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Cardiology in the Young

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Session 1: Fetal

01

Recurrence of congenital heart disease in fetuses with familial risk, referred for fetal echocardiography

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Objectives of the Study: To evaluate the recurrence of congenital heart disease (CHD) in a population of pregnant women with familial risk referred for fetal echocardiography.
Material and Methods: Retrospective/prospective study of the data of 1131 pregnant women with familial risk for CHD studied in our Center between January 1990–December 2001 by fetal echocardiography. Twelve pregnant women were followed-up in two consecutive pregnancies. 1033 cases had single familial risk, 98 cases multiple risk (78 – double risk, 19 had 3 and 1 had 4 relatives affected). 612 cases had one previous child with CHD; 24 – 2 previous children; mother was affected in 163 cases (with another relative in 15); father was affected in 82 cases (with another relative in 14); the 2–3 degree relatives were affected in 298 cases, 45 with multiple risk. Fetal diagnosis was compared with postnatal findings at follow-up (check-up point 6 months, in order to detect minor CHD found postnatally).
Results: CHD was found in 47/1131 infants (4.15%): in 43/1033 cases with single familial risk (4.16%) and in 4/98 (4.08%) of cases with multiple risk. The recurrence of CHD in the 1st degree relatives was 39/886 (4.4%), 4.08% (25/612) when one previous child was affected, 13.3% (2/15) with 2 previous children; 3.9% (6/153) with mother alone affected, 1/8 (12.5%) with mother and another relative affected; 3/68 (4.4%) with father alone affected and 1/3 with father and another relative affected; 4/298 (1.3%) with distant relatives affected. Specific recurrence of a similar type of pathology occurred in index cases with hypoplastic left heart syndrome, tricuspid and pulmonary atresia in situs viscerum inversus, atrioventricular defect, ventricular septal defect, TF, ASD II (rate 3– 6%) and it was higher in HCMP (global recurrence 6/33 = 18.2% and specific recurrence 5/33 = 15%).
Conclusions: Recurrence rate of CHD in our population is higher than in previously published fetal data; we confirm the specific transmission of a similar CHD in some families to indicate genetic predisposition.

02

Spectrum and one-year outcome of prenatally diagnosed cardiac malformations

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Objective: Definition of the spectrum of prenatally diagnosed structural heart defects, comparison with the postnatal diagnosis and description of one-year-outcome.
Background: The reliability of prenatal diagnosis, the spectrum of malformations and the perinatal approach all influence mortality and morbidity of neonates with structural heart defects.
Methods: Retrospective analysis of 111 fetuses with cardiac malformations prenatally diagnosed between 1998 and 2002. Comparison of the diagnosis established by fetal echocardiography with the postnatal "golden standards" 2-d echo, intraoperative or post-mortem finding. Follow-up of all available patients for at least one year.
Results: At the first fetal echocardiography, mean gestational age was 27 weeks (SD 6 weeks). The most frequent diagnose were septal defects, closely followed by right and left heart obstructions and complex cardiac lesions. In 18 cases, pregnancy was terminated, in 7 cases intrauterine death occurred, the remaining 93 pregnancies were continued, in a multidisciplinary fashion. All duct-dependent lesions were provided with i.v. prostaglandins immediately after birth; in all cases neonatal and pediatric-cardiology service was present. Of all 111 cardiac lesions, 91 had been correctly diagnosed in every detail, in 7 cases small concomitant lesions had been missed, in 2 cases prenatal diagnosis was not correct, in 11 cases complete follow-up was not possible. One year survival rate of the 86 neonates was 87%, the best in septal lesions (96%), the worst in left heart obstruction (29%); the numbers were 94% for right heart obstruction and for 70% complex lesions, respectively.
Conclusions: Close multidisciplinary approach has improved one-year-outcome of infants with prenatally diagnosed structural heart defects. Unfavourable courses are still present in the setting

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of simultaneous extracardiac or complex cardiac lesions and in situations where parents deny operative options, like in hypoplastic left heart.

03
Prenatal study of fetal endocardial echogenicity and its association with maternal toxoplasmosis

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The clinical significance of the intracardiac echogenic foci, or "golf balls", are still controversial. They are considered by many authors as a normal variant, and by others as possible markers for chromosomal abnormalities, or else for cardiac diseases. No possible correlation between these findings and fetal cardiac commitment by Toxoplasma gondii is related.
Objective: Compare a group of fetuses with acute or recent maternal toxoplasmosis and one group without maternal disease, evaluating the presence of fetal endocardial echogenicity
Methods: Ninety-one fetuses whose mothers had a clinical or laboratorial diagnosis of acute or recent toxoplasmosis, detected by soroconversion or by increased titers of IgM and IgG confirmed by the capture test, were compared to a control group of 182 fetuses, selected from a low-risk population participating in a program of prenatal screening for congenital heart disease.
Results: No statistical difference was observed in maternal and gestational ages between both groups. Areas of endocardial hyperechogenicity were observed in 69 fetuses with maternal toxoplasmosis (75.8%) and in only 6 fetuses of the control group (3.3%) ($p < 0.001$, chi-square test). In 52 cases of the study group (75.4%) the endocardial echogenicity was diffuse and in 17 (24.3%) it was focal. In the control group, a focal distribution was observed in 5 fetuses (83.3%).
Conclusion: The prenatal echocardiographic image of focal or diffuse endocardial hyperechogenicity is more prevalent in pregnancies with maternal toxoplasmosis than in normal pregnancies and there is association between the presence of fetal endocardial hyperechogenicity and maternal toxoplasmosis.

04
Relationship of maternal autoimmune response to clinical manifestations in children with congenital complete heart block

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The aims of the study were to study the autoimmune response in mothers with children with congenital heart block (CHB) with different clinical manifestations. Clinical data and sera for the determination of immunological tests were available from 104 mothers of 113 children diagnosed with CHB before the age of 16 years. Prenatal diagnosis was performed in 74 (65%) children of 65 mothers and 39 (35%) had postnatal diagnosis of CHB. Maternal antibodies to 52kd and 60kd SS-A, and to 48kd SS-B were determined by time resolved fluoroimmunoassay (TR-FIA) and to antinuclear antibodies (ANA) by immunoflurescense (IF). Out of the 65 mothers of children with in utero diagnosed CHB, 88% had antibodies to 52kd SS-A and 83% had ANA. Antibodies to 60kd SS-A and 48 kd SS-B were less frequently present, in 48% and in 54% of the mothers, respectively. Seven (11%) of the mothers were negative by all immunoassays. Of the 13 mothers of children with in infancy diagnosed CHB one mother had high-titer ANA. After one year of age CHB was diagnosed in 26 children; at 1 to 6 years in 16 and after 7 years in 10 children. The prevalence of antibody positivities were 6% and 10%, respectively. In all twin pregnancies ($n = 4$) and in all families with recurring cases with CHB ($n = 5$) maternal antibodies were positive at least in one assay. The titer of 48 kd anti-SS-B-antibodies was significantly higher in children with cutaneous neonatal lupus (98.1 vs. 41.0; $p = 0.02$). All mothers, whose children died before the age of 4 years ($n = 8$), and 85% (11/13) of mothers whose children developed cardiomyopathy had elevated antibody titers at least in one assay. However, we could not find any prognostic value of maternal antibody levels or specificities on the clinical outcome of the children with CHB. Although rare, late detection or postnatal progression of CHB in antibody-mediated CHB should be taken into consideration. Maternal antibody levels or specificities have prognostic effect neither on the clinical outcome of the child with CHB nor on the risk of reappearance in the same family.

05
Prenatally diagnosed complete atrio-ventricular block with and without structural heart disease in the 1990s: management and outcome

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Complete AV block (CAVB) is associated with significant perinatal morbidity and mortality. The emergence of transplacental treatment modalities (steroids; beta-adrenergic agents) and progress in neonatal care may have improved the prognosis of affected fetuses in recent years. Reviewing the experience of 2 large fetal cardiac programs for the years 1990–2000, we sought to determine the prenatal management and outcome of prenatally diagnosed CAVB.
Results: Fifty fetuses were detected at 24 ± 5 weeks gestation. CAVB associated with structural heart disease was diagnosed in 22 (44%): 15 had left atrial isomerism (LAI) and 3 congenitally corrected transposition (l-TGA). At diagnosis, 10/22 (45%) fetuses had junctional or ventricular escape rates of <55 beats/minute, 5 of whom were in hydrops. Beta-adrenergic drug therapy was initiated in only 2 (9%): both had LAI and died as neonates. Overall, 19/22 (86%) with structural heart disease, died either in utero (LAI: 11/15) or as neonates (LAI: 3/4; l-TGA: 1/3). Isolated CAVB was diagnosed in 28 (56%) fetuses: usually (92%) this was associated with the presence of maternal anti-Ro/anti-La autoantibodies. Thirteen of the 28 (46%) had junctional or ventricular escape rates <55 beats/minute, but only 1 (4%) was hydropic at presentation. Compared to CAVB with structural heart disease, transplacental drug treatment was attempted more commonly and more successful. In cases with heart rates <55 beats/minute, the fetal and neonatal survival was better with maternal beta-adrenergic treatment when compared to untreated pregnancies (6/7 versus 1/6; $P < 0.05$). Maternal dexamethasone was added to 17 of the 24 (71%) pregnancies with immune-mediated CAVB: only 3 (18%) did not survive the neonatal period, while 4 of 7 cases (44%) without steroid administration had died ($P < 0.02$).
Conclusions: CAVB associated with structural heart disease continues to be associated with poor fetal and postnatal outcome. In this non-randomized study, transplacental anti-inflammatory treatment with fluorinated steroids, combined with beta-sympathomimetics at fetal heart rates <55 beats/minute appears to improve the survival of CAVB and structurally normal hearts.

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06

Treatment of fetal tachycardia with sotalol: transplacental pharmacokinetics and pharmacodynamics

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Background: Maternally administered, intra uterine therapy of fetal tachycardia is dependent on the transplacental passage of the antiarrhythmic agent. In this study, the pharmacokinetics and -dynamics of sotalol and digoxin are therefore investigated.
Methods: A prospective study of patients treated for fetal tachycardia with sotalol and/or digoxin. Concentrations of sotalol and/or digoxin were determined in maternal, umbilical and neonatal blood and in amniotic fluid and the relationship between these concentrations and the occurrence of conversion to sinus rhythm, was investigated. Results Nineteen patients were studied, 10 with atrial flutter and 9 with supraventricular tachycardia. Fourteen were treated with sotalol; 13 converted to sinus rhythm, of whom 2 relapsed. There was one intra uterine death. Four patients were treated with sotalol and digoxin of whom 2 successfully. One patient was unsuccessfully treated with digoxin and flecainide. Mean birth weight was 3306 gram. The daily maternal sotalol dose was linearly related to the maternal plasma concentration. The mean fetal/maternal sotalol plasma concentration (F/M ratio) was 1,1 and the mean amniotic fluid/fetal blood ratio of sotalol was 3,2. The effectiveness of sotalol therapy could not be extrapolated from maternal blood levels. The sotalol T1/2 in neonates varied from 5,9 to 48 hours. Digoxin passes the placenta partially with a mean F/M ratio of 0,53.
Conclusions: Sotalol is a potent anti-arrhythmic agent in the treatment of fetal tachycardia. The placental transfer is excellent with an approximately 1:1 F/M ratio at the time of delivery. It accumulates in amniotic fluid but not in the fetus itself. Therefore it seems that renal excretion in the fetus is efficient and greater than the oral absorption by fetal swallowing. The maternal blood level is not a reliable predictor of the chances of success of therapy. Sotalol is not associated with fetal growth restriction. Pharmacokinetics in the neonate show that the renal excretion in the neonate is adequate and comparable to that in adults.

