation, following Mustard procedure. In our series asymptomatic women with good echocardiographic function tolerated pregnancy well. The natural history of patients following Mustard procedure suggests that earlier pregnancy should be advised.

P138
COArctation Long-term Assessment (COALA-Study): incidence of restenosis and hypertension after surgical repair
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Background: To assess the incidence of restenosis and arterial hypertension in patients after surgical repair of aortic coarctation in a cross sectional study.
Patients and Methods: From 1974 to 2000, 405 patients born before 1.1.1965 underwent surgery for isolated aortic coarctation in our hospital. From those 383 who are still alive 26 moved to remote or unknown areas and 83 denied a follow up examination at our institution. The study group of the remaining 274 patients (16–73 years old, 90 female, 184 male, 72% of those that are still alive), 18.4 ± 5.9 years (range 1–27 years) after surgery, underwent a structured clinical investigation with a Doppler sonographic measurement of the blood pressure at all limbs, exercise test, and ambulatory blood pressure measurement.
Results: 29 patients (11%) already underwent surgery for restenosis, another 20 patients (7%) had a leg-arm gradient of more than 20 mmHg suggesting restenosis now. Risk factors were young age at the first repair and first repair without end-to-end anastomosis. 67 Patients (24%) were already on antihypertensive drug therapy, another 64 patients (24%) had a mean systolic blood pressure of more than 133 mmHg or a diastolic blood pressure of more than 78 mmHg in ambulatory blood pressure measurement and should therefore also be classified hypertensive. Furthermore, another 26 patients (9%) showed a blood pressure during exercise exceeding 2 SD of reference values. All in all, only 117 patients (43%) had a normal blood pressure reaction. The only independent risk factors for an abnormal blood pressure was present status with prosthetic in situ (prosthetic tube graft, patch, stent). Other risk factors like age at repair, age at investigation, and body weight at investigation were not significant in a multiple regression analysis anymore.
Conclusions: In the long-term follow up 18% of patients after surgery for aortic coarctation show restenosis. An arterial hypertensive blood pressure reaction is much more common in long term follow up with an incidence of 57%. In only few patients hypertension is due to restenosis. Best results were achieved in patients in whom an end-to-end anastomosis could be performed and in whom repair was not performed at a very young age.

P139
Mitral regurgitation in adult patients, operated in the past for their complete or partial atrioventricular septal defect
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Purpose: The degree of left AV valve regurgitation is known to be the most important determinant for long-term prognosis in

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patients operated upon for both partial and complete atrioventricular septal defect (AVSD). The surgical techniques for correction of AVSDs have changed over the past 30 years, aiming at a reduction of left-sided atrioventricular valve regurgitation. The aim of the study was to assess the extent of left AV valve regurgitation in adult patients with both partial and complete AVSD.
Methods and Patients: All 102 patients with an AVSD who were seen in the past 5 years at the outpatient clinic adult congenital heart disease of our institution were evaluated retrospectively. Of these, 83 had an echocardiographic examination. Twenty-six had a complete AVSD, 76 a partial AVSD, and 25 had Down syndrome. Twenty-five patients had >1 operation, of which 12 (48%) had a second left sided AV valve plasty and 8 patients (32%) an artificial valve. Age at first surgery was 16,8 ± 15,6 years. Age at last follow up was 30,4 ± 13,6 years.
Results: Mitral regurgitation was absent or mild in 50 patients (60%), moderate (grade 3 regurgitation) in 27 (33%) and severe in 6 patients (7%). The site of maximal regurgitation was at the tip of the leaflets in 33%, and trough a cleft in the anterior mitral valve leaflet (AMVL) in 67%. The mean left atrial (LA) dimension was 42 ± 11 mm, 25 patients (30%) had a LA >45 mm. The mean left ventricular (LV) end-diastolic diameter was 51 ± 8 mm, 29 patients (35%) were >p95 for body length. The end-systolic diameter was 33 ± 8 mm.
Conclusions: A substantial number of patients has a moderate or severe left AV valve regurgitation, mainly through a cleft in the AMVL. The dimensions of the LA and LV are at the upper limits of normal or over it. Since this is an attractive substrate for successful valve repair, these patients should be considered for redo surgery, weighing the disappointing natural history of chronic mitral regurgitation versus the surgical risks, which are low, nowadays.

P140
Evaluation of biventricular function using serial MRI in a patient with atrial septal defect and restrictive left ventricle before and after "conditioning therapy"
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Objectives: Congestive left ventricular (LV) failure after closure of an atrial septal defect (ASD) particularly in the elderly has been described. Administration of diuretics and positive inotropic medication prior to the intervention ("conditioning") might help to prevent congestive heart failure. The aim of this study was to assess the effects of the "conditioning" in a patient with large ASD II and restrictive left ventricle prior to its definite transcatheter closure.
Methods: Serial cardiac MRI was performed in an elderly patient (76 years of age) with ASD. MRI was done without prior medication with open ASD and during balloon occlusion first, then 3 days after "conditioning" with furosemid (0.5 mg/kg/d), dobutamine (5 μg/kg/min) and milrinone (0.5 μg/kg/min) with open ASD and then immediately after transcatheter closure.
Results: With open ASD, good biventricular function and a left to right shunt with Qp:Qs of 2.8:1 was measured. After balloon occlusion of the defect, right ventricular stroke volume (RVSV) and ejection fraction (RVEF) decreased from 126 ml to 60 ml and from 64% to 40%, respectively. Left ventricular stroke volume (LVSV) remained unchanged, but ejection fraction (LVEF) decreased from 68% to 59%, thus, the enddiastolic volume increased due to enhanced preload, but no increase in stroke volume resulted. Three days after "conditioning", Qp:Qs changed from 2.8:1 to 2.4:1. RVSV and RVEF showed no different behaviour, however, LVSV increased 10% and LVEF from 68% to 78%. Immediately after closure of the defect LVSV increased 20% and LVEF remained stable at 77%. RVSV and RVEF decreased similarly as prior seen.
Conclusions: In this patient with large ASD we could demonstrate the inability of a restrictive left ventricle to take over an enhanced preload adequately. An anti-congestive "conditioning therapy", however, enables the restrictive ventricle to adapt promptly to the sudden increase in preload immediately after transcatheter closure of a large ASD. Serial cardiac MRI is an accurate method to measure these changes under variable loading conditions.

Session 11: Psychosocial and Socio-economic Problems

P141
Quality of life in children with congenital heart disease: an individual perspective
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Objectives: To examine Quality of Life (QoL), self-perceived competence and personality characteristics in children with Congenital Heart Disease (CHD) and to explore the role of personality in understanding quality of life.
Study Design: Eighty-nine children with CHD between 6 and 12 years were compared with eighty-one healthy children between 8 and 12 years using 3 psychometric instruments. The mothers of the CHD children were administered the Dutch Pediatric Quality of Life Inventory (PedsQL; Koot & Batiaansen, 1998), the Self-Perception Profile for Children (SPPC; Dutch translation, Veerman, 1997) and the Hierarchical Personality Inventory for Children (HiPIC; Mervielde & De Fruyt, 1999). The CHD children, older than 8 years also described themselves on the PedsQL and the SPPC.
Results: CHD children experienced a higher quality of life and self-perceived competence than the referent group, whereas no differences were found for personality characteristics. Complexity of cardiac surgery, postoperative status and physical complaints varied among the children with CHD. However, only the presence of physical complaints was significantly related to a lower QoL for both parental and self-ratings. Parent-child agreement across groups was high, with the CHD group presenting significantly higher agreement than the referent group. Results indicated that child Neuroticism is a significant predictor of QoL in children with CHD.
Conclusions: The results demonstrate a higher QoL and self-perceived competence in children with CHD. Further research is recommended in children with complex CHD and associated residual complaints.

P142
The participation in summer camp as a part of rehabilitation programme of improving the physical activity in children after cardiosurgical correction of congenital heart defect
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Objectives: To examine the usefulness of participation in summer camp in improving the physical abilities of the children after cardiosurgical correction of congenital heart defect.

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Patients: The group consists of 50 children with the congenital heart defect, most of them after the cardiosurgical correction.. Before the camp patients were classified into proper AHA categories of physical activity on the base of detailed history taking. Participants underwent testing according to exercise protocol. Heart rate and blood pressure were measured before and after each physical activity. The exercise protocol included 60 meters race, 4 kilometers walk on the beach, basketball and/or volleyball match, table tennis or badminton competition and 15 minutes bath in the sea.
Results: All children after correction of CHD performed the programmed exercises without any symptoms of heart insufficiency and with the level of tiredness adequate to effort and actual exercise capacity. The electrocardiograms of all children before and after finishing each physical activity showed neither features of overload nor heart rhythm disturbances. The heart rate and blood pressure during the exercise were within the normal ranges. The distinct difference between the levels of physical abilities was observed while comparing the previous and camp activity of children after cardiosurgical treatment.
Conclusions:

1. The physical activity of children after surgical complete correction of congenital heart defect is often limited only by psychological or social reasons.
2. Participation in the camp supervised by pediatric cardiologist could be the part of standard rehabilitation programme after cardiosurgical operation.

P143

Chest scar and heart scar: how influence of the operative scar on quality of life in adult patients with congenital heart disease

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Background: Advances of medical and surgical management have brought the extended life expectancy of children with congenital heart disease who could not survive without surgical treatment. Currently, the importance would be focused on the quality of life after the surgery. In this regard, we speculated the operative scar may influence on the daily activity or life style in adolescents or young adults.
Objective and Method: To evaluate the awareness for their own operative scar and to clarify the relationship between the severity of scar and their awareness, questionnaires were handed at the outpatient clinic after the routine physical examination. Questionnaires included that how they feel their operative scar, the obstacles or worries for daily life related with operative scar, current physical activity, patient personal profile and wishes for the future operation. The location, length and severity of operative scar were evaluated by 2 cardiologists independently. Questionnaires were handed for consecutive 211 post operative patients, and completed by 138 (65%) patients. There were 46 men and 92 women, and their median age was 28 years. Their cardiac diagnosis was tetralogy of Fallot in 43, VSD in 30, ASD in 17, after valve replacement in 11, complex cyanotic heart disease in 19, and others in 20.
Results: The degree of worries for their operative scar was, mild worries in 41%, moderate in 33%, severe in 6% and completely no worries in 20%. Women had more worries their scar than men had (22% vs. 51%, $p < 0.05$). Such complaints were not related with severity of disease or current NYHA classification. In addition, the complaints were not correlated with the severity, size or location of scar. However, if the analysis limited in only women, the intercostal scar had fewer worries compared to the midsternum scar (15% vs. 65%, $p < 0.05$). The most worried ages was just after the operation in 18%, primary school in 32%, junior high school in 23%, high school in 16% and after high school in 12%. The transition of worries was no change in 55%, decrease in 36%, and increase in 11%. Cosmetic surgery was considered only in 6%.
Conclusions: Complaints of surgical scar was not correlated with the severity or size of scar. Especially in women, the surgical chest scar was very important heart scar in their school age regardless of its severity.

P144

Socioeconomic status and psychosocial behaviour of schoolchildren and adolescents with congenital heart disease in Croatia

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The aims of this study were: to find out about socioeconmic status of the families of the children with congenital heart disease, and to establish the influence of the disease on their physical and psychosocial behaviour in everyday life.
Patients and Methods: The study subjects comprised 153 children with congenital heart disease, median age 11,92 years, 88 female (58%) and 65 male (42%). Number of 106 pts (69%) belongs to urban society and 47 pts (31%) belongs to rural society. The control group consists of 153 healthy schoolchildren and adolescents. For determination of socioeconomic status of the families we used questionnaire fulfilled by parents and for establishing the influence of the disease on physical and psychosocial behaviour we used questionnaires fulfilled by patients.
Results: School achievement of chronic patients is significantly lower than of healthy children. The biggest number of patients with congenital heart disease achieved satisfactory (70% of them, D and C grades), whereas the healthy examinees achieved excellent (48%, A and B grades) and very good results (27%, $P < 0,01$, $CI = 0,48$). Also, children with congenital heart disease are statistically more likely to fail a year (than the healthy ones) (11%:0%; $P < 0,01$, $CI = 0,35$). Their motivation is also decreased, which is shown in lower attendance of extra-curricular activities (B 44%:Z 60%, $P < 0,05$, $CI = 0,22$), as well as in the need to associate with their peers ($P < 0,01$, $CI = 0,81$). The primary disease of chronically ill children also affects the relationships in the family. Parent–child relationship is crucial, especially mother–child relationship ($P < 0,05$, $CI = 0,31$). Excessive protection is present in 66% in patients, and 44% among healthy children, which resulted in some personality changes – lack of independence (82%), tearfulness, sociableness and depression (60%). The family of a chronically ill child with congenital heart disease has considerably lower standard of living in 25% cases, whereas in 36% it is significantly burdened by the disease and its treatment.
Conclusions: It is necessary to change social partners relation towards children with congenital heart disease and their families. This is the only way to avoid problems of social adaptation, integration, and possibilities of further education and future employment.

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P145

Intellectual and social status of schoolchildren and adolescents with congenital heart disease in Croatia

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The aims of our study were to establish the intellectual development of schoolchildren and adolescents with congenital heart disease, and to establish the social maturity.
Patients and Methods: The study subjects comprised 153 children with congenital heart disease, (median age 11,92 years), 88 female (58%) and 65 male (42%), who lived to their school age or adolescence. The survey excluded all patients with congenital heart disease related to syndrome disease which include mental deficiency. The control group consists of 153 healthy schoolchildren and adolescents who are of the same age and sex as the patients involved in the study. For evaluation of cognitive functions of a child following tests were used: a) progressive colour matrix (Raven), in which is by choice of the coincident shapes determined capability of visual perception as a measure of QI, and b) scale of social maturity by Doll, by which is social age evaluated and compared with chronologic age and so determines quotient of social maturity.
Results: The mental status (QI) of children with congenital heart disease who have become chronic patients, significantly differs for the whole group (QIh = 104,97: QId = 99,24, $P < 0,01$), as well for both sexes (men QImh = 105,6: QImd = 99,25, $P < 0,01$), (girl QIgh = 106,25: QIgd = 99,82, $P < 0,01$). The coefficient of social age (SA) of chronic patients with congenital heart disease is significantly different for the whole group (SAh = 13,43: SAd = 10,24, $P < 0,01$), and each of the genders (SAmh = 13,11: SAmd = 9,63, $P < 0,01$), (SAgh = 13,67: SAgd = 10,57, $P < 0,01$). Chronic patients, due to congenital heart disease, have significantly lower social quotient (SQ) compared with the healthy children, applies to the whole group (SQh = 114,59: SQd = 89,29, $P < 0,01$), as well as for both sexes (SQmh = 114,46: SQmd = 86,77, $P < 0,01$), (SQgh = 115,48: SQgd = 90,53, $P < 0,01$).
Conclusion: Teamwork of all the subjects involved – paediatricians, cardiologists, cardio-surgeons, psychologists, social workers, psychoterapeutists, nurses and parents – is essential if we are to solve this problem. Congenital heart diseases are public health problem due to the fact that an increasing number of these children become chronic patients in adult age, proven by study.

Session 12: Developmental Anatomy and Morphology of CHD

P146

The prevalance and spontaneous closure rate of isolated ventricular septal defects in newborns by echocardiographic screening

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Aims: This study was designed to determined the prevalance, risk factors and timing of spontaneous closure (SC) of isolated ventricular septal defects (IVSD) in neonates by using color flow Doppler echocardiographic screening.
Methods: 1074 neonates (128 preterms and 946 terms) were examined within the first 96 hours by CFDE. Detailed history was taken to evaluate the risk factors and the cases with IVSD were followed to determine the rate of spontaneous closure during the first years of life.
Results: IVSD was found in 51 of 1074 neonates, in a prevelance of 47.4/1.000 live births, 8 cases were preterm neonates (<37 weeks) and a prevalance of 62.5/1.000 live births and 43 cases were term neonates and a prevalance of 45.4/1.000 live births. Gestational ages and birth weights of IVSD cases ranged 32 to 41 weeks and 1519 to 4100 gr, respectively. Only one baby's mother had ventricular septal aneurysm and one baby with IVSD had Down syndrome. By echocardiography, 45 cases had a trabecular (apical: 25 and midmuscular: 20), 5 cases had perimembraneous and one cases had doubly-committed VSD. 9.8% cases had two IVSDs. The defect's sizes ranged 0.1 to 6.8 mm in diameter. 49% defects were detectable only with CFDE imaging. Minor cardiac defects (peripheric pulmonary arter stenosis and atrial septal aneurysm) were determined in 7.8% cases. During the first years of life, SC was seen in 89.5% (n: 43/48) cases. SC rate was found 100% for preterms and 87.5% for term babies. SC was most commonly 72% occured within the 6th months. Closure rate of trabecular IVSD was 87.2% (apical VSDs: 82.8% and midmuscular VSDs: 84.2%). Closure rate of perimembraneous and subpulmonic IVSD were 80% and 100% respectively. One cases with large perimembraneous IVSD underwent an operation at the 6th months, but died at the 9th postoperative days.
Conclusions: The prevalance of IVSD was considerably high in neonates ecpecially preterms when routine echocardiographic examination was performed. Despite increased prevelance of IVSD spontaneous closure rate was remarkably high within the first year of life. These defects seem to result from delayed physiologic development and have fairly good prognosis.

P147

Ghrelin expression during prenatal cardiac development

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Ghrelin is a novel growth factor acting on GH-IGF-1 axis, with cardioprotective effects, which has been proposed as a therapeutic approach for the treatment of heart failure in adults. Since GH and IGF-1 are implicated in cardiac development, our aim was to investigate cardiac ghrelin expression during prenatal development.

Sprague-Dawley rats were mated and checked daily for introit plugging. Whole embryos and fetal cardiac tissue were harvested at 12, 14, 16, 18, 20 and 22 dpc (days post-conception). After fixation, 12 dpc embryos were stored at −20°C or immediately used for whole mount in situ hybridization. Remaining embryos/foetuses were dissected, fixed and prepared as paraffin sections for in situ hybridization. Ghrelin riboprobe was synthesized from plasmid pGEM-T#ghrelin containing a 247-bp fragment of rat preproghrelin cDNA. Following immunocytochemical reaction with anti-digoxigenin antibody, mRNA was labelled by NBT/BCIP complex. Paraffin sections were counterstained with Nuclear Fast Red. Adult gastric fundus sections were used as positive controls and non-specific probes were used as negative controls.

Ghrelin expression in the heart, during embryonic and fetal development, has a distinct pattern according to stage. Early, at 12 dpc, ghrelin was identified in aortic arches, suggesting its involvement in migration of cardiac neural crest cells. In cardiac structures, ghrelin expression begins at 14 dpc and remains until the term of gestation, at 22 dpc. Its expression is maximal at stages 14 and 16 dpc. Ghrelin expression is restricted to myocardium, being entirely

absent from endocardic cushions or valvular structures, with a sharp boundary between expressing and nonexpressing cells. Ventricular expression is more relevant than auricular; however no left-right asymmetry was detected.

We describe, for the first time, ghrelin expression during cardiac prenatal development. Our results suggest that ghrelin may play an important role in heart and great vessels morphogenesis in early stages and lately in myocardial growth. Additional studies are requested in order to clarify ghrelin's rule in the aetiology of congenital heart diseases and cardiomyopathies with neonatal presentation.

P148

Spontaneous closure of apical muscular ventricular septal defects

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Background: The rate of spontaneous closure of apical VSD's is not defined very well and may have been underestimated in the past.
Methods: To determine the frequency of spontaneous closure, we retrospectively analyzed 45 cases with apical VSDs diagnosed by color flow Doppler echocardiography and they were followed up at least 2 years to determine the closure rate.
Results: Ages of 45 cases ranged between 1 day and 13 years old. Initial examination was performed in 22 cases within the newborn period, in 9 cases between 1 and 6 months of age and in 14 cases $>$ 6 months of age. Spontaneous closure was seen in 24 of 45 cases (53.3%) between 1 and 36 months of ages and it was most commonly recorded during the first 6 months. It was remarkable that spontaneous closure was occurred in 20 of 22 cases (90%) diagnosed in the neonatal period.
Conclusions: The rate of spontaneous closure in cases diagnosed in the neonatal period is higher than previously believed. It is advisable to follow up patients to determine the spontaneous closure especially within the first two years of life.