07

Congenital heart disease and outcome in Turner's syndrome diagnosed prenatally

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Overall outcome and the incidence and type of heart disease were investigated in fetuses diagnosed with Turner's syndrome in early fetal life. Seventy-eight cases of Turner's syndrome were identified after karyotyping. Of these, 61 were examined echocardiographically by abdominal ultrasound, which was performed before a karyotype result was known in 59. Fifty-four (88.5%) cases were referred because of increased nuchal translucency thickness (NT) of >4mm, six cases because of fetal hydrops and one case because of the karyotype result itself. Seventy-three of the total group of 78 (93.5%) fetuses had a 45XO karyotype and the other 5 were mosaics. Diagnostic cardiac images were obtained in 54 of the 61 fetuses at initial echocardiography, which was performed prior to 14 weeks gestation in 47/54. A cardiac abnormality was detected in 33/54 fetuses (61.1%). The most common diagnosis was coarctation of the aorta in 24/54 (44.4%), followed by the hypoplastic left heart syndrome in 7/54 (13%). The mean NT was significantly higher in fetuses with a heart defect than in those with normal echocardiography. Termination of pregnancy occurred in 61/78 (78.2%) and intrauterine death in 15 (88%) of the 17 continuing pregnancies. Both survivors had normal hearts and mosaic karyotypes, the abnormality being confined to placenta in one of them. There was a higher incidence of heart defects in Turner's syndrome identified prenatally when compared to postnatal reports (61% vs 20–30%) and the defects tended to be more severe. The commonest heart defects detected prenatally were hypoplastic left heart syndrome and coarctation of the aorta, in contrast to postnatal life where a bicuspid aortic valve is the commonest diagnosis. Of 15 continuing pregnancies with an 45XO karyotype, none survived intrauterine life. Turner's syndrome, as it usually presents to a fetal referral centre, with increased nuchal fluid or hydrops and with a typical XO karyotype, has an extremely poor chance of intrauterine survival.

08

Chromosome aberrations in children with congenital heart defects. Analysis of microscopic and submicroscopic abnormalities

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Congenital heart malformations are heterogeneous in their origin. Originally 85% were classified as multifactorial, 5% as exogeneous and only 10% as genetic dispositions. The majority of genetic defects was contributed to well defined syndromes as trisomy 21. But by improving the applied techniques it became obvious that the percentage of genetic disorders in children with congenital heart disease is much higher. Our investigations comprise 365 children with different types of heart malformations, excluding the probands showing a defined syndrome such as trisomy 21, 13 or 18. Conventional chromosome investigations showed aberrations in only 9%. Specific microdeletion analysis having regard to the phenotype of the patients or their structural heart anomalities revealed aberrations in 23% in those with a normal appearing karyotype after conventional investigation. Excluding the patients with pathologic findings in conventional and microdeletion analysis, 25 of the remaining group were investigated by subtelomeric screening. Of these three patients (8%) with different abnormalities were detected [del(1q), del(14q), der(8)]. Molecular analysis to characterize the aberrations are in progress. This systematic approach in three steps, developed in Bonn, seems to have a high efficiency in detecting an underlying genetic disorder in congenital heart disease. (Supported by the Richard-Winter-Stiftung)

09

Foetal and neonatal cardiomyopathies: a series of 81 patients

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Objective: To describe the spectrum of phenotypes, the aetiologies and the prognosis of cardiomyopathies (CMP) diagnosed during f3 mitochondrial respiratory chain defects fœtal life or within the first month of life.
Population: Retrospective analysis of the charts of 42 fœtal and 39 neonatal CMP diagnosed over a period of 7 years. CMP associated

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with a heart defect, systemic hypertension, maternal diabetes mellitus, twin-twin transfusion syndrome or tachyarrhythmias were excluded. Patients were screened for inherited metabolic diseases, known genetic syndrome and for family history of CMP (ECG and echocardiography of siblings and parents).
Phenotypes: 38 (47%) had dilated CMP, 35 (43%) hypertrophic CMP and 8 (10%) others [3 right ventricular dysplasias, 5 non compact myocardium].
Aetiologies: A cause to the CMP was found in 57% of the cases. For hypertrophic CMP (known cause in 57%): 7 dominantly inherited cases (20%), 6 Noonan syndromes (17.5%), 3 mitochondrial respiratory chain defects (9%), 1 Pompe's disease, 1 Williams syndrome, 2 polymalformative syndromes. For dilated CMP (knwon cause in 34%): 7 myocarditis (19%), 4 mitochondrial respiratory chain defects (11%), and 2 dominantly inherited cases.
Prognosis: 7 pregnancies were terminated. Overall mortality was 46% after a median follow-up of 3.6 years. There is no significant difference of mortality between hypertrophic and dilated CMP. All patients with right ventricular dysplasia and 3/5 of the non compact myocardium patients died. At one year of age, 37.5% of the survivors have a normal left ventricular function on echocardiography (20% of hypertrophic and 58% of dilated CMP).
Conclusion: The spectrum of aetiologies for CMP in foetuses and new-borns is comparable to the one of older infants. A large proportion remains of unknown cause. Mortality is high in the first weeks of life but improvement of left ventricular function is not rare. These issues make the prenatal counselling and decision making with regards to termination of pregnancy difficult to the physician as well as the parents.

Session 2: GUCH

10

The changing practice of GUCH surgery 1990–2002

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Objectives: The improved outcomes of paediatric cardiac surgery and the impact of interventional cardiology have created a changing population of adults with congenital heart disease requiring surgery. We have undertaken a review of GUCH surgery in a single centre for the past 12 years.
Methods: We reviewed the records of patients over 16 years of age undergoing surgery between January 1990 and June 2002 in a dedicated GUCH unit. Patients with ASDs were included, but not those with Marfan's Syndrome or undergoing a first procedure for bicuspid aortic valves.
Results: During this time period 434 operations were performed, with overall mortality of 7% (N = 31). The median age was 25 years (interquartile range 19 to 38). There were 166 redo operations (38%). When results were analysed according to era of operation the mean age of patients has decreased from 31.7 years in 1990–92 to 26.9 years in 1999–2002 ($p = 0.02$). Using the concensus classification the percentage of simple procedures has reduced from 41% in 1990–92 to 19% in 1999–2002 ($p = 0.002$), which is predominantly due to a decrease in the number of Secundum ASDs referred for surgical closure. The greatest area of increased practice has been the number of Pulmonary Valve Replacements (PVR) in the population of ageing Fallot (from 1% of cases initially to 9% of cases in 1999–2002). This is likely to continue with a further 140 patients likely to require PVR. Similarly the number of conduit changes in previously corrected Pulmonary Atresia and Truncus has increased from 6% in 1990 to 13% in 2002. However, Secundum ASD closures decreased from n = 25 in 1990–92 to n = 10 in 1999–2002. The mean hospital stay for the adult patients was 6 days (range 5–8) with median ITU days of 1 day. The estimated hospital cost (not including salaries/prosthetics) incurred by GUCH surgical patient was €3,435 compared to €3,961 for a CABG patient.
Conclusion: Despite the impact of interventional cardiology, the total number of surgical procedures remains unchanged. The complexity of the cases has increased particularly with redo surgery. Nevertheless, inpatient costs remain comparable to surgery for acquired disease.

11

Plamsa BNP level and chronic heart failure syndrome in adults with congenital heart disease

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Background: It is well established that the degree of B-Type Natriuretic Peptide activation (BNP) in CHF relates to functional capacity, the degree of left ventricular (LV) dysfunction and mortality. The role and value of this peptide in patients with chronic right ventricular (RV) dysfunction is not well investigated. The purpose of this study was to prove the relation between BNP and cardiopulmonary exercise capacity (VO_2 max) in asymptomatic adults with repaired tetralogy of Fallot (TOF).
Methods: NT-Pro-BNP was determined (Elecsys 2010, Fa. Roche) from n = 40 adults with TOF 22 ± 4 years after surgical repair (female n = 20; Age 27 ± 7 y; male n = 20; Age 30 ± 8 y). VO_2 max was performed by spiroergo-meter. All patients were in NYHA class I or II. All had RV-residual defects (mild pulmonary stenosis and mild to moderate Pulmonary regurgitation). LV showed in 2D-Echocardiogram normal size and function. NT-Pro-BNP was determined from 100 sex- and age-matched healthy control subjects. The normal range for VO_2 max was obtained from literature.
Results: Taken as a whole, TOF patients had significantly higher BNP level as the control subjects (TOF female 161 ± 75; TOF male 133 ± 110, control subjects 38 ± 22 pg/ml). The VO_2 max was significantly reduced in TOF patients (mean 23 ml/kg/min vs. 35 ml/kg/min). There was negative correlation between VO_2 max and NT-Pro-BNP ($r = -0.75$; $p < 0.01$).
Conclusion: The BNP activation in asymptomatic adult patients after surgical repair of TOF is elevated and the VO_2 max is impaired, so that the BNP is a surrogate parameter to estimate the cardiopulmonary capacity in patients with right ventricular dysfunction.

12

Cyanotic congenital cardiac disease in adults: new findings on hematologic and serologic variables

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Background: The diagnostic value of hematologic and serologic variables of adults with cyanotic cardiac disease in respect of iron deficiency remains unclear. The objective of this study was therefore to assess hematologic and serologic variables in respect of iron deficiency.

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Patients and Methods: 52 consecutive cyanotic patients with a right-to-left-shunt were included (median: 31 years). 25 had an Eisenmenger reaction. 34 patients had were s/p cardiac surgey, 18 patients were unoperated.
Results: Mean hemoglobin was 20.1 ± 2.7 g/dl, hematocrit 60.9 ± 8.4%, oxygen saturation 80.6 ± 7.7%, mean corpuscular volume (MCV) 91 ± 7.5 fl, and mean cell hemoglobin (MCH) 30 ± 3.1 pg. Latent or manifest iron deficiency was present in 24 patients (46.2%). Despite iron deficiency an expected hypochromasia was observed in only 11 patients, and microcythemia in five patients. Serum homocysteine ranged from 5.8 to 47.4 µmol/L (median 12.8 µmol/l). Hyperhomocysteinemia (13 µmol/L) was present in 27patients (51.9%). Extreme low values were found for serum folic acid in one patient, for erythrocyte folic acid in three patients, and for vitamin B12 in one patient.
Conclusions: In the current study 46% of adults with cyanotic CCD had iron deficiency. However, a large number of patients showed normo-/macrocytosis or normo-/hyperchromasia. Hyperhomocysteinemia was found in almost half of all patients. Therefore, normo-/macrocytosis or normo-/hyperchromasia in a cyanotic patient with iron deficiency may depict a combined effect of iron deficiency and folic acid or B-complex vitamin deficiency states. Folate and/or vitamin B deficiency, however, can be postulated but not be proven as long as no reliable reference ranges are available for these vitamins. Additionally, it is unclear, whether hyperhomocysteinemia is a risk factor for cardiovascular disease or an epiphenomenon in adult cyanotic CCD and whether cyanotic patients may benefit from pharmacologic reduction of homocysteine concentrations.