Session 13: General Paediatric Cardiology

P149

Comparative study of myocardial scintigraphic image and endomyocardial biopsy findings in patients with Kawasaki disease

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Purpose: To investigate the relationship between coronary artery lesions (CAL) and myocardial damages, 201TLCL (TL) and 123I-BNIPP (BMIPP) dual isotope single photon emission computed tomography (SPECT) and endomyocardial biopsy (EMB) were performed in the patients with Kawasaki disease (KD).
Materials and Methods: They included 20 patients with giant coronary aneurysm (G-AN); the male to female ratio was 15:5 and 32 patients with CAL; the male to female ratio was 23:9. Ages ranged 2 to 18 years. Histopathology was evaluated with histomorphometric method using computer to calculate the diameter of myocytes, % area of fibrous and fatty tissue and small vessel changes. A defect score(DS) for SPECT images was interpreted as normal: 0, mildly decreased: 1, moderately or severely decreased: 2, complete defect: 3. The difference in DS between TL and BMIPP was defined as the mismatch score (MS).
Results: Myocardial changes on EMB, such as fibrosis, degeneration, disarray and inflammatory cell infiltration, showed 57%, 38%, 65% and 26%, respectively in the patients with G-AN, while 54%, 38%, 38% and 6%, respectively in the KD patients with CAL, including suggested chronic myocarditis. Accumulation of myelin bodies in the myocytes, disarray of myofibrils, vacuoles and microangiopathy were found as ultrastructural changes. TL/BMIPP discrepancy was found 22% in KD with G-AN and 20% in KD with CAL. In the cases TL/BMIPP discrepancy ultrastructual changes revealed massive myelin bodies.
Conclusions: Myocardial changes in the patients with KD include fibrosis, hypertrophy of myocytes, degeneration of myocytes and postmyocarditic changes on light microscope and ultrastractural changes as microangiopathy and myelin bodies even in late stage. Discrepancy of dual SPECT image suggested that myocardial changes in long-standing Kawasaki disease might reflect not only myocardial ischemia but also disordered myocardial fatty acid metabolism following myocarditis.

P150

A clinical trial of steroids in the management of acute myocarditis

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Background: Acute myocarditis is a serious disorder and treatment options are limited and expensive. Myocardial damage in myocarditis is mediated, in part, by immunological mechanisms and high-dose intravenous gamma-globulin (IVIG) are proven to be beneficial. They are, however, very expensive and not feasible for a country like Pakistan where incidence of myocarditis seems to be much higher.
Aim: To determine whether steroids improve outcome in patients with acute myocarditis. Since October 2002, the routine management of presumed acute myocarditis at Children's Hospital, Lahore, Pakistan has included administration of steroids.
Methods and Results: We treated 42 consecutive children presenting with presumed acute myocarditis with IV methylprednisolone over 48–72 hours, in addition to anticongestive therapies. This was followed by oral prednisolone for a period of 3 months. A comparison group comprised 43 control patients meeting identical eligibility criteria but not receiving steroids therapy. Left ventricular function was assessed at admission, at 1 to 3 weeks (at discharge), 3 weeks to 3 months and 3 to 6 months. Follow-up at 6 to 12 months is under way at the moment. At presentation, the both groups had comparable left ventricular enlargement and poor fractional shortening. The mean hospital stay and need for prolonged inotropic support was shorter in steroid treated group as compare to non-steroid group ($P = 0.06$ and 0.05). As compared with the non-steroid group, those treated with steroids had a smaller mean adjusted left ventricular end-diastolic dimension and higher ejection fraction in the 3 to 6 months ($P = 0.005$ and $P = 0.035$, respectively). When adjusting for age, intravenous inotropic agents, and angiotensin-converting enzyme inhibitors, patients treated with steroids were more likely to achieve near normal left ventricular function during the first 6 months of presentation ($P = 0.04$). We observed no adverse effects of steroids.
Conclusions: Our results support routine use of steroids for treatment of acute myocarditis. It is associated with improved recovery

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of left ventricular function and with a tendency to better survival during the six months after presentation. Long-term data is being collected to validate these observations further.

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An experimental study on protective effects of Melatonin, L-Triptophan and Pentoxifylline on Doxorubicin cardiotoxicity

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Doxorubicin (DOXO) is an effective chemotherapeutic agent used in oncology. There are studies on the subject of protection of the most serious side effect of DOXO cardiotoxicity. The cardiotoxicity of DOXO is attributed to elevated free oxygen radicals and lipid peroxidation products, and decreased antioxidant enzyme activities. In this study we aimed to evaluate the protective effect of melatonin, L-triptophan and pentoxifylline on experimentally produced cardiotoxicity.

We gave DOXO in 15 days in 6 doses (15 mg/kg cumulative dose) to the four groups, which each composed of 10 rabbits. The first group received only DOXO, second group received melatonin, third group received L-triptophan and fourth group received pentoxifylline for 21 days addition to DOXO. At the beginning and the end of the study blood samples were taken, at the end of the study after the sacrification the myocardial tissue samples were taken. For determining the cardiotoxicity myocardial and plasma glutathion peroxidase (GSH-Px), superoxide dismutase (SOD), malondialdehyde (MDA), myocardial nitric oxide (NO), serum troponin-I (Tn-I) and creatine kinase-MB (CK-MB) levels were studied. Cardiotoxicity was evaluated with histopathologicaly.

In the first group severe cardiomyopathy was demonstrated with histopathologicaly, GSH-Px decrease, MDA and NO elevation detected. In DOXO-melatonin received group GSH-Px and SOD increase and NO and MDA decrease; in DOXO-L-triptophan received group GSH-Px and SOD increase, in DOXO-pentoxifylline received group GSH-Px and SOD increase, NO decrease detected. Histopathological examination reveled the protective effect of L-triptophan and pentoxifylline on severe cardiomyopathy. There were no significant difference between groups according to the GSH-Px, SOD and MDA levels. Serum Tn-I and CK-MB were elevated only in severe cardiomyopathy, but did not changed in antioxidant-received groups.

Our findings support that, melatonin, L- triptophan and pentoxifylline supplementation has a strong protective effect on DOXO cardiomyopathy.

P152

22q11.2 microdeletion syndrome in children with congenital heart diseases

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Congenital heart diseases (CHD), particularly conotruncal (cCHD) are characteristic for DiGeorge syndrome (DGS), Velo-Cardio-Facial syndrome (VCFS), Conotruncal Anomaly Face syndrome (CTAFS). Molecular studies demonstrated, that they are caused by 22q11.2 microdeletion and are the variants of the same disease with a large spectrum of clinical symptoms.

Aim: The aim of the study was to evaluate incidence of 22q11.2 microdeletion in pts with cCHD and in children with phenotypic symptoms of del 22q11.2 and CHD other than cCHD.
Materials and Methods: Clinical and genetic examinations were done in 250 children aged from 2nd day to 18 yrs. The 22q11.2 microdeletion is diagnosed in pts with a submicroscopic deletion of chromosome 22 detected by fluorescence in situ hybridization (FISH) using DNA probes from the DiGeorge chromosomal region. Clinical studies included: family history, physical and cardiological examination.
Results: All 250 children were divided into 2 groups. GROUP I: n = 204 pts with cCHD (TOF n = 103, TGA n = 55, DORV n = 22, AP + VSD n = 10, TAC n = 12, IAA type B n = 2). In 182 pts isolated cCHD, in 22 pts coexisted with others phenotypic features of del 22q11.2 (dysmorphic face n = 14, anus atresia n = 3, hypospadiasis n = 2, kidney agenesis n = 1, cleft palate n = 2) were observed. GROUP II: n = 46 pts with phenotypic features of del 22q11.2 (dysmorphic face n = 39, hypospadiasis n = 1, kidney agenesis n = 2, cleft palate n = 3, equinovarus feet n = 1) and CHD other than cCHD: VSD n = 10, ASDII n = 18, PDA n = 4, CoA n = 14. In all 250 children 22q11.2 microdeletion was found in 15 (6%) of pts, in 14 (7%) from GROUP I (n = 7 pts with TOF, n = 3 with TAC, n = 2 pts with AP + VSD, n = 1 with DORV, n = 1 with IAA type B) and in 1 (2%) pt from GROUP II (n = 1 child with PDA and dysmorphic face). In 9 out of 15 (64%) pts with del 22q11.2 CHD (mainly conotruncal) coexisted with other noncardiac clinical features typical for 22q11.2 microdeletion syndrome.
Conclusions:

1. The 22q11.2 microdeletion was found in 3% of children with an isolated conotruncal CHD and in 41% of pts with conotruncal CHD coexisted with others phenotypic features of 22q11.2 (DG/VCF) syndrome.
2. The 22q11.2 microdeletion was found in 2% of pts with phenotypic symptoms of DG/VCF syndrome and other than conotruncal CHD.
3. The 22q11.2 microdeletion was diagnosed in 10% of newborns, 8% of infants and in 4% of children older than 1 year.

P153

Age-associated changes of stiffness of abdominal orta

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Objectives: The assessment of the mechanical properties of the aorta in adults is important for early detection of atherosclerotic changes. Age-associated changes in pulsatile components of the arterial system have studied rarely in children. In this study, the changes in aortic stiffness and distensibility were established.
Methods: Aortic strain (S), pressure strain elastic modulus (Ep) and normalized Ep (Ep*) in 300 healthy children (20 equal gender children for each age; ranging between 2 and 16 years) were measured by using sphygmomanometer and two-dimensional transthoracic echocardiography (2DE). We recorded systolic (Ps) and diastolic (Pd) blood pressure and measured aortic diameter (Dd) at both minimum diastolic pressure and maximum systolic expansion (Ds) by echocardiography. These measurements were used to determine aortic strain S = (Ds − Dd)/Dd, pressure strain elastic modulus Ep = (Ps − Pd)/S and normalized Ep Ep* = Ep/Pd.
Results: It was found that aortic distensibility varies with age. The value of S was decreased while the value of Ep and Ep* was increased with age.

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Conclusions: Age-associated changes in aortic distensibility and stiffness were observed in childhood. These changes may be related with normal maturation or beginning atherosclerosis. These measurements can be used as reference ranges in later studies.

P154

Alterations of lung function in infants with univentricular circulation

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Infants with congenital heart disease and univentricular circulation often suffer from obstructive airway disease with the need for hospitalization. Optimized pulmonary function and circulation are essential for completion to modified Fontan. However, infant lung function tests are time-consuming and difficult to perform due to the lack of active cooperation and need for sedation.
Aim of Study: To trace measurable lung and airway alteration in these infants with univentricular circulation.
Patients and Methods: In a prospective clinical study om 25 infants (16 m, 9 f, mean weight 8.2 kg, mean SaO_2 82%) we investigated small and central airway function, forced lung volumes and -flows. Patients were examined by means of babybodyplethysmography, multiple breath inert gas washin/washout and raised volume rapid thoracic compression technique. We looked for obstruction, hyperinflation or gas mixing inhomogeneities.
Results:
(Mean ± SEM)

FRC_{pleth} %	FRC_{SF6} %	sReff %	$FEV_{0.5}$ %	MV/kg ml/kg/min	TV/kg ml/kg	BF /min
90.6 ± 6.7	109.8 ± 5.0	109.4 ± 13.3	78.4 ± 8.0	391.8 ± 19.5	10.8 ± 0.7	39.2 ± 2.9

FRC functional residual capacity, **pleth** plethysmographic, **SF6** inert gas washin/washout, **sReff** specific effective airway resistance, **$FEV_{0.5}$** forced expiratory volume at 0.5 sec (FEV0.5) ($p > 0.001$) indicating, **MV** minute volume, **TV** tidal volume, **BF** breathing frequency.

Discussion and Conclusions: In 25 infants with congenital heart disease and univentricular circulation we found a signifcant reduction of forced expiratory volume at 0.5 sec ($FEV_{0.5}$) ($p > 0.001$) indicating small airway obstruction. Mildly reduced functional residual capacity (FRC_{pleth}, static measurement) indicates significantly smaller FRC in the study group ($P = 0.034$, compared to normal) as a hint of retarded alveolar growth. As there is no significantly lower functional residual capacity in multiple breath inert gas washin/washout measurement when compared to FRC_{pleth}, no ventilation inhomogeneities or dystelectasis could be traced as reason for frequent infections. The shown small airway obstruction however explains the observed increased susceptibility to pulmonary infections of these infants.

P155

Screening for duct-dependent congenital heart disease with pulse oximetry: a critical evaluation of strategies to maximise sensitivity

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Background: With the increasing trend towards early neonatal discharge of well term babies, an increasing proportion of newborn babies are being discharged with unrecognised serious congenital heart disease. In our referral region a review showed that in 1999–2001 29% of infants admitted with critical congenital heart disease to our supraregional centre had left the neonatal nursery undiagnosed.
Aim: Evaluation of the feasibility of detecting duct-dependent congenital heart disease before hospital discharge by using pulse oximetry.
Methods: Pulse oximetry was performed in the right hand and one foot using a new generation oximeter (Masimo Radical SET) and a conventional oximeter (Datex-Ohmeda TuffSat). Two groups of patients were studied; 200 term newborns in the well baby nursery with echocardiographically normal hearts (median age 1 day) and 53 infants with critical congenital heart disease, CCHD (median age 3.5 days).
Results: Normal newborns had a median postductal saturation of 98% (94 to 100) with the new generation oximeter. Intra-observer variability showed a mean difference of 0% with a SD of 1.5%. The conventional oximeter showed too great a variability in the normal infants to be used for screening, with a significantly greater proportion of postductal values below 95% (41% versus 1%) compared to the new generation oximeter ($p < 0.0001$). The CCHD group had a median postductal saturation of 90% (46 to 99) with the new generation oximeter. Analysis of frequency distributions suggested a cut-off of <95% for screening, this however still gave 7/53 false negative patients with arch obstruction. Best sensitivity was obtained by combining two criteria: either a saturation <95% in both hand and foot or a difference of >3% between pre- and postductal saturation counting as a positive screening result. These criteria reached a sensitivity of 98%, a specificity of 96%, a positive predictive value of 87% and a negative predictive value of 99.5%. Repeated measurements on some false positive individuals suggest that the false positive rate could probably be reduced substantially by repeating the saturation measurement a few hours later in positive subjects.
Conclusions: Systematic screening for CCHD using a new generation oximeter is effective, and the detection of CCHD is substantially improved when saturations in both right hand and one foot are compared.

P156

Respiratory syncytial virus (RSV) infection in infants with significant congenital heart disease

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Background: Infants with congenital heart disease are considered to be especially at risk during RSV infections, but information of the course of RSV disease in this patient group is scarce. We found it important to study the occurrence and the course of RSV infections in infants with significant heart disease before possible prophylaxis with palivizumab (Synagis®) is started in this patient group.
Methods: Infants who had surgery for congenital heart disease during the first two years of life at our institution from January 1999 to June 2002 were studied retrospectively with regard to type of heart defect, surgery, diagnostic method of possible RSV infection, length of hospital stay due to RSV infection, need of oxygen treatment, CPAP – or mechanical ventilation treament, or concomitant extracardiac disease.
Results: 420 infants were included of whom 26 (6.2%) had been hospitalized due to RSV infection. The length of inhospital care varied from 3 to 95 days (median 18 days). 19 infants had oxygen treatment during 1–67 days (median 12 days). Eight patients were treated in an intensive care unit during 1–26 days (median 7.5 days).

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One child had CPAP treatment and five had mechanical ventilation support 1–22 days (median 15 days). The cardiac malformation was part of a syndrome or complicated by pulmonary disorders or other extracardiac diseases in 123 out of the 420 children (29.3%). Adjusted for follow-up time, significantly more children with extracardiac diseases were hospitalized due to RSV infection 17/123 (13.8%) compared to those without complicating disorders 9/297 (3.0%); $P < 0.001$ using a log-rank test. Among those needing inhospital care no specific heart defect was overrepresented. No child died in immediate conjunction with the RSV infection.
Conclusions: Among infants who had heart surgery before two years of age 6.2% were hospitalized due to RSV infection. The need of inhospital care was significantly more common in infants with significant congenital heart disease in combination with an extracardiac disorder (13.8%) as compared to heart diseased infants without any other disorder (3.0%, $p < 0.001$). This may be important to consider before recommending RSV-prophylaxis with palivizumab (Synagis®) to infants with congenital heart defects.

P157
Cardiologic findings in adolescents with anorexia nervosa

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Introduction: AN in adolescents can be associated with cardiac changes, such as thinning of the LV wall, reduction of the LV mass index (LVMI), pericardial effusion, mitral valve prolapse and ECG abnormalities.
Methods: 19 adolescent girls with AN (14.6 ± 1.52 y) BMI (15.8 ± 2.37 kg/m^2) with self-induced starvation were referred. None of them had clinical symptoms of cardiovascular disease and all were studied in the acute state of the disease.In the study we included 15 controls (10.1 ± 1.40 y) BMI (20.5 ± 3.78 kg/m^2). None of the controls had a family history of cardiac diseases or cardiotoxic medication. Patients and controls underwent a 2-D-echocardiography (Vingmed 725 Sonotron 3.25 transducer) and 12 lead ECG (Hp 4700 A system). QT intervals were measured manually in blinded fashion by 2 pediatric cardiologists and for each lead the average of 3 consecutive QT intervals was calculated according to Bazett's formula (QTc = QT/square root RR) and the QTc dispersion was calculated. Standard formulas were used to calculate the LVM = (0.8 × (1.04((LVEDd + IVSd + LVPWd)^3 − (LVEDd)^3)) + 0.6). To correct for linear growth LVM was converted to LVMC by the method of the Simone (LVM divided by height to the 2.7 power).
Results:

	Patients	Controls	p value
Number	19	15	
Age (y)	14.6 ± 1.52	10.1 ± 1.40	0.277
BMI (kg/m^2)	15.8 ± 2.37	20.5 ± 3.78	<0.001
HR (bpm)	56.1 ± 16.3	73.5 ± 0.6	0.001
SBP (mmHg)	96.9 ± 12.7	109 ± 11.2	0.008
Weight loss (%)	23.0 ± 8.94		
LVM/H2.7 (g/m2.7)	28.6 ± 6.37	31.5 ± 7.1	0.210
QTc disp. (msec)	42.2 ± 27.3	26.2 ± 11.0	0.010
R wave V6 (mm)	6.5 ± 3.01	15.6 ± 4.06	<0.001

Conclusions: SBP, HR, and R wave (V6) are all significantly reduced, while QTc dispersion is significantly increased in AN. (1) A possible mechanism influencing increased QTc is alteration of ion channels,active in cardiac repolarization. (2)Those findings are associated with an increased risk of ventricular arrhythmias and sudden death. This warrants a systematic cardiologic work-up in those patients.
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P158
Endothelial function and intima media-thickness, predicting the risk of atherosclerosis in children after aortic coarctation repair and in children with obesity

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Background: Children after repair of aortic coarctation (CoA) and obese children are under suspicion to be at high risk for early development of atherosclerosis for different reasons. A great number of factors define a potential atherosclerotic risk profile. Impaired flow-mediated vasodilation (FMD) and increased intima-media thickness (IMT), are known as surrogate parameters of coronary atherosclerosis and have been shown in children with certain risk factors (RF). Data if different RF profiles lead to certain vascular changes are lacking.
Methods and Results: We investigated 28 children late after successful repair of CoA, 20 obese children in comparison to 30 control subjects. All children underwent the same 2 day screening procedure, with a broad cardiovascular RF profile and FMD/IMT measurements. CoA-children as with obesity had significant ($p < 0{,}001$) impaired FMD (4,87 + 2,6%, 4,96 + 2,8% versus 10,2 + 3,1%) and higher values of IMT than controls (0,52 + 0,08 mm, 0,55 + 0,12 mm versus 0,43 + 0,06 mm), respectively. Measures of FMD and IMT in obese and CoA-children showed no difference but their risk profiles did. Blood pressure at rest and exercise and left ventricular mass were significantly elevated in both groups. Obese children had a number of additional elevated RF with influence on FMD/IMT.
Conclusions: This study has shown that nearly similar vascular wall changes are already present in children after CoA repair and children with obesity despite different risk profiles. A broad risk factor screening in combination with FMD and IMT is sufficient to define children at high risk for atherosclerosis and to control the effect of preventive actions on the vascular wall in childhood and youth.

P159
Changes in airway and parenchymal mechanics following surgical repair of congenital heart disease in children

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It has been now well established that pulmonary haemodynamical changes affect the lung function, however conflicting results have been reported on the effects of surgical repair of congenital heart diseases (CHD) in children. Nevertheless, changes in the lung

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function has been characterized by measuring global lung mechanical parameters. Therefore, we characterised the effect of changes in pulmonary haemodynamics on the airway and tissue mechanical parameters separately in two groups of children with CHD. Children with lesions associated with high pulmonary blood flow and/or pressure (group HP, n = 12), and children with hypoperfused lungs (tetralogy of Fallot or pulmonary atresia) (group LP, n = 11) were included in the study. Forced oscillatory input impedance of the respiratory system (Zrs) was measured between 0.4 and 12 Hz immediately before sternotomy and after chest closure. Airway resistance (Raw), tissue damping (G) and elastance (H) were extracted from the impedance spectra by fitting a linear model to the Zrs data. In group LP, a postoperative reduction in pulmonary blood flow and/or pressure resulted in an immediate decrease in the airway tone, which was reflected in the significant decreases in Raw (−29.0 ± 9.3%, mean ± SE). In contrast, in the children assigned to group LP, surgically re-established pulmonary haemodynamics induced increases in Raw (20.8 ± 18.1%). With substantial interindividual variability, no statistically significant change was observed in G in either group, HP (6.4 ± 13.1%) or LP (31.4 ± 24.6%). H increased in children of both HP (23.0 ± 8.1%) and LP (40.2 ± 6.1%) groups indicating a marked stiffening of the lungs during the surgery. These findings suggest that preoperative pulmonary haemodynamic conditions determines the changes in airway resistance and tissue damping and elastance: surgical repair of CHD leads to an improvement in lung function in children with congested lungs, whereas CPB and/or the mechanical ventilation deteriorate the airway and tissue mechanics in children with hpypoperfused lungs. Furthermore, changes in H are likely to be attributed to the adverse effects of mechanical ventilation and/or cardiopulmonary bypass (CPB) rather than to the modification of pulmonary flow and/or pressure.