13
Carotid and femoral intima-media thickness in adult patients succesfully operated for aortic coarctation

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Objectives: Structural changes and abnormal vascular reactivity persist in patients with aortic coarctation despite successful repair. This may be due to previous increased blood pressure in the pre-coarctatial arterial conduits and contributes to the increased late morbidity and mortality in post-coarctectomy patients. B-mode ultrasound imaging can describe status and changes in intima-media complex thickness (IMCT) of carotid and femoral arterial walls. Carotid and femoral IMTC was investigated in normotensive adult post-coarctectomy patients.
Methods: Carotid and femoral IMTC was measured in 26 normotensive (31.7(SD9.1) yrs) successfully operated post-coarctectomy patients. Normotension was defined as a daytime mean systolic blood pressure under 140 mmHg as registered on 24 hour ambulatory blood pressure monitoring. Patients were compared to 26 age and sex matched controls (33.9(9.9) years). A subject's IMTC was defined as the averaged measurements of three right, three left carotid, and two left and two right femoral arterial wall segments. In comparisons of the carotid and femoral arterial IMTC's between patients and controls, the averaged per subject IMTC measurements of each of the arterial conduits were used. Comparisons were done with unpaired Student t-tests.
Results: Overall IMTC of the normotensive post-coarctectomy patients (0.65(0.17) mm) was increased if compared to controls (IMTC = 0.59(0.11) mm): DIMT = 0.06 mm, p = 0.002. Carotid IMTC was 0.70(0.10) mm in patients and 0.60(0.09) mm in controls: DIMT = 0.14 mm, p < 0.0001; however the femoral IMTC's were similar: (0.56(0.10) mm in patients and 0.57(0.07) mm in controls: DIMT = 0.01 mm, p = 0.64.
Conclusions: Normotensive, post-coarctectomy patients presented with increased carotid IMTC, where the femoral IMTC is similar to controls. It may be concluded that the vascular damage in post-coarctectomy patients is more apparent in the vascular bed proximal of the former aortic coarctation. This finding reflects the increased risk for cardiovascular and cerebral events in these patients. Moreover, the results illustrate the need to investigate multiple vascular beds according to a standardized protocol in order to describe mechanism of disease and improve disease prevention in those at vascular risk.

14
Spontaneous decrease in severity occurs frequently in pulmonary but not in aortic valvar stenosis

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The aim of the study was, to compare the time course of spontaneous changes in disease severity in isolated aortic (AS) and pulmonary valvar stenosis (PS). Patients with isolated AS or PS (excluding neonatal critical stenosis) followed in our institution between 1990 and 2000 and who had serial echocardiographic evaluation of transvalvar gradients were included. Children with gradients > 16 mmHg were grouped according to their age at presentation and initial severity of stenosis. Decrease or increase of severity of stenosis was defined as change in gradient >10 mmHg between 2 exams.
Results: 102 AS patients and 74 PS patients were followed for a mean period of 7.8 years. PS was much more frequently diagnosed in infancy (58% of patients) than AS (22%; p < 0.01). The proportion of patients who showed progression of the disease during follow-up was equal (47% in AS and 48% in PS). Over the total follow-up only 4% of AS patients had a decrease in severity of stenosis compared to 46% of PS patients (p < 0.01). The transvalvar gradients did not change during follow-up in 49% of AS patients compared to only 6% of PS patients (p < 0.01). The most rapid changes in severity were observed in the first year of life, where 67% of instances of PS regression occurred but also 82% of rapid increases (>20 mmHg/year) of PS. In contrast to PS patients, rapid progression in stenosis gradients occurred through all age groups in AS patients. A slightly larger proportion of PS patients required intervention in follow-up (22/74 or 30%) as compared to AS patients (22/102 or 22%; p = ns). It is concluded that a decrease in disease severity is frequent in PS but not in AS and that in PS but not in AS the most dramatic changes in the degree of stenosis occurred in the first year of life.

15
Bicuspid Aortic Valve (BAV): median and long term follow up

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Aim of the Study: Median and long term f/up of subjects with BAV diagnosed in paediatric age.
Study population: One hundred and sixty pts out of 250 were diagnosed before age 10 of BAV: they represent our study population. Male 111(69%); mean age at last examination 13.7 yrs (range 0.5–33 yrs); mean f/up period 8.8 yrs (range 0.5–23 yrs). All pts underwent clinical evaluation, ECG, complete 2D echo and when feasable exercise treadmill test. Morphologic anatomy of the aortic

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valve; presence and degree of valvar dysfunction; aortic diameter at annulus, sino-tubular and proximal ascending aorta level were evaluated.
Results: The aortic valve showed normal function in 43 pts(26.9%); in 86 (53.7%) there was aortic regurgitation (AR), mild in 71(82.5%) and severe in 6(17.5%). Valvar stenosis (AS) was present in 31 (19.4%), mild in 18(58.1%), moderate in 7(22.5%) and severe in 6(19.4%). All pts with dysfunctioning valve and only 3(7%) of those with normally functioning valve had a median anterior raphe. Dilation of aorta was present in 27/160 and in all cases it was at both the sino-tubular junction and at ascending aorta level. In 15/27 the valve showed mild AR or normal function. During the f/up period all pts remained in NYHA functional class 1. 4/160 with coarctation of aorta were on medical treatment. Progression of valvar dysfunction occurred in 7/31 pts with AS (mean gradient from 35.5 to 71 mmHg) and in 3/36 with AR from mild to moderate. During the study period 4 pts underwent surgery (3 valvar replacement and 1 Ross operation). Increase in aortic dilation was documented in only 2/160 pts(1%).
Conclusion: Valvar dysfunction and dilation of aorta appear relevant in a young population affering to tertiary care paediatric cardiology unit. Progression of valvar dysfunction occurs in a minority of pts whereas progression of aortic dilation seems to occur when increased diameter is already present at first examination. Surgery was electively undertaken in all cases.

16
Health status and quality of life in children post-Fontan procedure – comparison to their healthy siblings

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Purpose: The Fontan procedure has become the definitive surgery for patients with congenital heart defects (CHD) who have single ventricle physiology, yet long-term health status and health related quality of life (HRQL) has not been determined. The purpose of this study was to determine HRQL in children following the Fontan procedure.
Methods: Medical records were reviewed for Fontan survivors aged 10 to 18 years, and consenting families were mailed questionnaires. Parents completed the parent version of the PedsQL, and patients completed the PedsQL, Child Health Questionnaire (CHQ) and a validated disease-specific measure called the CHAT. Where available, a sibling closest in age completed the CHQ and PedsQL for comparison.
Results: Of 95 eligible patients, 68 families participated including 40 siblings. Mean patient age was 13 ± 4 years. Physical disabilities were reported in 21%, 67% were on medications, and 39% were getting extra help in school with 25% reporting learning disability. On the PedsQL, parents' assessments were slightly but not significantly lower than the patients' self-assessment. However, siblings scored themselves significantly ($p < 0.001$) higher than patients for physical (mean 97.9 v. 94.3/100), emotional (93.9 v. 90.2), social (96.5 v. 90.4) and school functioning (91.9 v. 88.0). Normalized CHQ scores showed patients to have normal physical functioning, general and mental health and freedom from bodily pain, but greater than normal freedom from behavior problems and social limitations. However, compared to their unaffected siblings, patients had significantly lower global health ($p < 0.001$) and greater social limitations due physical problems ($p = 0.03$). Disease-specific issues from the CHAT showed 35% of patients to have important tiredness (nearly all mild), with 47% having limitations to vigorous activity. Cardiac concerns were the cause of reported anxiety in 20%, school absence in 21% and feelings of difference in 23%.
Conclusions: While generally good, children and adolescents after the Fontan have significantly lower HRQL, particularly when compared to their unaffected siblings. Factors associated with HRQL in these patients need to be defined.

17
Long-term outcome and quality of life after Fontan operation

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Background: The first successful Fontan operation was performed in 1971 and now the first cohort of Fontan patients reach adulthood. The aim of this study is to evaluate our adult Fontan patients, who are seen in the outpatient clinic for congenital heart disease, to assess mortality, morbidity and quality of life.
Methods: All patients, who have undergone a Fontan procedure and are seen in the adult outpatient clinic, were included. The population was divided into two groups: (1) patients who had a Fontan operation before the age of 18 years and have reached adulthood ($n = 29$) and (2) patients who underwent a Fontan operation at adult age (>18 years) ($n = 7$). We studied the long-term follow-up from Fontan operation until last follow-up or death, to assess the incidence of reoperations, arrhythmias, hospitalisation and thromboembolic disease, and performed a quality of life study.
Results: Of the initial 36 patients, 10 died(28%) 10 years (range 0–21 years) after surgery and one patient underwent cardiac transplantation. The mean follow-up period of all patients after Fontan operation was 15 years (range 0–23 years), for group (1) 17 years and for group (2) 6 years. Reoperations were performed in 21 patients(58%), the most common reason for reoperation was revision of the Fontan connection. A supraventricular tachycardia was observed in 20 patients(56%) with an increased incidence of arrhythmias with longer duration of follow-up. Thromboembolic events were detected in 9 patients(25%) of which 5 had adequate anticoagulants levels at the time of the events. The thromboembolic event was fatal for 3 patients. All patients needed at least one hospitalisation after their Fontan operation. A total of 195 hospital admissions were recoreded, including 46 cardioversions. In the quality of life assessment physical functioning, mental health and general health perception were significantly lower for patients compared to the Dutch population.
Conclusion: We found an ongoing mortality and very high morbidity during long-term follow-up after Fontan operation. Quality of life is compromised by arrhythmia, risk for thromboembolic events, recurrent hospitalisations and reoperations.

18
Right ventricular function analysis after repair of Tetralogy of Fallot (TOF) by an angiocardiographic pressure-volume loop: Options and limitations

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Background: The assessment of function of the volume loaded and enlarged right ventricle (RV) in patients late after repair of TOF is still a matter of concern. The options and limitations of analysing a single pressure-volume (PV) loop in order to assess RV function will be presented.

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Methods: Since 1998, 22 patients were examined at a median of 6.8 years after corrective surgery by cardiac catheterization. A single beat PV loop was constructed using biplane angiocardiographic volumetry to measure RV volume combined with simultaneous RV pressure measurement. RV stroke work (W) was calculated by planimetry of the area enclosed by the PV loop and normalised to body surface area (W/BSA) and RV enddiastolic volume (W/EDV). Additionally, the maximum velocities of RV systolic outflow (−Vmax) and diastolic inflow (+Vmax) were calculated from RV volume-time curve as measure for systolic and diastolic function respectively. Flow velocities were normalised for RV stroke volume (SV). Linear regressions were calculated between these variables and RV enddiastolic volume (RVEDV).
Results: There was a significant positive correlation between W/BSA and RVEDV ($r = 0.62$, $p < 0.01$) and W/EDV remained constant with increasing RVEDV ($r = -0.12$, NS). No correlation was found between the normalised in- and outflow velocities and RVEDV (+Vmax/SV vs. RVEVD: $r = 0.002$, NS; −Vmax/SV vs. RVEDV: $r = 0.08$, NS).
Conclusion: The analysis of a single PV loop allows to quantify the abnormal loading conditions of the RV after TOF repair by W/BSA. As expected W/BSA increases with RV enlargement as a consequence of increased volume load. The lack of decrease of W/EDV even in patients with large RV indicates that the volume loaded RV is capable to perform adequate work in a wide range. The analysis of a single PV loop does not allow to estimate myocardial performance. Therefore the recording of multiple PV loops during load change is necessary.