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P160

Anterosuperior diverticula of the right ventricle

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Objectives: In the majority of cases anterosuperior diverticula of the right ventricle (RV) are associated with congenital heart disease, especially ventricular septal defects or tetralogy of Fallot. Almost always RV diverticula have been diagnosed unexpectedly during preoperative evaluation or surgical correction of the associated cardiac anomaly.

Methods: In this retrospective study we analysed the data of all 5 pts. with RV diverticula who were diagnosed in 2 tertiary referral centers from 1982–2003.

Results: Mean age at diagnosis was 5.7 months. None of the pts. had symptoms related to the diverticulum. The diagnosis was established by cardiac cath in all. Retrospective analysis revealed, that the diagnosis could have been established in each case by subcostal echocardiography. In all pts the diverticulum was situated at the junction of the inflow and infundibular portions of the RV and contracted during systole. 4 pts had a large perimembranous vsd, in 2 of them the RV opening of the diverticulum was in close proximity to the vsd resulting in a direct communication of the LV with the diverticulum. In the remaining pt the diverticulum was connected with the LV by a tunnel-like structure originating from the left ventricular outflow tract. In this pt the diverticulum communicated with the RV by a restrictive opening. 4/5 pts underwent surgical correction (mean age 7.2 mths): 1 pt underwent patch closure of the vsd, the diverticulum was left untouched. In 3 pts the vsd was closed through an incision of the diverticulum followed by patch closure of the communication to the RV in two of them. There were no postoperative complications especially no arrhythmias during a mean follow-up of 7.6 years. Surgical correction was rejected by the parents in the pt with the diverticulum connected to the LV, the girl remains asymptomatic at the age of 5.4 years.

Conclusions: Although exceedingly rare anterosuperior diverticula of the RV represent a specific congenital cardiovascular anomaly. There are significant variations however in the connection of the diverticulum to the RV requiring different surgical strategies. In pts undergoing surgery of vsd without invasive diagnosis, anterosuperior RV-diverticula should be excluded by subcostal echocardiography, full evaluation of RV-diverticula still requires angiography.

P161

An experimental study on protective effects of melatonin, L-Triptophan and Pentoxifylline on hypoxic heart

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Hypoxia has different degrees of effects on all organ systems. The pathogenesis of the hypoxic heart injury is related with elevation of free oxygen radicals and lipid peroxidation products. In this study we aimed to evaluate the protective effect of melatonin, L-triptophan and pentoxifylline on experimentally produced hypoxic cardiac injury.

In this study four study groups composed of 10 rabbits were taken. The first group only exposed to hypoxia, second group received melatonin, third group received L-triptophan and fourth group received pentoxifylline for 3 days addition to hypoxia. At the beginning and the end of the study blood samples were taken, at the end of the study after the sacrification the myocardial tissue samples were taken. For determining the effects of hypoxia on heart myocardial and plasma glutathion peroxidase (GSH-Px), superoxide dismutase (SOD), malondialdehyde (MDA), myocardial nitric oxide (NO), serum troponin-I (Tn-I) and creatine kinase-MB (CK-MB) levels were studied. Cardiotoxicity was evaluated with histopathologicaly.

In the first group severe cardiomyopathy was demonstrated with histopathologicaly, GSH-Px and SOD activity decrease, MDA and NO elevation detected. Significant troponin-I, CK-MB and LDH elevations were detected in this group. In second group: hypoxic and melatonin received group, the serum troponin-I levels increased clearly, myocardial NO and MDA levels increased slightly. In third group: hypoxic and L-triptophan received group, there was a slight increase in NO and MDA levels, and the change in the other parameters was insignificant. In fourth group: hypoxic and pentoxifylline received group, the change in myocardial and plasma parameters were insignificant. Histopathological examination reveled the protective effect of melatonin, L-triptophan and pentoxifylline on severe cardiomyopathy.

Our findings support that, there is a clear effect of free oxygen radicals and lipid peroxidation products on hypoxic cardiomyopathy and melatonin, L-triptophan and pentoxifylline supplementation has a strong protective effect on hypoxic heart.

P162

Thromboembolic complications after the Fontan operation: results of a risk-adjusted prophylaxis

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Background: Thrombotic/thromboembolic events had been reported as a major cause of morbidity after the Fontan procedure. There is no consensus concerning the postoperative mode and duration of anticoagulation prophylaxis. In a combined retro- and prospective study we evaluated the results of a risk-adjusted prophylaxis taking into account potential predisposing risk factors or specific sequelae.
Methods: We evaluated 142 surviving patients who had undergone a total cavopulmonary anastomosis from 1988 through 2002: A lateral tunnel procedure was performed in 21 patients (38 with fenestration), an extracardiac autologous conduit in 21 patients. Mean follow-up was 91.1 + 43.9 months. Anticoagulation strategies were chosen according to potential risk factors: 40 patients after a lateral or autologous tunnel procedure were free of any anticoagulation treatment. 74 patients with prosthetic material for creation of the intraatrial tunnel and/or tunnel fenestration received acetylsalicylic acid while 28 patients with specific risk factors or complications were placed on coumadin.
Results: Thrombotic events occurred in 10 patients (7%) with systemic venous thrombus formation in 8 (5.6%) and stroke in 2 patients (1.4%) with a peak incidence during the first postoperative year. Expected freedom from thromboembolic event was 92% at 5 years and 79% at 10 years. One of 40 patients without any anticoagulation medication, but none of the 74 patients receiving acetylsalicylic acid presented with thrombotic complications. From 28 patients under coumadin therapy one asymptomatic patient with severe protein-losing enteropathy developed a right atrial thrombus. 8/10 patients were under heparin-therapy mainly for temporary postoperative immobilization. There was no association to coagulation factor abnormalities. Protein-losing enteropathy was present in 4/10 patients.
Conclusions: After total cavopulmonary anastomosis the majority of thrombotic/thromboembolic events occured during immobilisation despite heparin-therapy. A risk-adjusted anticoagulation strategy taking into account surgical technique and potential risk factors proved effective in the prevention of late thrombotic complications. Anticoagulation medication is not routinely necessary during long-term follow-up.

P163

NT-proBNP levels in umbilical cord blood

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Aims: To establish reference values for umbilical cord NT-proBNP levels; to investigate placentar physiology of NT-proBNP; to investigate the relation between umbilical cord blood pH and NT-proBNP.
Methods: Blood pH and NT-proBNP levels were measured in venous and arterial samples drawn from the umbilical cord in 56 successively born neonates with gestational age ranging from 26 to 42 week. NT-proBNP levels were measured using the Roche Elecsys' method. Umbilical cord pH was bedside monitored
Results: NT-proBNP levels were not significantly different for venous and arterial umbilical cord blood samples: venous blood 79.9 ± 50.0 pmol/l (mean ± SD) and arterial blood 79.6 ± 42.9 pmol/l. We did not find any relationship between NT-proBNP and umbilical cord blood pH or gestational age. Preliminary results show that the NT-proBNP levels in serum from healthy mothers are around 10 pmol/l, which is signficantly lower than the levels found in umbilical cord blood.
Conclusions: (1) NT-proBNP concentrations in umbilical cord blood are not correlated to perinatal asphyxia as detected by a decreased pH. (2) Arterial and venous umbilical cord blood NT-proBNP levels are not significantly different. (3) The high fetal cardiac load is reflected in the high level of umbilical cord blood NT-proBNP concentration. (4) The observed difference between the NT-proBNP levels in mother and child suggest that NT-proBNP does not have placental exchange.

P164

The specifics features of the etiological diagnosis and antiviral immunomodulator treatment at children with acute viral myocarditis

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Many specialiyed studies proved the main role of Coxsackie B virus (CVB) in human myocarditis. The clinical diagnosis of viral myocarditis in children is based on a complex clinical and laboratory markers, but the confirmation of the viral agent is very difficult.
Objective: We proposed to appreciate the presence of viral trunks of Coxsackie B in the cases of acute myocarditis in children and efficiency of the antiviral therapy.
Methods: We have observed 65 patients with the diagnosis of acute viral non rheumatic myocarditis by the affirmed data of clinical examination, echocardiography, electrocardiography, laboratory tests with definition of activity of enzymes are shown: total LDH, its main isoenzymes LDH1, LDH2 and CFC, fraction MB-CFC. We have determined the following immunologic parameters: immunoglobulins of classes A, M and G, and T-lymphocytes and population O-helpers, O-suppresser, index O-h/T-s and also interferon's status with definition of serum IF and the level of IFg and IFa. Alongside with these examinations we were performed the virology researches on extraction and identification of enteroviruses from stool samples and glut's wads. We performed the SN tests for detection of the titres of antibody to Coxsackie A5. In depending on type of therapy the patients were divided into 3 groups: I – receiving antiviral treatment such Interferon a (Viferon in rectal suppositories); II – standard therapy; III – non treatment.
Results: The virology tests have determined by increase of an antiserum capacity against a virus Coxsacki A5 more than 4 times in 52, 94 % of cases. From bioassays we didn't detect the enteroviruses. The estimation of immunological parameters and interferon's status has defined a secondary immunodepression with a different degree of manifestation. For patients receiving antiviral therapy (drug Viferon) the activity of acute process has decreased faster in comparison with other two groups. We did not detect any adverse effect at application of Viferon in suppositories.
Conclusions: Our study confirm the virus Coxsacki B5 frequently implication in acute myocarditis in children. The application of antiviral treatment such as Interferon a (Viferon) contributes to

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the improvement of clinical status and the prevention of the possible complications.

P165

Glycemia and QT interval in young type 1 diabetic patients: what is the relation?

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Objectives: The influence of changes in glycemia on the length of the QT or QTc interval remains a controversial issue. We evaluated this relation combining QT values of 24 hours Holter recordings and continuous glucose monitoring values in children and adolescents with type 1 diabetes.
Methods: A 24 h ECG was recorded in 8 (M/F: 5/3) diabetic children and adolescents (median 12 y, range 9 to 19), with automatic QT, RR and QTc measurements. Simultaneously, glycemia (interstitial glucose value using a subcutaneous abdominal catheter) was measured using the Continuous Glucose Monitoring System (CGMS, Minimed®). This technique provided 12 glucose measurements hourly.
Results: High values for QT and QTc were found, with % QTc > 440 ms up to 56%/24 h and % QTc > 460 ms up to 31%/24 h, with a maximum up to 578 ms. A diurnal pattern was found for glycemia, with the lowest values at night and in the early morning. The opposite curve was observed for QT pattern over 24 h, with the higher QT values at night and in the morning, partially leveled out when correcting for heart rate (QTc = QT/square root RR). Plotting QT, QTc and QTc max against glycemia, a significant correlation was found between QT and glycemia in 5/8 patients, between QTc, QTc max and glycemia in 3/8 patients.
Conclusions: QT and QTc prolongation was confirmed in young type 1 diabetic patients. This prolonged repolarisation seems to be related to low glucose values in some of our patients. These data support the evidence of other reports that prolonged cardiac repolarisation contributes to sudden death associated with nocturnal hypoglycemia in young people with diabetes.

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Long-term follow-up after coarctation of the aorta repair

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Aim of Study: Long-term evaluation of patients after coarctation repair.
Methods: 104 patients (69 M, 35 F), aged 4,4 to 48,1 years (mean 16,4 years), after repair of coarctation of the aorta in 1967–1999, were included in the study. Mean age at time of operation was 6,1 years (range 12 days to 17 years), mean age at follow-up was 10,0 years (range 4 to 36 years).
Results: Patients were divided into 3 groups. Group I – normotensive patients with good hemodynamic result (72 pts, 69%), group II – hypertensive patients with good result (25 pts, 24%) and group III – hypertensive patients with recoarctation (7 pts, 7%). Suspected risk factors of recoarctation (age at surgery, the years when operation was performed and methods of treatment) and late hypertension (preoperative blood pressure values, age at operation and duration of follow-up) were analysed. In our data predictors of recoarctation were: repair before 1970, repair < 1 month of life and treatment with balloon dilatation of native coarctation. In patients with good hemodynamic result the risk for being hypertensive at late follow-up was increased for those operated in older age and those with higher preoperative blood pressure values. Percentage of hypertensive patients increased significantly with the follow-up period. We also observed regression of left ventricle hypertrophy. At the time of repair LVH was present in 60% of patients comparing to 18% at our last examination. Six subjects had to be treated because of coexisting aortic valve abnormalities and five patients were operated on because of aortic aneurysm.
Conclusions:

1. Results of coarctation of the aorta repair are satisfactory in most patients.
2. Some of the patients require reintervention because of recoarctation in repair site or in transverse aortic arch, aortic valve abnormalities or aortic anerysm.
3. Recoarctation rate is higher in patients operated in neonatal period and in those treated with native balloon dilatation.
4. Systemic hypertension is the main late cardiovascular complication. It may be caused by restenosis of the aorta, but occurs also in many subjects without signs of recoarctation. Older age at repair and higher preoperative blood pressure values were associated with an increased risk of hypertension at follow-up. Hypertension rate increased significantly with the follow-up period.

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Low (300 Hz) versus very high (4000 Hz) sampling rate electrocardiograms in children: an intra-subject comparative study

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Aim: The normal variation of ECG measurements with age is essential for proper interpretation by the clinician. Electrocardiograms were previously recorded with low sampling rates. Introduction of standard higher sampling rates can potentially unmask higher voltage or different ECG duration parameters. We set out to evaluate intra-subject differences in ECG parameters obtained with a low and a very high sampling rate ECG recording.
Methods: We prospectively enrolled 60 subjects, 48 healthy and 12 with heart disease. For each subject two 12-leads ECG's were recorded, one with a digital acquisition system with a sampling rate of 300 Hz, another with a digital recording system at a sampling rate of 4000 Hz per channel. For both systems frequency response: 3 dB at 0.01 to 150 Hz. The same electrode leads were used for the two recordings stored on disk using a scale 20-mm/1 mV. P, Q, R and S amplitude were measured in each praecordial and limb lead including PR, QRS and QT duration, QTc Bazett calculated and QRS and T axis. Descriptive and analytical statistics were calculated, significance level set at $p = < 0.05$.
Results: Mean age 7.68 years, median 7.3, range 0.15–16.9 years. A significant difference between ECGs were found for QAVF ($p = 0.028$), RV1 ($p = 0.049$), RV6 ($p = 0.044$), SAVR ($p = 0.001$) and QRS duration ($p = 0.005$) values being higher for ECG sampling rate of 4000 Hz. Differences were unrelated to age or presence of pathology.
Conclusion: This intra-subject study comparing ECG sampling rate shows significantly higher Q and R waves voltages in some leads and prolonged QRS duration with ECGs obtained at 4000 Hz. This needs to be taken into account in clinical practice. A study

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comprising a large group of subjects is required to validate these differences and establish the need for new norms. Further study is in progress in the very young and low birth weight babies, where these differences may be more important.

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Simple exercise testing is useful for the diagnosis of pulmonary AV malformations in patients with left atrial isomerism

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Background: Residual cyanosis after superior cavopulmonary anastomosis (CPA) in patients with left atrial isomerism (LAI) and interrupted inferior vena cava can progress because of pulmonary arteriovenous malformations (PAVM's). Resolution can occur after redirection of hepatic venous blood in the pulmonary circulation. Our aim was to find a simple clinical tool for early diagnosis and follow up of patients developing PAVM's.
Patients and Methods: We reviewed a now 3.5y. old male and a 9.5y. old female with LAI, polysplenia, interrupted IVC with azygos continuation and complex congenital heart disease who had developed PAVM's after CPA (age at operation 1.0 and 2.8y. respectively). Regular exercise testing (ET) consisted of running up and down two flights of stairs. In our male patient resting saturations dropped from 93% (after ET: 92%) shortly after CPA to 84% (67%) 15 months later. In our female patient saturations were 94% (94%) after CPA and during the first 48 postoperative months resting saturations decreased to 84% (80%). After 60 months they were 80% (64%). When saturations dropped < 70% at ET (at 15 and 60 months postoperative) pulmonary angiography and pulmonary bubble contrast echocardiography were performed and demonstrated significant PAVM's. Surgical inclusion of hepatic veins in the pulmonary circulation was performed at the age of 2.9 and 8.5 years respectively. Postoperative saturations increased to 92% (87%) at 7 months in the male patient and to 90% (88%) at 2 months in the female patient. Peripheral vein contrast echocardiography was positive in both patients at last follow up.
Conclusions: Simple exercise testing is feasible in young children and is a sensitive diagnostic tool to reveal significant PAVM's. Despite acceptable saturations at rest severe desaturation on exercise (<70%) can occur early or late after CPA. It should lead to further invasive investigation with bubble contrast echocardiography and pulmonary angiography. Lack of significant desaturation on exercise can be found early after completion of CPA despite positive peripheral bubble contrast echocardiography.

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Home INR monitoring for children on warfarin

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Objectives: To evaluate the accuracy, acceptability and feasibility of using home INR monitoring in children on warfarin in the setting of a regional paediatric cardiology unit. To describe our experience of home INR monitoring in twenty five children.
Methods: Six children (5–12 years) had INRs measured at home by their parents using a CoaguChek (Roche) device. Immediately following this, the INR was measured at the hospital clinic (Manchester capillary INR). After the initial 3 months INR was measured using the at home device only. The patients underwent structured interview to assess the acceptability of home INR monitoring. Following this home INR monitoring has been offered to all our children on warfarin.
Results: The difference between home and hospital INR values in the children was never sufficient to alter the advice regarding warfarin dose (coefficient of correlation R = 0.87). All six children and families expressed a strong preference for the home testing method. Twenty five children have since used or are using home INR monitoring (median age at starting 9.6 years, range 3.0–16.0 years). During a total of 95 patient years' experience with the system no child has had problems related to thrombo-embolism. One fifteen year old had significant bleeding following termination of pregnancy. Once this was controlled her INR could be rapidly stabilised at home in view of the ease of INR monitoring. There have been no other complications with excessive bleeding. Two children have stopped using the device because of compliance problems. A further infant's family preferred to continue with hospital INR monitoring until he is older.
Conclusions: At home INR monitoring is reliable and usually preferred by the child and their family. It is feasible in the context of a regional paediatric cardiology unit. It should be the recommended method of INR monitoring in the majority of children on warfarin.

P170

Spiro-ergometry for the assessment of physical capability during physical rehabilitation in patients after surgery for congenital heart disease

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Background: Prior to entering the rehabilitation program "Physical Training in Children with Congenital Heart Disease" of the German University of Sprts, spiro-ergometry was performed to assess baseline physical capability in children after correction of ventricular septal defect (VSD) and tetralogy of Fallot (TOF), and compared with normal values.
Patients and Methods: Three patient groups were studied: group 1 consisted of 14 patients (mean age 12.4 years, range 6.8 to 16.8) following surgical closure of VSD; group 2 consisted of 14 patients following repair of TOF (mean age 11.7 years, range 6.5 to 16.6), and group 3 consisted of 18 matched control patients (6.5 to 16.5 years, mean 11.8) with minor abnormalities or complaints such as atrial premature beats, mitral valve prolapse, or noncardiogenic syncope in whom exercise capacity may be expected to be unimpaired. All patients in groups 1 and 2 were in NYHA class 1, without hemodynamically important residual sequelae. Exercise testing was performed in a standardised manner on a bicycle ergometer. The load was increased every 2 minutes by 0.5 Watt/kg upto the endurance limit. Heart rate, oxygen uptake, oxygen pulse and respiratory quotient at maximal load were analysed.
Results: Maximal work was either low normal (groups 1 and 3) or subnormal (group 2) compared with published normal values: 2.0–3.0 Watts/kg (mean 2.1) in group 1 versus 2.5–3.5 Watts/kg (mean 2.6) in group 2 versus 2–4 Watts/kg (mean 2.6) in group 3. Maximal heart rate was below normal in all 3 groups: 128–208 (mean 174) in group 1 versus 140–216 (mean 171) in group 2 versus 162–204 (mean 182) in group 3. Oxygen consumption (27–70 ml/Watts/kg, mean 40 ml/Watts/kg) and oxygen pulse were

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similar in all 3 groups and did not differ from normal published values.
Conclusions: These results indicate that most children have normal physical capability following VSD closure or repair of TOF. However, they also suggest that the normal values need to be reexamined, and that published data derived from highly motivated normal subjects may not be the appropriate reference standard for comparison.

P171
Serum insulin-like growth factor-1, insulin-like growth factor binding protein-3, and growth hormone levels in children with congenital heart disease
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The influence of insulin-like growth factor (IGF) in the pathogenesis of failure to thrive, commonly seen in congenital heart disease (CHD), have recently been reported. Chronic hypoxemia could directly or indirectly reduce serum insulin-like growth factor-1 (IGF-1) concentrations and this may be a cause of the growth failure in patients with cyanotic congenital heart disease.