Session 3: Arrhythmias/Pulmonary Hypertension

19
ECG Differentiation of AV node reentrant tachycardia from AV reciprocating tachycardia mediated by concealed accessory pathway in children

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The utility of the electrocardiogram (ECG) in children with supraventricular tachycardia (SVT) is unclear. The non-invasive differentiation of typical atrio-ventricular node reentrant tachycardia (AVNRT) and atrio-ventricular reciprocating tachycardia mediated by concealed accessory pathway (AVRT) is clinically important as it helps in counseling and potentially facilitates ablation procedures. One hundred and forty-eight electrocardiograms showing narrow QRS complex SVT were obtained from children (age: 10.6 + 3.9 years), prior to successful radiofrequency catheter ablation. An initial 102 ECGs were analyzed by 3 blinded observers to assess the utility of various ECG findings. No ECG criteria were found to discriminate between SVT mechanisms on 1- to 3-channel Holter/event recorder tracings (n = 32): their interpretation mainly (55%) resulted in a wrong SVT diagnosis. On 12-lead ECG (n = 70), the 2 arrhythmias were accurately assigned in 76%; 5 electrocardiographic findings were found to be discriminators of tachycardia mechanism. Predictors of AVRT were visible P-waves in 74% (sensitivity 92%; specificity 64%), RP interval >100 ms in 91% (sensitivity 84%; specificity 91%) and ST-segment depression > 2 mm which persisted 80 ms after the J point in 73% (sensitivity 52%; specificity 82%). Pseudo-r′ waves in V1 and pseudo-S waves in the inferior leads during tachycardia predicted AVNRT in 100% (sensitivity 55% and 20% respectively; specificity 100% for both). Based on these results, we developed and tested a new diagnostic 12-lead ECG algorithm for the appearance of pseudo-r′/S-waves, RP-duration and ST-segment depression during tachycardia. Stepwise analyzing allowed to differentiate AVNRT (n = 21) and AVRT (n = 25) in 42 of 46 (91%) additional cases. Thus, the stepwise use of diagnostically relevant 12-lead ECG parameters helps to more accurately differentiate mechanisms of reentrant SVT. By contrast, interpretation of SVT mechanism on Holter/event recorder tracings is frequently misleading.

20
Cardiac resynchronization therapy in congenital heart disease

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Introduction: Usefulness of cardiac resynchronization therapy (CRT) in congenital heart disease (CHD) has so far not been demonstrated.
Methods: 8 pts after surgery for CHD with systemic right (N = 6) or left (N = 2) ventricular dysfunction and spontaneous (bundle branch block, N = 2) or conventional DDD pacing induced (3rd degree AV block, N = 6) systemic ventricular conduction delay aged 6.3–29.2 yrs underwent biventricular pacemaker implantation/upgrade using epicardial (N = 7) or transvenous (N = 1) systemic ventricular leads and were followed-up for a median of 4.5 mo.
Results: There were no procedure related complications. Following biventricular pacing QRS duration decreased from 158 ± 24 to 111 ± 20 ms ($p < 0.005$), systemic ventricular ejection fraction (radionuclide ventriculography) increased from 41 ± 8 to 48 ± 5%, systemic AV valve regurgitation dv/dt from 37 ± 7 to 54 ± 12 m/s^2 ($p < 0.05$ for both) and systemic ventricular filling period from 43 ± 8 to 48 ± 8% of respective RR interval ($p < 0.005$). A rate dependent increase in arterial pulse pressure ranging from a mean of 6.2% at 50% (pNS) to 18.0% at 80% ($p < 0.05$) of calculated maximum age related heart rate was observed during acute hemodynamic testing after implantation.
Conclusion: CRT is feasible in CHD with the failing right or left systemic ventricle and improves systemic ventricular filling, ejection fraction and pulse pressure over short-term follow-up. Long-term benefit of this new heart failure therapy in terms of reverse remodeling needs to be established. (This study was supported by the Research Project of University Hospital Motol No 000 000 64 203)

21
Multicenter cryoablation experience in pediatric patients

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Transcatheter cryoablation (cryo) is now available for tachydysrhythmia therapy. We report the first multicenter cryo experience in pediatric (<18 years) patients (pts).
Methods: Data collected prospectively since 4/02 were available for 45 pts aged 14 ± 3 (mean ± SD) years undergoing cryo at 10 centers participating in the CryoCath® International Patient Registry. Dysrhythmia duration was 4.9 ± 4.3 years, with diagnoses of AV node reentry (AVNRT, 22 pts), anteroseptal (8 pts) or other (11 pts) accessory pathway (AP) mediated AV reentry, and ventricular tachycardia (VT, 4 pts). Cryo was offered by cardiologist preference

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with written informed procedural consent of each patient and/or legal guardian. The cryo system was approved for use in each country from which data were received. Cryo was delivered under general anesthesia (19 pts) or conscious sedation (18 pts) using CryoCath® (Kirkland, Canada) 7F Freezor® 4 mm tip or Freezor Xtra® 6 mm tip catheters via femoral (36 pts) and/or other (4 pts) venous and/or femoral arterial (1 pt) access. Cryomapping was performed at −30°C and permanent ablation delivered with 4 minute (min) applications at −75°C. Acute success was determined by criteria used in radiofrequency ablation (RFA) therapy.
Results: Summary data are shown in the Table. No significant complication or sedated pt discomfort occurred. RFA applied in 10 cryo failures was successful in 4/4 AVNRT and 5/6 AP pts.

Substrate	Success	Maps	Lesions	Proctime	Fluorotime
AVNRT	17/22	5(1–12)	2(0–9)	169(51)	20(12)
AntrosptAP	5/8	9(5–11)	4(1–17)	222(77)	44(27)
OtherAP	3/11	6(1–22)	3(1–20)	226(96)	48(32)
VT	3/4	0	9(9)	238(88)	28(3)

Conclusions: Cryo is a safe and well-tolerated alternative to RFA in pediatric pts based on our initial experience. Success is highest in AVNRT and in substrates recognized as technically challenging or risky for RFA, such as antero- or midseptal AP where a 9% incidence of RFA-induced heart block has been reported.

22
Influence of bradycardia on right ventricular function after the atrial baffle procedure for transposition of great arteries

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Aim: Patients after the Mustard or Senning procedure frequently have sinus node and right ventricular (RV) dysfunction. Indications for antibradycardia pacing are not well defined. This study evaluates influence of bradycardia on RV function during longitudinal postoperative follow-up.
Methods: RV function was repeatedly assessed by radionuclide ventriculography in 73 pts at a median of 4.4 and 13.2 yrs postoperatively. Degree of bradycardia was quantified by serial 24-hour Holter monitoring as an average of a mean of 2.7 (2–7) evaluations/pat during the same interval. Both uncorrected and age corrected minimum, average and maximum heart rates (HRc) and maximum RR intervals (RRc) were used for analysis.
Results: RV ejection fraction (RVEF) and echocardiographic grade of tricuspid regurgitation (1) evaluation: mean 42.2 ± 6.2% and 1.0 ± 1.0; (2) evaluation: mean 44 ± 7.6% and 1.3 ± 1.0, p = NS) did not correlate with the degree of bradycardia. In 26 pts with RVEF decrease (mean 5.2, range 1–17%) minimum (38.0 ± 6.4 vs 39.8 ± 6.5 bpm) and average (70.9 ± 11.8 vs 67.4 ± 13 bpm) HRc and maximum RRc interval (1.8 ± 0.4 vs 1.6 ± 0.4 s) did not differ from the rest of the group (NS for all). In 15 pts with significant bradycardia (minimum HRc < 35 bpm) change in RVEF (−0.1 ± 7.8 vs 2.4 ± 6.4%, p = NS) and increase in tricuspid regurgitation (by 0.4 ± 1.0 vs 0.2 ± 0.8 grades, p = NS) did not differ from the remaining pts.
Conclusion: Asymptomatic bradycardia is not a risk factor for the development of RV dysfunction after the atrial baffle procedure for transposition of great arteries and does not represent a specific hemodynamic indication for permanent antibradycardia pacing.
(This study was supported by the Research Project of University Hospital Motol No 000 000 64 203.)

23
Automatic implantable cardioverter-defibrillators in pediatric patients in The Netherlands

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Background: The indication for automatic implantable cardioverter defibrillators (AICD) in children as therapy for life-threatening arrhythmias is infrequent and the experience of each single institution is limited. This emphasizes the need for a nationwide pediatric AICD registry that includes follow-up data.
Methods and Results: All pediatric cardiology centers in the Netherlands contributed to the database. Since 1995 until June 2002; 23 pediatric AICD implantations were performed. Mean age was 11.3 ± 5.3 yrs, range 0.5–18 yrs, mean weight 46 ± 25 kg. The indication for AICD was aborted sudden death (n = 14), syncope (n = 5) or positive family history (4). Diagnosis included primary electrical disease (n = 16), hypertrophic obstructive cardiomyopathy (n = 3), OCTN2 deficiency (n = 1) and congenital heart disease (n = 3). Electrophysiological studies were performed in 12/23 pts and VF/VT was inducable in 5 pts. 18/23 AICD systems were placed transvenously while an epicardial or subcutaneous system was used in 5 pts. The generator was implanted subpectorally in 15/23. There were no major complications and the mean defibrillation threshold was 15 ± 6 J at the time of implantation. The mean follow-up was 29 ± 28 mos (0–7 yrs). One infant with LQTS and repetitive polymorphic VT and appropriate shocks died 4 mos after implant. The other 22 pts were in good clinical condition and 13/23 used antiarrhythmic drugs during follow-up. Appropriate shocks occurred in 5/23 after a mean follow-up of 9 mos. Inappropriate shocks took place in 7/23 due to sinus tachycardia (n = 4), supraventricular tachycardia (n = 1) or T-wave sensing (n = 2) usually in the first mos after implantation. Revision of the AICD system was necessary in 4/23 due to transvenous lead fracture (n = 1) and battery depletion (n = 4). There were no infections.
Conclusion: AICD therapy is infrequent in the pediatric population and is usually indicated in children with primary electrical disease. Inappropriate shocks frequently occur shortly after implantation. It is safe and effective and the complication rate during follow-up is low.

24
Cardiac index and endurance time to exercise test during vvir and av sequential pacing in children with VDD/DDD pacemakers: the effect of atrioventricular delay settings

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AV sequential pacing (AVSP) more closely resembles the normal physiology of cardiac activation than VVIR pacing. This beneficial effect may not be apparent at rapid rates. The aim of this study was (1) to measure the cardiac index at rest and exercise performance in both pacing modes and (2) to evaluate the influence of atrioventricular (AV) delay on cardiac index (CI) during AVSP at rest. Thirteen children (eight girls, five boys, mean age of 13.8 ± 4.3 years, mean weight of 45.1 ± 18.4 kg) with a VDD/DDD pacemaker were evaluated by Doppler echocardiography and exercise stress test. Cardiac output was calculated from aortic velocity time integral and heart rate and it was divided body surface area for

calculating CI. During AVSP with 100, 125, 150, 200 ms AV intervals, and VVIR pacing, CI was measured at rest. Endurance times to exercise, with a treadmill using modified Bruce protocol, were assessed during both in AVSP with the optimal AV delay and VVIR pacing. The optimal AV delay which provides highest CI was 100 ms in four, 125 ms in two, 150 ms in four and 200 ms in three patients during AVSP. VDD/DDD pacing with 100 ms, 125 ms, 150 ms and 200 ms AV intervals resulted in a significantly higher CI (6.59 ± 2.95, 6.49 ± 2.39, 6.27 ± 2.30, 6.65 ± 2.65 l/min/m^2 respectively) than VVIR pacing (5.22 ± 2.23 l/min/m2) at the rest ($p < 0.05$). However endurance times to treadmill exercise were similar in both the AVSP mode with the optimal AV delay (22.4 ± 3.4 minutes) and VVIR pacing mode (21.7 ± 3.8 minutes) ($p = 0.093$). 1. Doppler echocardiographic measurement of CI is a simple, noninvasive method for estimating optimal AV delay in children with AV sequential pacemakers. 2. Each child should be programmed according to optimal AV delay measurement. 3. AVSP with VDD/DDD pacemakers with the optimal AV delay seems superior to VVIR pacing with respect to CI at rest. 4. The benefit of AVSP, which were proven at rest with echocardiography, were not occult during conventional exercise test. These results may be evaluated further with the metabolic parameters during exercise test, since conventional exercise test may not reflect true exercise performance.

25

Neurophysiologic abnormalities in children treated with amiodarone

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Amiodarone, a highly effective drug in the treatment of refractory cardiac arrhythmias, is known to cause many neurologic side effects. Among those, tremor, ataxia and peripheral neuropathy with or without spinal cord involvement are the most common neurotoxic findings. With increasing use, a number of peripheral neuropathy cases associated with amiodarone treatment have been reported. The reported prevalence of the peripheral neuropathy varies from 2% to 10%. The aim of this study was to evaluate the side effects of amiodarone on peripheral nerve fibers in children. Fourteen children with a mean age of 9.3 ± 5.2 years (1 to 16 years) treated with amiodarone for cardiac arrhythmias were included in this study. The median duration of treatment was 8 months (3 months–10 years), and the mean cumulative dose 2286 ± 1291 mg/kg (ranged 300 to 6935). Patients were examined for concurrent factors or disorders causing polyneuropathy. Neuroelectrophysiological studies were performed using Medelec Synergy System. Nerve conduction velocities (NCV) were measured from one upper and one lower extremity. Motor and sensory NCV, distal latency and compound muscle and sensory action potential amplitudes were measured. Median, ulnar, common peroneal and tibial motor nerves and median, ulnar and sural sensory nerves were evaluated with surface stimulating and recording techniques. The electrophysiological variables were compared with normal reference values obtained from pediatric nerve conduction studies. An electrophysiological diagnosis of polyneuropathy was made when conduction velocity, or distal latency, or amplitude, were abnormal in at least two nerves. Two patients (14%) had abnormal electrophysiological findings. NCVs were decreased in one patient and compound muscle action potential amplitudes were diminished in the other. These two patients had been taking a cumulative amiodarone dose of 1650 and 4375 mg/kg. In this study we detected both slowing of conduction velocity and decrease of compound muscle action potential amplitudes that is evidence of demyelinization and axonal loss respectively. This study suggest that in patients treated with amiodarone periodical electrophysiological studies are required for early detection of peripheral neuropathy.

26

Current experience with Bosentan (Tracleer) in the treatment of primary and secondary pulmonary hypertension in children

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Introduction: Pulmonary hypertension (PH) is a rare but serious disease. The most effective therapy to date has been continuous intravenous epoprostenol, a relatively complicated therapy with risk of sepsis. Other options are limited in children with severe disease. In adults treatment with the orally active ET-A and ET-B receptor antagonist Bosentan improves exercise capacity and cardiopulmonary haemodynamics[1]. Little is known about its effect in children. Elevation of liver enzymes was the commonest side effect in adults.

Aims: Examination of the safety and efficacy of Bosentan in children with PH. Characteristics of patients treated: 17 children aged between 9 months and 16 years, mean 6.7 years (6 males and 11 females) were treated with Bosentan for 1–12 months (mean 4.6 months). 7 children had primary and 9 had secondary PH. All had recent clinical deterioration and limited exercise tolerance. 8 children had symptoms before 1 year of age, 7 at 1–5 years and in 2 developed symptoms after 6 years of age (mean age 2.2 years). Echocardiography showed moderate to severe right atrio-ventricular valve regurgitation and a dilated and hypertrophied right ventricle in all children with a velocity above 4 m/sec in 14 cases. The pulmonary artery pressure was ⩾ systemic pressure in 14 children. The pulmonary vascular resistance was elevated in all and ⩾11 units/m^2 in 12 children. It didn't fall significantly in response to acute vasodilator testing.

Results: Most children showed clinical improvement as indicated by improved exercise tolerance and a beneficial shift in NYHA classification (Fig. 1). 5 children showed a reduction in TR velocity jet. No adverse effects have been noted to date.

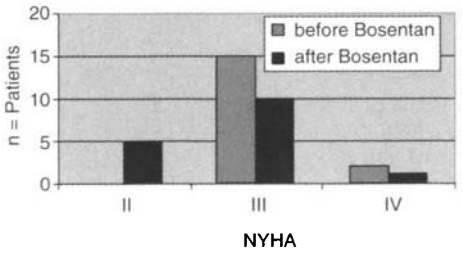

Fig. 1 NYHA Classification before and after Bosentan treatment

Conclusion: Our experience with Bosentan in treating severe primary and secondary PH in young children is encouraging. Clinical improvement including improved exercise capacity and cardiopulmonary haemodynamics is seen as early as one month after the commencement of treatment. Chronic administration of an oral rather than an intravenous drug is a huge advantage, particularly in children

Reference:

1 Rubin, LJ, David B, Badesch M.D et al; Bosentan therapy for pulmonary artery hypertension, N Engl J Med, Vol. 346, No 12, March 21, 2002

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27

Aerosolized versus intravenous iloprost in secondary pulmonary hypertension in children with congenital heart disease

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In this study, we compared the effects of inhaled and intravenous application of iloprost, a stable analogue of prostocyclin, on pulmonary hemodynamics in children with pulmonary hypertension secondary to congenital heart disease. Twelve children (age ranged 6 months to 16 years) with pulmonary hypertension secondary to congenital heart disease were prospectively studied. Patients were first given intravenous iloprost 25 ng/kg/min for 10 minutes; then, after baseline values were reached again, they were given aerosolized iloprost at 25 ng/kg/min for another 10 minutes. Hemodynamic measurements were performed prior to and after each application. Both aerosolized and intravenous infusion of iloprost caused significant decrease in pulmonary artery pressure (PAP) and pulmonary vascular resistance (PVR) ($p < 0.05$). Intravenous infusion caused prominent decrease in mean systemic arterial pressure (SVR) ($p < 0.05$), this was only slightly affected by aerosolized iloprost ($p > 0.05$). Aerosolized iloprost caused a significant decrease in PVR/SVR ratio ($p < 0.05$), however intravenous infusion increased this ratio. Aerosolized iloprost also increased the pulmonary to systemic blood flow (Qp/Qs) ratio ($p < 0.05$), but intravenous infusion slightly decreased this ratio ($p > 0.05$). Aerosolized iloprost was well tolerated by all patients, however clinically apparent hypotension was seen in six patients during the intravenous application. In conclusion, intravenous application of iloprost decreases SVR more effectively than PVR, and deteriorates pulmonary homodynamics. In children with pulmonary hypertension secondary to congenital heart disease, aerosolized iloprost is effective in selectively lowering PVR.

	Mean PAP	Mean SAP*	Qp/Qs	PVR/SVR
Baseline	59.3 ± 14.3	70.8 ± 11.5	1.8 ± 0.9	61.9 ± 46.8
Inhalation	46.2 ± 16.3	70.1 ± 9.2	3.4 ± 2.3	38.6 ± 64.9
Intravenous	40.3 ± 18.9	49.3 ± 13.2	1.7 ± 0.8	108 ± 239

*SAP: Systemic arterial pressure.

Session 4: Basic Sciences

28

Increased myocardial MMP-2 activity after resuscitation with 100% O_2 in hypoxic newborn piglets

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Background: Matrix Metalloproteinases (MMPs) play an important role in cellular and structural changes in heart disease, such as myocardial infarction, congestive heart failure and cardiomyopathy. Perinatal asphyxia is associated with cardiac dysfunction secondary to myocardial ischemia. During resuscitation it is important to prevent further myocardial damage due to reperfusion. The nature, however, of hypoxemia/reoxygenation injury is still not fully understood. *Objective:* To assess cardiac remodelling by measuring MMP-2 and MMP-9 activity in neonatal porcine myocardium after global ischemia, and subsequent resuscitation with ambient air or 100% O_2. To study any effect of differences in pCO_2-levels. Blood samples were analysed for cardiac Troponin I (cTnI).

The Model: Newborn piglets (12–36 h) were exposed to hypoxaemia by ventilation with 8% O_2 in nitrogen. There were no differences between the groups in number of animals, age, weight, hemoglobin or hypoxemia time. When mean arterial blood pressure had fallen to 15 mmHg, or base excess < -20 mmol/L, the animals were randomly resuscitated by ventilation with 21% O_2 (group A, n = 28) or 100% O_2 (group B, n = 29) for 30 minutes. Afterwards they were observed in ambient air for another 150 minutes. During resuscitation the two groups were randomly divided into 3 subgroups with low, normal and elevated CO_2 -levels. MMP activity was analyzed by gelatine zymografi, and the results were calculated by setting one normal control sample not included in the study to 1, used as an internal standard on every zymography run. All values are given as mean (±SEM).
Results: MMP-2 activity more than doubled (0,51 ± 1,5 – 1,15 ± 5,8) during the experiment. ($p < 0,001$) There were significant differences in MMP-2 activity ($p = 0,012$) between the group resuscitated with 100% O_2 (1,29 ± 0,08) compared to the group resuscitated with ambient air (1,0 ± 0,07). The CO_2 level did not influence on myocardial MMP-2 activity. MMP-9 was below level of detection. CTnI increased more than 10 fold (0,96 µg/L ± 0,01 – 2,18 µg/L ± 0,37), $p < 0.001$, indicating marked myocardial affection, although circulation was rapidly restored (blood pressure/heart rate/Base Excess).
Conclusion: After global ischemia and subsequent reoxygenation, myocardial MMP-2 activity was higher in piglets resuscitated with 100% O_2 than those resuscitated with ambient air.

29

Vascular mediators in the flow- or pressure-overloaded pulmonary circulation

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Background: Endothelial activation and alterations in vasoactive mediators like endothelin (ET) and nitric oxide (NO) have been implicated in the regulation of pulmonary vascular tone and the development of pulmonary hypertension (PH). Therefore, soluble markers of endothelial damage and vasoactive compounds were compared in patients with increased pulmonary flow (atrial septal defect, ASD) and with pressure overload due to increased pulmonary resistance (Rp).
Patients: This study included 20 patients (median: age 6.1 years [range 3.5–17.1], Qp/Qs 2.1, Rp/Rs < 0.12) before and after interventional closure of an ASD and 20 patients with PH (median: age 8.1 years [1.2–13.5], Rp/Rs 1.1 (0.36–1.79).
Methods: Plasma NO_2, NO_3 (GC-MS) and the endogenous NO inhibitor asymmetric dimethylarginine (ADMA) (HPLC) were analyzed in the pulmonary (PA) and femoral artery (SA). Big-ET, ICAM and VCAM were measured in PA samples (ELISA).
Results: (1) ASD patients showed increased Big-ET (1.45 fmol/ml, 0.51–5.69, control: 0.95 ± 0.68, $p < 0.001$) and a significant transpulmonary ratio of 1.34 for NO_2 (median, $p < 0.01$ vs 1.0), but not for ADMA (1.05) or NO_3 (1.01). (2) ASD closure resulted in a decrease of Big-ET (1.22, 0.42–4.92) and the NO_2 transpulmonary ratio (0.89, $p < 0.05$), indicating a switch from NO_2 production to consumption. ADMA (1.00) and NO_3 (0.99) remained unchanged. (3) In PH, significant transpulmonary ratios were observed for ADMA (1.11, $p < 0.05$) and NO_3 (1.03), but not for NO_2 (0.84) reflecting inappropriate NO_2 production. Big-ET (2.08, 0.97–23.90) and VCAM (967 pg/ml, 348–2617 vs 770, 407–1412 in ASD patients, $p < 0.05$) were increased.