To aim of this study was to evaluate serum IGF-1, insulin-like growth factor binding protein-3 (IGFBP-3) and growth hormone (GH) levels in CHD patients and also evaluate relationship of these parameters with anthropometric findings, cyanosis and malnutrition.

In this study, serum IGF-1, IGFBP-3 and GH levels were studied in 94 patients (36 female, 58 male; 20% was cyanotic CHD), with congenital cardiac malformation and 76 control subjects (35 female, 41 male). Thirty-nine percent of children with CHD and 57.8% of children with cyanotic CHD has malnutrition. Serum IGF-levels were lower in CHD group than control subjects ($p < 0.001$). Serum IGFBP-3 and GH levels were higher in CHD group than control subjects ($p > 0.05$ and $p < 0.05$). Serum IGFBP-3 levels were higher in malnourished CHD group than well-nourished group ($p < 0.05$). In cyanotic CHD group, serum IGF-1 levels were lower and serum GH levels were higher than control subjects ($p < 0.01$ for both). Serum IGF-1 levels were positively correlated with height, weight, and age in both cyanotic and acyanotic CHD groups.

Recent report demonstrated that malnutrition and growth failure are important conditions associated CHD, especially cyanotic group. Our results suggested that GH-IGF axis are influenced in CHD patients, especially in the cyanotic group, but serum IGF-1 levels were also lower in acyanotic CHD group. We can not explain the decline of serum IGF-1 levels in CHD with only cyanosis, other factors may be related.

P172
Natriuretic peptide type B before and after exercise in surgically palliated patients with functionally univentricular hearts at a long-term follow-up
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Good ventricular function is mandatory for long time survival in patients with univentricular hearts. Ventricular function may deteriorate over time influenced by primary causes such as ventricular morphology and operative technique. The use of natriuretic peptide type B (BNP) as a marker of ventricular function has gained increased interest during recent years.
Aim: To study BNP levels in plasma (P) before and after exercise in surgically palliated patients with functionally univentricular hearts at a long term follow-up.
Patients and Methods: Samples of P-BNP were obtained before and after exercise test (bicycle ergometer) from 15 patients with univentricular hearts, with a median age of 18.3 years (12.5–54.2) of which 6 were operated upon by total cavopulmonary connection (TCPC) (follow-up time 10.2 years (8.3–11.8)) and 9 by classical Fontan procedure (follow-up time 18.8 years (11.1–20.3)). Cardiac index (CI) was determined by the dye-dilution technique.
Results: The P-BNP values were significantly higher in the Fontan group of patients both before 131.8 ng/L (0.5–296.4) and after 108.1 (0.1–235.9) exercise, as compared with the TCPC group 12.8 (0.5–39.1) ($p = 0.02$) (Mann-Whitney U test) and 9.7 (2.7–26.2) ($p = 0.03$) respectively. The reference interval for P-BNP was 0–18.4 ng/L. Exercise did not increase the P-BNP levels significantly in any of the groups with a difference of the P-BNP between before and after exercise of 5.9 (−23.7–31.0) in the Fontan group and −1.0 (−12.0–3.9) in the TCPC group. Workload and CI at rest and during exercise were the same in both groups 1.4 W/kg (1.0–2.1), 2.3 l/min/m^2 (1.5–2.6) and 4.7 l/min/m^2 (3.7–6.3) in the Fontan group and 1.5 W/kg (1.0–1.7), 1.9 l/min/m^2 (1.3–3.5) and 5.5 l/min/m^2 (4.2–6.8) in the TCPC group ($p = 0.6$), ($p = 0.9$) and ($p = 0.3$), respectively.
Conclusions: Patients with univentricluar hearts operated upon by the classical Fontan procedure had increased P-BNP levels as compared to those operated upon by TCPC which may be explained by older age at surgical palliation, longer follow-up time and type of surgical procedure per se. Exercise did not increase the P-BNP levels in the classical Fontan patients nor the TCPC patients which may indicate that ventricular overload is not the major cause of the decreased working capacity observed in patients with univentricular hearts.

P173
Evidence of endothelial dysfunction in patients with univentricular physiology before completion of the Fontan operation
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Objective: Postoperative thrombosis after the Fontan procedure has been widely described. Coagulation abnormalities seem to occur early in the course of patients with univentricular physiology and may precede surgery. Endothelial abnormalities due to chronic hypoxia and hyperviscosity may take place in this scenario.

The purpose of this study was to investigate if patients with a bidirectional superior cavopulmonary anastomosis have altered plasma levels of endothelial and coagulation markers.
Methods: Ten patients aged 4–19 years (median 12) were compared to six normal controls (aged 9–16 years, median 12). Plasma levels of von Willebrand factor antigen (vWF:Ag), thrombomodulin, tissue plasminogen activator (t-PA), plasminogen activator inhibitor-1 (PAI-1) and D-dimer were measured with enzyme-linked immunosorbent assay.
Results: Increased vWF:Ag ($p = 0.01$) and t-PA ($p = 0.01$) and decreased thrombomodulin ($p = 0.03$) were found in patients when compared with controls while PAI-1 levels were not different. D-dimer levels were within the normal range. T-PA levels had a positive correlation with platelet count ($r = 0.78$, $p = 0.007$), suggesting that platelets may play a role in the endothelial activation.

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Conclusions: Altered plasma levels of endothelial markers in the presence of normal D-dimer levels suggest that endothelial dysfunction may precede the occurrence of intravascular coagulation and thrombosis in patients with univentricular physiology. These observations may have therapeutical implications.

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The fate of children with microdeletion 22q11 syndrome and congenital heart defect: clinical course and cardiac outcome

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Background: Congenital heart defects (CHD), particularly conotruncal malformations are a major feature of the microdeletion 22q11 syndrome (22q11). The prevalence of CHD and the overall clinical features are well known but information about the clinical course is scarce.
Aims: To analyze the cardiac outcome of children with 22q11 and CHD during long-term follow-up.
Methods: Retrospective assessment of the clinical history of 47 children with 22q11 and CHD, the median age at diagnosis was 3 d (range 1 d–3 mo), the weight 2.9 kg (1.3–4.8 kg). CHD consisted in tetralogy of Fallot in 12, interrupted aortic arch in 10, pulmonary atresia in 10, ventricular septum defect in 8, truncus arteriosus in 6 and double aortic arch in 1 patient. Extracardiac anomalies were present in 46 of 47 children.
Results: Median follow-up time was 8.5 y (3 mo–23.5 y). Thirty-five children underwent surgical repair of CHD, age 7.5 mo (2 d–5 y). Hospitalization time was 35 d (7–204 d), ICU stay 15d (3–194 d). Significant complications were observed in 12 (34%) patients, including tracheotomy in 7 due to tracheo-bronchial malformations in 4 and prolonged ventilation in 3. Surgery for extracardiac malformations was required in 23 (49%) patients. Overall mortality was 23% (11/47), median age of 8 mo (3 d–6 y). Cause of death was cardiac in 5, withdrawal of care by multiple malformations in 4 and immunologic in 2 cases. Cardiac re-interventions were performed in 18 patients (51%), including re-operation in 12 and interventional catheterization in 10. Five patients underwent more than one re-intervention. Significant residual cardiac findings were present in 25 (71%) patients at end of follow-up.
Conclusions: Even considering the severity of CHD, the clinical course of children with 22q11 and CHD is characterized by high mortality and morbidity, both conditions aggravating each other.

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Important comorbidity due to upper airway anomalies associated with relevant congenital heart disease

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Few data exist on the clinical course and outcome in children with significant congenital heart disease (CHD) and associated clinically relevant upper airway anomalies. A retrospective study was performed covering an 8-year period ending in 2003 evaluating all patients with relevant CHD and with associated upper airway anomalies, diagnosed and assessed at least by bronchoscopy. 11 patients were identified accounting for 1.5% of a total of 764 patients requiring surgical treatment of CHD during the observation period. The spectrum of CHD was wide (VSD 3, transposition 2, Fallot 2, anomalous pulmonary venous connection 2, truncus 1, coarctation 1). Heart surgery was performed neonatally in 6 patients and by age 5 months in the remaining. Diagnosis of airway disease was made prior to heart surgery in 5 and in the postoperative course in 6 patients. In the latter patients, failure of early extubation was the leading symptom in all. Diagnosis was made by bronchoscopy in all 11 and by additional bronchography in 2 patients. Treatment of airway disease was surgical in 4 patients (including tracheostomy in 2) and conservative in the remaining. Patients with airway anomalies had complicated management of cardiac operations with duration of perioperative intubation significantly prolonged (median 24 days, range 4–34) and also prolonged perioperative hospital stay (median 72 days, range 14–147). Overall outcome was good with 10 patients being alive, one newborn was denied surgical correction of total anomalous pulmonary venous connection because of severe airway hypoplasia and died. After a median follow-up of 17 months, 4 of the survivors have respiratory symptoms, none has cardiac symptoms. 3 infants had respiratory syncitial virus infections (2 prior to heart surgery).
Conclusion: Upper airway anomalies did not lead to increased mortality in our series but perioperative management for correction of CHD was complicated by prolonged duration of intubation and intensive care stay.

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The evaluation of cardiac effects of perinatal asphyxia by biochemically

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The effects of asphyxia on heart are transient myocardial ischemic effects. Fetal and neonatal myocardium is quite resistant to the hypoxemia and most of the effected myocardium result in cure spontaneously. We investigated the effect of the asphyxia on perinatal heart by biochemically.

Forty-five term hypoxic infants were enrolled for the study group and control group. Study group was composed of 3 subgroups: Group I – grade I asphyxia (n = 15), Group II – grade II asphyxia (n = 15) and Group III – grade III asphyxia (n = 15). The biochemical examinations were done in all infants in the first hour: AST; ALT, ALT, CK, CKMB, LDH, and troponin-T. In asphytic infants were repeated on the 3rd day, at the end of the first week and 15th day.

In group I infants the biochemical parameters were decreased from the first day to 15th day, but the decrease was more significant from the 3rd day to 15th day. The troponin-T levels remained under 0.1 ng/dl; the first day mean value was 0.07 ± 0.03 ng/dl and the 15th day mean value was 0.02 ± 0.01 ng/dl. In group II the parameters other than LDH were decreased from the first day to 15th day like group I and the decrease was more significant from the 3rd day to 15th day. In first 3 days the LDH levels increased, and reached to peak, and after the third day the LDH levels decreased significantly. The mean troponin-T levels on first and 3rd day was higher and the 7th and 15th day mean values were under 0.1 ng/dl. In group III infants like group II, the first 3 days the LDH levels increased, and reached to peak, and after the third day the LDH levels decreased significantly. The mean values of troponin-T on first, 3rd and 7th days were higher and on 15th day the mean value was under 0.1 ng/dl. Other parameters on first,

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3rd and 7th days were higher than the other asphytic groups and there was a significant decrease from first day to 15th day. The increase in LDH and troponin-T levels was significant on 7th day, when compared with other parameters. The asphyxia related cardiac changes were more significant in grade III asphytic infants and these changes were reversible.