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Conclusions: Different loading conditions of the pulmonary circulation are reflected by load-dependent changes in the intravascular hemeostasis of the Big-ET and NO pathways and activation of the pulmonary endothelium.

30
The myocardial expression of growth factors in infants with congenital cardiac defect involves the activation of the Jak/STAT pathway

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Background: Cardiac remodeling and hypertrophy in response to hemodynamic overload due to congenital cardiac defect could involve the up-regulation of growth factors such as vascular endothelial growth factor (VEGF), interleukin (IL)-6 and cardiotrophin (CT)-1, and the regulation of apoptosis. Our study was intended to test this hypothesis and to identify the signaling pathway involved.
Methods and Results: Concentrations of IL-6 and CT-1 were measured by ELISA and/or Western blot in the right ventricular myocardium sampled during corrective cardiac surgery of infants with tetralogy of Fallot (TOF) ($n = 6$) or with ventricular septal defect (VSD) ($n = 6$). Infants with TOF had higher levels of IL-6 ($p < 0.05$) but similar concentrations of CT-1, as compared to patients with VSD. Concentrations of IL-6 correlated with right ventricular pressure (Spearman: 0.68, $p < 0.05$). Levels of phosphorylation of the signal transducer and activator of transcription (STAT)-3 was measured by Western blot and was higher in infants with TOF than in those with VSD ($p = 0.1$). Myocardial levels of the apoptosis inhibiting protein Bcl-xL and of VEGF, both target genes of STAT-3, were measured by Western blot and were higher in infants with TOF than in those with VSD ($p < 0.05$; $p < 0.1$, respectively).
Conclusions: In infants with congenital cardiac defect, mechanical stress due to hemodynamic overload activates the Jak/STAT pathway in the myocardium. This leads to the up-regulation of the growth factors IL-6, CT-1 and VEGF and of the apoptosis inhibitory protein Bcl-xL. Up-regulation of growth factors and inhibition of apoptosis are likely to be involved in myocardial remodeling and hypertrophy in this age group.

31
Myocardial volume and organization are changed by failure of addition of secondary heart field myocardium to the cardiac outflow tract

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Cardiac neural crest (CNC) ablation in chick embryos results in the loss of cardiac outflow tract septation and malalignment of the outflow tract (1). In addition to the loss of aorticopulmonary septation, absence of cardiac neural crest cells in the pharynx of neural crest-ablated chick embryos results in primary myocardial dysfunction initiated prior to the time that neural crest cells normally populate the heart (2). This dysfunction is accompanied by failure of the secondary heart field to add the definitive myocardium to the cardiac outflow tract (3). The current study was undertaken to understand the changes in myocardial characteristics in the heart tube including volume, proliferation and cell size in sham-operated and neural crest-ablated embryos by using methods such as Magnetic Resonance Microscopy, digital microscopic analysis of myocardial proliferation, confocal microscopy for cardiac morphometry, immunohistochemistry and cell size measurements of freshly dissociated myocardial cells. The volume of myocardium in the looped heart was dramatically reduced after neural crest ablation, especially in the outflow tract and ventricular regions. Myocardial cell size was increased and the cells in the looped tube showed increased proliferation in all regions of the heart, especially in the outflow tract. At the same time, myocardial cell death appeared to be normal. Even with elevated proliferation and normal cell death, the compact layer of the myocardial wall was thinner in neural crest-ablated embryos during and just after looping. By incubation day eight when the heart would have normally completed septation, the anterior (ventral) wall of the right ventricle and right ventricular outflow tract was significantly thinner in the neural crest-ablated embryos than normal. Since myocardial cell death was not elevated and cell proliferation increased, the decreased volume of myocardium in the heart tube is most likely due to the failure of secondary heart field to add myocardium to the outflow tract.

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32
Characterization of chronic right ventricular pressure overload in young lambs: evidence of a hyperplastic response

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Objective: In the neonatal period, cardiac pressure overload is present in several types of congenital heart or lung disease. Cardiac function might be affected by the type of cellular adaptation of the overloaded myocardium (i.e. hypertrophy/hyperplasia) as well as by (re)expression of fetal genes.
Methods: Young lambs underwent adjustable pulmonary artery banding for 8 weeks to obtain peak right ventricular (RV) pressures equal to left ventricular (LV) pressures after which transmural cardiac tissue samples from the RV, LV and interventricular septum (IVS) were subjected to morphometric and biochemical analysis. The latter involved determination of DNA concentrations, isoform distributions of cardiac myosin heavy chain (MHC) and troponin I (TnI).
Results: Chronic RV pressure overload resulted in a significant increase in RV-to-LV wall thickness ratio (0.43 ± 0.04 to 0.94 ± 0.15, $p < 0.01$). Morphometric analysis revealed no changes in myocardial tissue composition (expressed as relative volume fractions) or individual myocyte dimensions (length, width, area). As to the distributions of myocyte area, no differences between the control and RV overload group were observed. However, in the latter group, the proportion of binucleated myocytes at any given myocyte size had decreased as compared to control ($p < 0.05$). Myocardial DNA concentrations in any of the samples studied were not significantly different from control values. Chronic RV overload significantly decreased the a-MHC isoform in favour of the b-MHC isoform in RV and IVS samples. Western blotting revealed two TnI protein bands in both groups, a prominent one

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at 31 kD and a faint band at approximately 28 kD. The latter was more pronounced in the RV overload group and probably represents reexpression (or delayed downregulation) of a fetal TnI isoform, slow skeletal TnI.
Conclusions: The young heart is able to respond to increased work load by means of myocyte hyperplasia which is accompanied by a shift in the MHC isoform expression towards a state in which contraction is energetically more efficient under conditions of increased workload. In addition, chronic RV pressure overload induces reexpression (or decreased downregulation) of the fetal TnI isoform which may underlie diastolic dysfunction following chronic RV pressure overload as has been demonstrated by us previously.

33
Systolic left ventricular dysfunction and compensatory dilatation in juvenile rats treated with glucocorticosteroids in neonatal period

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Background: In preterm infants chronic lung disease (BPD) is a major cause of morbidity and mortality. Glucocorticosteroid (dexamethasone) treatment has been successful in weaning very low birth weight infants from mechanical ventilation. However, major concern has emerged about negative effects on the cardiovascular system. Short-term reversible cardiac hypertrophy has been described. Recent studies in rats indicate hypertension, alterations in myocyte structure later in life, and reduced life expectancy.
Study Objectives: Hemodynamic studies will be performed in three age-groups: juvenile (4 wks), post-puberty (8 wks) and "adults at risk" (50 wks) to explore GC-induced changes in cardiac function during the span of life. The present study reports our initial findings in the juvenile rats.
Methods: Rat pups were injected (IP) with dexamethasone (0.5, 0.3 and 0.1 μg/g) on day 1, 2 and 3 of life. In the control group saline was used. At the age of 4 wks the animals were anesthetized (dormicum/hypnorm), intubated and ventilated. A miniature (1.4 F) pressure–conductance catheter was introduced into the left ventricle (LV) via the right carotid artery to measure LV pressure–volume loops. The left jugular vein was cannulated for injection of hypertonic saline to calibrate the conductance measurements. A loading intervention was performed by compression of the vena cava inferior via a small skin incision below the diaphragm. Cardiac function was quantified by systolic and diastolic LV pressure–volume relations obtained from pressure–volume loops registered during the loading intervention.
Results: Body weight was significantly lower in the dexamethasone-treated animals (N = 10) compared to age matched controls (N = 8) from day 2 up to day 63. In the 4 week old rats, end-diastolic volume (EDV), end-systolic volume, stroke volume and cardiac output were significantly higher in the GC-group. The contractile state was significantly lower in the GC-group, as evidenced by the slope (Ees) of the end-systolic pressure–volume relation (9.4 ± 2.7 vs 17.6 ± 7.7 mmHg/microL, p = 0.03) and the slope of the dP/dtMAX – EDV relation (719 ± 295 vs 1410 ± 662 mmHg/s/μL, p = 0.03).
Conclusion: Our findings indicate that the reduced life expectancy and myocardial degeneration previously found in aged rats (treated by dexamethasone in neonatal period) are preceded by systolic dysfunction and compensatory cardiac dilatation in juvenile (4 weeks old) rats.

34
Bradykinin prevents arterial endothelial dysfunction caused by ischemia-reperfusion in humans

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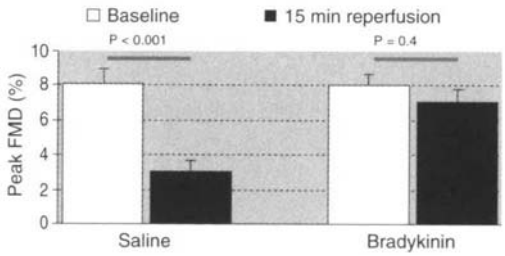
Background: Endothelial dysfunction of epicardial coronary arteries is an ubiquitous finding during open heart surgery (OHS) in children, and emerges as a crucial mechanism in the pathogenesis of reperfusion-induced myocardial damage. Earlier studies using young animal models of ischemia-reperfusion (I/R) via cardiopulmonary by-pass, which is customarily used during OHS, have shown that intracoronary preinfusion with 5 μg/ml bradykinin (BK) could alleviate the myocardial damage caused by I/R. Using an experimental human model of I/R, we investigated whether intra-arterial pre-infusion with 5 μg/ml BK could prevent I/R-induced arterial endothelial dysfunction.
Methods and Results: The left radial artery (LRA) from sixteen healthy male volunteers, 18–30 years old, was submitted to I/R by completely occluding the left brachial artery with a pressure tourniquet for 20 min (ischemia), followed by its release (reperfusion). Half of the subjects were randomly assigned to receive either BK (5 μg/ml) or saline, both being infused into the brachial artery (0.5 ml/min, 10 min). The infusion was followed by a 10 min drug-free period. The endothelial function of the LRA was studied by measuring in a blind fashion the flow-mediated dilatation (FMD) at baseline (prior to drug infusion), and at 15 min of reperfusion. Plasma nitrate and von Willebrand factor were measured at these time points, and immediately before I/R. FMD was significantly decreased by I/R in the saline-pretreated group, whereas no such effect of IR was observed in the BK-pretreated group (Figure). Plasma nitrate was higher in BK-pretreated group than in saline-pretreated group ($p < 0.05$).



Conclusion: This is the first study to show that a small dose of BK could protect the arterial endothelial function against the damaging effects of I/R in humans, possibly via increased availability of endothelial NO. The finding supports the use of BK as an additive to cardioplegic solutions in the prevention of I/R-induced tissue injury during OHS.