With the respect of our study, in the evaluation of the degree of asphyxia related cardiac changes, the troponin-T level may be a more reliable test.

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Hyperhomocysteinemia in patients with congenital cardiac malformations: relationship with vitamin B12, folic acid levels and cyanosis

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Hyperhomoccysteinemia is defined as an independent new risk factor for ischemic heart disease, stroke, atherosclerosis and venous thrombosis.

We determined plasma total homocysteine (tHcy) levels by ELISA method in 142 children which 37 of them were cyanotic, with isolated congenital cardiac malformations and 71 healthy subjects. Hyperhomocysteinemia was defined if tHcy levels was above 85% value of the controls (13.7 mg/L). While, median tHcy levels were not different between patient group (11.3 mg/L) and control (9.95 mg/L) ($p > 0.05$), hyperhomocysteinemia frequency was higher in patient group (40.7%) ($p < 0.001$). Vitamin B12 and folic acid levels were lower in patients than controls ($p < 0.05$, $p < 0.001$, respectively). In patients, these vitamin levels were negatively correlated with tHcy levels ($r = -0.2$, $p < 0.05$, for both). No correlations were found between tHcy levels and body weight, body mass index, BUN, creatinine, T3, T4 , TSH levels ($p > 0.05$). Patients with cyanosis had higher tHcy levels (median: 18.7 mg/L) than acyanotic group and control subjects ($p < 0.001$ for both). Hyperhomocysteinemia frequency was 78% in cyanotic group. Vitamin B12 and folic acid levels were not different between patients with or without cyanosis ($p > 0.05$). In conclusion, hyperhomocysteinemia is found more frequently in children with isolated congenital cardiac malformations. It is related with vitamin B12 and folic acid levels indicating that these vitamins's supplement would be useful to prevent hyperhomocysteinemia in these patients. On the other hand, it is not clear why plasma tHcy levels are higher in patients with cyanosis.

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Anomalous origin of the left coronary artery from pulmonary artery

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This study examines the clinical experience of one centre with anomalous origin of the left coronary artery from the pulmonary artery.

Twenty four consecutive patients were referred for anomalous origin of the left coronary artery. Tachypnea was observed in 13 patients, feeding difficulties in 12 patients and severe congestive heart failure in 3. The electrocardiogram was characteristic in all showing at least a lateral infarction pattern. Each had moderate cardiomegaly on chest radiographs. In the most recent patients, diagnosis was confirmed by echocardiography ($n = 12$) showing the abnormal origin of the left coronary artery with typical retrograde flow into the pulmonary trunk on colour flow velocity mapping. This later could replace the need for cardiac catheterization angiography.

Three patients died before surgery. The remaining patients underwent surgical correction at the age of 46 ± 51 months, median 33 months. Direct reimplantation of the left coronary artery in the aorta was realised in 13 patients, 5 patients had coronary artery bypass with interposition of saphenous vein graft ($n = 3$) or subclavian artery graft ($n = 2$), and 3 patients had intrapulmonary tunnel of the left main coronary artery to the aorta (Takeuchi procedure). Associated repair of mitral valve was performed in 5 patients and one had resection of apical aneurysm. Two patients died after surgery.

Follow-up was available in all but 4 and included clinical examination, exercise stress testing, myocardial scintigraphy and cardiac catheterization. On control echocardiography, the shortening fraction improved in all patients. Two patients had occlusion of the proximal left coronary artery requiring mammary artery bypass graft and another one had repair of mitral valve.

To conclude, anomalous origin of the left coronary artery from the pulmonary artery is rare and diagnosis can be performed by echocardiography without the need of cardiac catheterization. Direct reimplantation of the left coronary artery to the aorta is the procedure of choice. Long-term follow-up is required because of sequelae of myocardial infarction and risk of coronary artery occlusion.

P179

Patient and family awareness of endocarditis risk associated with body art, including tattooing and body piercing. A questionnaire on a cohort of children with congenital heart disease

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Introduction: Body art in the form of tattoos and piercings have become increasingly popular with youngsters and are, nowadays, more socially acceptable. The risks associated with body art have been recently highlighted in the media with tissue destruction and liver damage topping the list. Two deaths were also reported in Europe alone but no specific attention has been drawn to the potential risk of endocarditis in susceptible individuals.
Method: We decided to investigate the awareness of patients with congenital heart disease and their families to the potential link between body art and endocarditis. We supplied 500 questionaires to clinic patients who were potentially at risk from endocarditis asking whether they had any piercings or tattoos, whether they had any intention to have any in the future and whether they thought there could be a link between body art and endocarditis. The underlying cardiac conditions were divided into the following groups: (1) Cyanotic (2) Shunt (3) Obstructive lesion (4) Regurgitant lesion (5) Prosthesis and some had a combination of these.
Results: Of the 500, there were 350 returns and of these, 42 declined to reply. Of those who replied 150 were female and 158 male and the average age was 8.12 years. Eighty six percent were filled by a parent and the rest by the patient. The ethnicity was identified in 267 returns (optional). Seventy eight admitted to having piercings and 1 had a tattoo. The main underlying conditions which this cohort of patients sufferred from consisted of cyanotic 96, shunt 78, obstructive lesion 76, regurgitant lesion 20 and prosthesis 11; the remaining had a combination of these.
Conclusion: Many were bemused about the questionaire. Few were aware of the potential risk of infection from body art let alone the

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possible link with endocarditis. Better knowledge is required about the link between body art and endocarditis in order to provide guidelines for doctors and patients. This also has public health implications as body art parlours are licenced by city councils

P180

The relation between serum digitalis-like immunoreactive substance (DLIS) levels and cardiac performance in the transient tacyphnea of the newborn infants

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Objectives: Transient tacyphnea of the newborn (TTN) is the most common reason for respiratory distress in newborn infants. Digitalis-like immunoreactive substances (DLIS), a kind of adrenocortical hormone, are biological inhibitor of Na/K ATPase. In this study, we have studied the relationship between cardiac performance, volume overload and DLIS levels in TTN cases.
Methods: The study group was composed of 12 TTN cases (8 male, 4 female) and compared with 12 healthy newborn (7 male, 5 female) as a control group. We have studied DLIS levels, FeNa (fractional Na excretion), serum Na, serum osmolarity, blood pressure and left atrial diameter (LAD), left ventricular end-diastolic diameter (LVEDD) with echocardiography for both groups, in the first and seventh days.
Results: The mean gestational age and birth weight of study group and control group were 37.4 ± 0.0 weeks and 36.8 ± 0.9 weeks, 2867 ± 191 gr and 2848 ± 237 gr respectively. In first and seventh day, DLIS levels and serum Na were found higher in TTN cases than the control group. The first and seventh day DLIS levels were 0.57 ± 0.36 ng/ml and 0.27 ± 0.23 ng/ml in TTN cases ($p < 0.01$) and 0.23 ± 0.22 ng/ml and 0.06 ± 0.05 ng/ml in healthy newborn ($p < 0.001$). We have seen a correlation between DLIS levels and LAD, LVEDD, FeNa in first day and, LVEDD and FeNa in the seventh day for both groups. LAD and LVEDD (10.53 ± 1.34, 14.66 ± 1.46) were found higher in TTN group than the control group (10.3 ± 1.05, 12.33 ± 0.83) in the first day, but in seventh day no statistical difference was found between these measurements.
Conclusions: DLIS, serum Na level, LAD and LVEDD were found to be higher in TTN cases. In TTN cases this result indicates volume overload and cardiac effect in TTN. High DLIS levels may be a result of volume overload and increased serum Na level. These findings are thought to be important to explain the pathophysiology of TTN.

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Primary heart tumors in the pediatric age group: a 16-year review

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Primary heart tumors in the pediatric age group are extremely rare. The rhabdomyosarcoma, fibroma and myxoma are the most common. Their clinical manifestations vary widely from asymptomatic presentations to life-threatening cardiac events. As the clinical picture of cardiac tumors is varied they cause many diagnostic problems to primary medical care.

The aim of this research was to analyze the clinical course and the treatment in newborns, infants and children with heart tumors hospitalized in the Department of Pediatric Cardiology of Poznan Medical University between 1987–2003. The study group consisted of 10 newborns, 9 infants and 4 children (12 males and 11 females). The age on the admission ranged from 1 day to 16.5 years.

The majority of the newborns was referred to the Clinic with the suspicion of congenital heart disease based on cyanosis, tachycardia and cardiomegaly in chest x-ray, in 3 patients the heart tumor was diagnosed prenataly. In the group of infants only 1 was referred with the suspicion of heart tumor, the others were referred with the suspicion of pneumonia or myocarditis. In the group of children all were referred with arrhythmias. The clinical examination revealed the symptoms of cardiac failure including tachycardia and tachypnoe in 7 cases, in all the cases the heart murmur was audible. In 15 cases arrhythmia was present, including supraventricular tachycardia treated pharmacologically in 3 newborns. The abnormalities in ECG and Holter monitoring examination varied in 5 patients conduction abnormalities including atrioventricular block first-second degree occurred, in 2 patients sinus bradycardia, in 13 patients presence of ventricular and supraventricular extrasystolic beats, in 3 patients supraventricular tachycardia were diagnosed.

The diagnosis of the heart tumor was established after performed echocardiography. Tumors were located most frequently in the right ventricle (14) and left ventricle (6) with multiple tumors being present in 11 cases.

The surgical treatment was performed in 4 newborns, 3 infants and 2 children. The histological examination revealed: rhabdomyosarcoma (3), rhabdomyoma (3), myxoma (2) and endothelioma (1). 4 patients died, including 1 newborn before the surgery. The rest of the study group reminds in the care of cardiologic Outpatient Clinic. In 5 patients spontaneous regression was observed.

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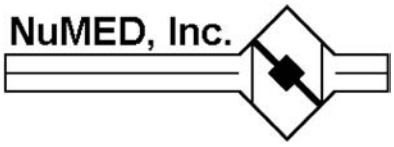

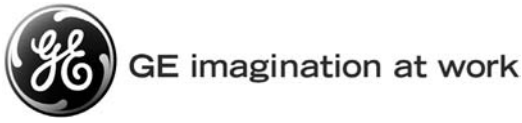

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