35
Acute infections in children promote oxidative modification of LDL, and are followed by thickening of carotid intima-media

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Background: Atherosclerosis begins in early childhood. Infection and inflammation have been hypothesised to contribute to the

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pathogenesis of atherosclerosis in part through increased lipid oxidation. We investigated whether acute infections in children could alter the carotid wall morphology and the lipid profile.
Methods: Mean carotid artery intima-media thickness (IMT) was measured by high-resolution ultrasound in twenty-eight hospitalised children (mean age: 5 ± 2 y.) who fulfilled the diagnostic criteria of acute infections (body temperature > 38°C, C-reactive protein > 15 mg/ml, and clinical), and in twenty age- and gender-matched controls. Antibodies against oxidized low-density lipoprotein (anti-oxLDL antibodies), as well as total and high-density lipoprotein cholesterol (HDL-C) were analyzed in all children. The infection group was investigated both during the acute illness and 3 months after clinical recovery (post-infection).
Results: During the acute illness, the infection group had elevated anti-oxLDL antibodies and decreased HDL-C, as compared to those obtained at 3 months and in controls ($p < 0.05$). These changes in the infection group were followed, at 3 months, by thickening of carotid intima-media. Those who received antibiotics during their acute illness had less carotid thickening than those who were not treated with antibiotics ($p < 0.05$).
Conclusion: Acute infections in children are accompanied by enhanced oxidative modification of LDL and by decrease of HDL-C. These lipid changes may be followed by thickening of carotid artery intima-media, and antibiotic treatment during the infectious illness appears to lessen the post-infection thickening. Subsequent acute infections, which are frequent in childhood, may, thus, pose additional burden and increase the risk for atherosclerosis.

36
Carvedilol improves I123-metaiodobenzylguanidine (MIBG) uptake and left ventricular ejection fraction in children with dilated cardiomyopathy

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Background: Beta-blocking agents reduce the risk of hospitalisation and death in adult patients with heart failure. Little information is available in children with dilated cardiomyopathy (DCM).
Objective: To evaluate the efficacy of the non-selective b-blocker carvedilol on left ventricular systolic function and MIBG uptake in children with DCM.
Patients: 25 children (median age 3 years, range 2 months–14 years) with heart failure (NYHA II[n = 10]; III[n = 15]) resulting from dilated cardiomyopathy: 11 idiopathic (3 familial cases), 4 mitochondrial DCM, 2 anthracyclin DCM, 5 after repair of congenital heart defect, and 3 various other disorders.
Methods: At the initial examination, echocardiographic variables, cardiac MIBG uptake and isotopic left ventricular (LV) ejection fraction were recorded. Carvedilol in addition to standard medical therapy (diuretics, ACE inhibitors) was given at an initial dose of 6.25 mg/1.73 m^2, uptitrated over 2 months to an average maintenance dose of 49.2 mg/1.73 m^2. After 3 months on carvedilol, initial measurements were repeated.
Results: Initial LV parameters were (mean ± SD): Z value of diastolic diameter +5 ± 1.7, fractional shortening 17 ± 2%, isotopic ejection fraction 27 ± 3%, MIBG uptake 164 ± 2%. Carvedilol had to be discontinued in 2 patients because of worsening cardiac failure. After 3 months on carvedilol, LV parameters improved (mean ± SD/delta of improvement): Z value of diastolic diameter +3.4 ± 3.9/−1.2 ($p = 0.03$); fractional shortening 25 ± 12%/+7% ($p = .01$); isotopic ejection fraction 40 ± 2%/+12% ($p = 0.002$); MIBG uptake 205 ± 68%/ + 48% ($p = 0.0003$).
Conclusion: Carvedilol as an adjunct to standard therapy for paediatric heart failure improves LV function and MIBG uptake. As MIBG uptake has been shown to be the only predictor of unfavourable outcome in children with DCM, carvedilol might improve adverse outcome.

Session 5: Intervention

37
Coil closure of very large (>5 mm) patent ductus arteriosus by bioptome assisted simultaneous multiple coil delivery technique

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Transcatheter closure of very large patent ductus arteriosus (PDA) is possible with the occlusive devices. However, the cost of the device is an important limiting factor, especially in developing countries. We present our experience in closing very large ducts using coils. Between July 1998 and December 2002, 712 PDAs were referred to our institution for closure. 110 (15.4% of them were closed surgically and the remaining 612 (84.6%) were closed by transcatheter techniques. 96 PDAs were measuring >5 mm, 61 of them were surgically managed and 35 were closed by transcatheter means. Young age and shallow ampulla were the main indications to subject a child for surgical closure (median age 6 months (1 month–13 years)). Six patients underwent Amplatzer device closure and 30 underwent coil closure. Gianturco coils were used in all and initial coil delivery was done from the venous end with the assistance of a bioptome. Two to six 0.052-inch coils were delivered simultaneously via a long sheath (7–12 F) placed across the duct. Whenever residual flow was noted, additional coils were delivered from arterial end. The median age in the coil group was 9.0 years (5 months–52 years). The median size of the duct was 6.0 mm (5.0–10.2 mm). The median fluoroscopic time was 13 minutes (4.1–53.2 minutes) and the median number of coils used per patient was 3.0 (1–11 coils). There were 7 instances of coil embolizations and retrieval of the coils was successful in all the times. Eight (27%) patients had a small residual flow immediately after the procedure with hemolysis in 2 patients (one required repeat deployment of additional coils). At 6 months follow-up, only 2 showed persistent mild residual flow. Three patients developed mild LPA origin narrowing with Doppler gradients of 13, 10 and 12 mmHg respectively. There was one instance of procedure failure in a child with 8.3 mm duct. In conclusion, transcatheter coil closure of very large PDAs is feasible as a less expensive alternative to surgery and occlusive devices. The limitations of coil occlusion technique include residual flows with the potential for hemolysis and a high rate of coil embolization.

38
Stent implantation in the ductus arteriosus versus surgical aorto-pulmonary shunt in duct dependent congenital heart disease – a retrospective study

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Stent implantation in the ductus arteriosus (DA) for pulmonary circulation has become a reasonable alternative to surgical

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aorto-pulmonary shunts with improvements in stent technology and increased investigator experience. Our study compares short and midterm outcome of neonates where the transcatheter approach or surgery were performed.
Patients: From 2/2001 to 12/2002; 25 neonates with duct dependent pulmonary circulation were admitted to our institution. Mean age was 25dd, mean weight 3.2 kg. Diagnoses were: Pulmonary atresia (PAT) with VSD (n = 2), PAT with intact ventricular septum (n = 1), complex PAT (n = 14),Tetralogy of Fallot (n = 2), Ebstein's anomaly (n = 2), Complete transposition (n = 1), Tricuspid atresia (n = 3).
Methods: In 13 pts. mainly premounted coronary stents were implanted in the DA with a final diameter of 4 mm. 3 pts had additional balloon valvuloplasty of the pulmonary valve, all procedures were performed in analgosedation. In 12 pts a central surgical shunt of 3–4 mm (PTFE) was installed, partially with cardiopulmonary bypass, in 2 of them DA stenting had been attempted before.
Results: There were no procedural deaths in both groups. In the stent group ductal patency was achieved for up to 456 days. Re-intervention was necessary in 5 pts (redilation n = 2, second stent n = 3). Early aorto-pulmonary shunt obstruction occurred in 4 pts, re-operation was effected in 3, balloon dilatation in 1 pt. Secondary pulmonary artery branch stenosis was observed in 3 pts. Cardiopulmonary resuscitation occurred in 2 pts, pericardial effusion in 6 pts after surgery and none in the interventional group. After ductal stenting, 1 pt had corrective surgery within the follow up time of at latest 1.3 years, 2 pts Fontan type procedures, 3 pts are solely cured by intervention, 7 are awaiting surgery. In the shunt group 2 had corrective and 3 palliative surgery, 7 are awaiting operations. Intensive care stay was 5dd (median) in the shunt group, none in the stent group. Pulmonary artery growth was comparable in both groups.
Conclusion: In neonates with duct dependent pulmonary circulation ductal stenting is a minimal invasive alternative to surgery. Depending on ductal morphology the transcatheter approach is not applicable to all patients.

39
Stenting the neonatal arterial duct in duct dependent pulmonary circulation: new techniques, better results

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Objectives: To assess the feasibility and safety of a new approach and technique to stent the arterial duct in neonates with a duct dependant pulmonary circulation.
Background: Previous attempts to stent the neonatal arterial duct were unsatisfactory: the early generation rigid stents were relatively large, bulky, required stiff wires and long sheaths, and only partially covered the duct. Learning from these failures, we speculated that covering the complete length of the duct with current low profile stents might avoid previous problems.
Methods: 10 neonates with duct dependant pulmonary circulations through a short straight duct were treated with stent implantation. The duct was crossed with an atraumatic 0.014" wire. A low profile premounted coronary stent (outer diameter < 4Fr, length 18–21 mm, diameter 3.0–4.0 mm) was positioned within the duct, not protected by a sheath; care was taken to cover the complete length of the duct from the aorta-ductal junction until well within the pulmonary trunk.
Results: All stents could safely be deployed with adequate pulmonary flow in early and medium term follow-up. There were no procedure related complications; 1 patient died early from sepsis. All patients had adequate relief of cyanosis for at least 3–4 months. During follow-up the pulmonary vasculature bed growed without distortion. Acute occlusion of a stented duct was not observed. Ductal flow progressively decreased slowly over several months by luminal narrowing (intimal proliferation or peal formation), until the stented duct either had become redundant, was dilated/restented, or elective staged surgery was performed.
Conclusions: With current technology, complete stenting of a short straight duct is a safe and effective palliation, allowing adequate growth of the pulmonary arteries.

40
Evolving use of stent implantation for preserving patency of the arterial duct in cyanotic neonates

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Stent implantation into the arterial duct (ST-AD) may avoid systemic to pulmonary artery shunt operations duct-dependent cyanotic heart disease. We have compared the approach and outcome of ST-AD in two periods using different stents. Earlier era: Between 1991–1996, we implanted 4–6 mm diameter Tower 1.2–1.6 cm long retrievable stents in 10 patients aged 2–46 days with pulmonary atresia (PAt) in 6, subpulmonary stenosis in 3 and critical pulmonary stenosis (PS) in 1. In 4 pts, prior balloon dilation (BD) of the pulmonary valve was performed (2 after radiofrequency perforation RF. 5 stents had to be snared and removed because of malposition or embolisation: 2 patients remained duct-dependent and required further stents at a second procedure. Aspirin and warfarin were continued after the procedure. 1 pt died suddenly at 24 hours in ventricular fibrillation and 1 pt died at 8 weeks from inadequate pulmonary blood flow. 2 pts required Blalock-Taussig shunts at 1 & 6 weeks. 2 pts with complex PAt had central pulmonary artery reconstruction at 12 & 16 months. 1 pt had TOF repair at 4 months. In 2 pts, no longer duct dependent, the stents occluded at 9 and 25 months after warfarin was discontinued. 1 pt had a Glenn shunt. Current era: Between 1997 and 2002, we used 4–6 mm Corinthian premounted stents in 6 pts with pulmonary valve atresia or critical pulmonary valve stenosis. Three patients underwent RF and 3 had balloon dilation and in 4, the duct was stented at the same procedure. There were no stent related complications in these patients, although 1 patient developed septicaemia and jaundice and recovered over 6 weeks. All the patients remain well palliated with a patent stent except one patient who has needed a Glenn shunt. ST-AD with Corinthian stents is an alternative to neonatal shunt operations particularly when patency is only required for a short period while waiting for improvement in right ventricular compliance in critical PS and PA-IVS.

41
In-vivo results of MRI guided stent placement using patient safe automated catheter visualization and slice tracking with an optically switchable resonant marker

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Purpose: Catheter visualization and automated scan plane positioning are essential to support MR-guided interventions including

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endovascular stent delivery. Currently, active tracking techniques yield patient hazards, whereas passive methods do not deliver co-ordinates as required for fast and precise device localization. Purpose of this study was to test patient safe catheters with optically switchable resonant markers (OSRM). Catheters were assessed for their robustness in automated tracking technique and applicability for endovascular stent placement.
Materials and Methods: Five in-vivo experiments were performed. OSRMs were mounted on the tip of 6F catheters. For stent delivery OSMRMs were positioned distal and proximal to the loaded stent on 6F stent delivery systems. Interventions were performed at a 1.5T system (Intera, Philips). For image-guidance, an interactive real time bFFE sequence was used (15 frames/sec). Catheters were introduced into femoral vein and artery sheaths and guided to the iliac, carotid and pulmonary arteries, where selfexpanding nitinol stents (diameter 8–10 mm) were placed. After intervention position and patency of the stents were evaluated with bFFE and 3D angiogramms. Blood flow volumes through the stents were quantified using VEC MRI.
Results: Slice tracking performed robustly in the abdominal and thoracic vasculature. Pulsatile flow in the cardiac approximately caused considerable signal changes of the blood resulting in artifacts in the tracking projections. Bandpass-filtering improved the quality of the data and enabled slice tracking in most cases. Breathing motion did not influence tracking. Visualization of the OSMRMs was fast and unequivocal. Stent delivery was precisely performed at all interventions. Postinterventional assessment of stent position, visualization of its lumen and assessment of quantitative blood flow through the lumen was feasible in all cases.
Conclusion: Reliable slice tracking can be performed in-vivo with an optically switchable resonant marker. Moreover, the marker makes the tip of catheters and the position of loaded stents in stent delivery systems readily perceptible to the observer. It is expected, that a future version of such catheter hardware will allow MRI controlled intravascular interventions in patients while preserving their safety.

42
Preliminary results of genesis stents in native and post-operative pediatric congenital cardiac lesions: a multi-institutional study

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Background: Intravascular use of stents in pediatric population is limited because most available stents require large sheath size, are rigid, have decreased hoop strength, and cannot be dilated to adult size. Recently a new stent, Genesis XD, became available (Cordis, Sommerville, NJ). Genesis stent is a flexible, low profile stent with minimal foreshortenhing at larger diameter, it's hoop strength similar to Palmaz stent and, it can be dilated to 20 mm. This report describes the results of initial experience wtih the Genesis stent in native and postoperative congenital cardiac lesions.
Methods: Ninety-three (pts) underwent placement of 113 stents. The patients were divided into 4 subgroups according to the location of the lesion. Fifty patients had 66 stents placed in branch pulmonary arteries (BPA), (median weight 18 kg). Twenty-seven stents were placed in coarctation of aorta (COA); 7 patients had 9 stents placed in the right ventricular to pulmonary artery conduits; and 8 pts had 10 stents placed in Fontatn circulation. Twenty-six stents were placed in native and 87 stents in post-operative lesions.
Results: All targeted lesions were successfully dilated using sheath size that was 1 to 2 French smaller than the sheath size recommended for the Palmaz stent. In the BPA group, the mean pulmonary artery diameter increased from 4.7 to 9.4 mm ($p < 0.001$), in the COA group, the mean diameter increased from 7.5 to 12.7 mm ($P < 0.001$), in the conduit group the diameter increased from 7.3 to 12.9 mm ($P = 005$) and in the Fontan group the diameter increased from 6.6 to 12.2 mm ($P = 0.006$). Stent embolization requiring a second stent placement occurred in 4 patients. There were no other complications.
Conclusions: The initial experience with Genesis XD stents suggests that these stents are ideal for post-operative and native pediatric congenital cardiac lesions. Small delivery sheath size and flexibility of these stents allows easy approach to smaller vessels with tortuous pathways. The ability to ultimately dilate the stents to adult size is advantageous in rapidly growing pediatric patient.

43
Endovascular stents for coarctation of the aorta: initial results and intermediate-term follow-up

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Forty-eight patients were treated with stent implantation for different pathologies of the aorta. The purpose of the study was to assess the value of balloon expandable intravascular stents in transcatheter treatment of native and recurrent aortic coarctation. Between 1996 and 2002, 31 patients, 19 with native and 12 with recoarctation after various types of surgical repairs were successfully treated with Palmaz (25) and CP (6) stents in three Polish cardiac centers. Mean age was 14,3 ± 0,7 years (range 5–52 yrs). In 12 pts aortic coarctation was diagnosed incidentally. Mean stent to narrowest aortic diameter ratio was 3,0 ± 0,21. After stent implantation peak systolic pressure gradient decreased from 41 ± 0,7 mmHg to 7,6 ± 5,66 mmHg ($p < 0.001$), narrowest diameter increased from 6 ± 2,12 mmHg to 15,6 ± 4,24 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 ± 38,28 mths (range: 2–84 mths). Six patients underwent 9 successful stent re-dilations 3 -54 months after initial implantation. In 1 pt CP stent fracture was detected 16 mths after implantation with no clinical significance. Mean systolic blood pressure decreased from 147 ± 7,07 mmHg before stent placement to 123 ± 3,3 mmHg at follow-up ($p < 0.001$). Thirteen patients (42%) require pharmacological blood pressure control. CT angiography performed in 17 pts showed normal location of stents with no evidence of aneurysm, in 3 pts neointimal hyperplasia was documented during follow-up (successful balloon redilation).
Conclusions: Balloon expandable intravascular stents placement for native and recurrent aortic coarctation has good results in the short and intermediate terms. Successful redilation of stents implanted to aorta is possible. Technical complications during stent implantation can occur. Long-term outcome has to be evaluated for determining the role of transcatheter stent therapy for native and recurrent coarctation of aorta.

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44

Middle aortic syndrome in children – results of interventional treatment (single centre experience)

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The purpose of the study is to evaluate results of percutaneous treatment in middle aortic syndrome. Twelve patients aged 3–17 yrs (mean – 10,6) underwent interventional treatment (8 had stent implantation (S) and 3 balloon angioplasty (BA), 1 had BA followed by S). 1pt had previous correction of ToF, 1pt (ToF + PA) was after BT shunt (angiography in neonatal period – normal aorta, BA for middle aortic syndrome at the age of 3 yrs). All pts had narrowing of aorta (length 3,5–9 cm, minimal diameter 1,5–5 mm). Pressure gradient ranged between 40 and 90 mmHg (mean-57). 9pts underwent S (1pt – previous BA of aorta and renal arteries followed by renal arteries transplantation, 1pt after renal arteries transplantation). During the procedure 1pt had additional BA of renal arteries and 1pt stent to truncus coeliacus. 11Palmaz and 1CP stents were expanded with 7–14 mm balloons. Pressure gradient after implantation was 0–45 mmHg (mean – 15,0). Heparin (48 hours), aspirin and acenocumarol (3–6 mths) were administered after the procedure. In 1pt thrombosis of the stent occurred 6 days after implantation, successfully treated with local rt-PA infusion followed by BA and second stent implantation. In 3–99 mths (mean – 55,8) follow-up 3pts had elective stent redilation (after 4, 6 and 89 mths), 3pts underwent 4 successful redilations due to neointimal hyperplasia (after 9, 12, 19, and 42 mths). 1pt had second stent implantation 6 mths after procedure due to aneurysm formation. BA was performed in 4pts. In 3pts BA of aorta and renal arteries enabled pharmacological control of severe arterial hypertension (in 1pt BA was followed by transplantation of renal arteries and stent implantation to aorta). In 3 yrs old pt with PA + VSD, BA of aorta was performed and repeated after 4 mths, and gradient decreased from 50 to 15 mmHg, minimal diameter increased from 1,5 to 5 mm. Stent implantation produces satisfactory early and long-term results. Neointimal hyperplasia causing arterial hypertension can develop and can be successfully treated with repeat balloon dilation. In complex middle aortic syndrome, BA can improve pharmacological blood pressure control but long term result is uncertain, some pts require complex treatment – intervention/surgery. In young patients BA can be the first step followed by stent implantation.

45

Rapid right ventricular pacing is an alternative to adenosine in catheter interventional procedures for congenital heart disease

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Introduction: Adenosine has been shown to be safe and effective in creating transient cardiac standstill and is used during balloon dilatation procedures and stent implantations. However, there are some disadvantages such as a wide variation of the effective adenosine dose and significant variations of the time intervals from adenosine injection to asystole and duration of asystole. Ventricular extrasystoles triggered by the interventional procedure are not suppressed. We describe a different approach to create transient low cardiac output and prevent unwanted device movement by rapid right ventricular pacing.

Methods: This was a prospective pilot study. The method was used in patients with congenital aortic stenosis elected for aortic valve balloon dilation in whom the first dilation maneuvre failed due to slipping of the balloon through the valve. Balloon dilation was then repeated during rapid right ventricular pacing at a rate of 220 beats per minute.
Results: From 09/2001 till 12/2002 there were 22 patients (15 infants and 7 children) who were elected for aortic valve balloon dilation. The procedure was feasible in all. The balloon stayed in place in all but one infant but moved out of the valve in six of seven children. Therefore, rapid pacing was performed in seven patients. The duration of the maneuvre was less than 16 seconds and the balloon maintained a stable position during the procedure in all of them. There were no arrhythmias after cessation of ventricular stimulation. There were no other procedure related complications.
Conclusion: Rapid ventricular pacing is a safe and effective method for creating transient low cardiac output during balloon dilation of the aortic valve. This achieves stability of the dilation balloon which is likely to improve results and reduce complications. It may have applications in other fields of catheter intervention where low cardiac output is desirable to maintain stable positions during the critical phase of the procedure.

Session 6: Surgery

46

Risk factors for postoperative morbidity and length of stay in 99 consecutive patients following Stage I Norwood reconstruction

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Introduction: Stage I Norwood reconstruction is performed for a heterogeneous group of defects including single ventricle with obstruction to systemic blood flow. The Norwood procedure is associated with improving operative mortality but significant perioperative morbidity. Risk factors for mortality have been shown to include weight <2.5 kg and additional major cardiac or non-cardiac anomalies.
Purpose: To review the course in the cardiac intensive care unit (CICU) to determine the incidence of major postoperative morbidity, and to determine risk factors for prolonged postoperative length of stay (LOS).
Methods: The medical records of all patients born between 09/2000–09/2002 who underwent stage I reconstruction were retrospectively reviewed for details of the perioperative course. Risk factors for LOS analyzed by univariate and multivariate analyses.
Results: Ninety-nine patients met inclusion criteria. Hospital mortality was 12.1%; 5.7% in "standard-risk" (n = 53) and 19.6% in "high-risk" (n = 46) patients (p = 0.061). Delayed sternal closure was performed in 18 patients, at a median of 1.5 (0–5) days until closure. Other reoperations and significant postoperative morbidity are listed in the table (excluding cannulation for extracorporeal membrane oxygenation [ECMO]). Excluding deaths, the median total duration of mechanical ventilation was 3.8 days (range 1 day–4.2 months). Reintubation was performed in 34 patients, in 13 more than once; the only risk factor identified was diaphragmatic paresis. Median postoperative LOS in the CICU was 11 days (range 1 day–4.2 months); excluding deaths it was 10 days (4 days–4.2 months). Lower weight at surgery, anatomic diagnosis and "high-risk" status were not related to postoperative LOS. Risk factors for a prolonged postoperative LOS included longer aortic cross clamp time (p = 0.035), postoperative reoperation (p = 0.017)

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or reintubation (p = 0.002), renal dysfunction (p = 0.037) and greater fluid retention in the first 48 hours after surgery (p = 0.042). Nasogastric feeding was necessary at discharge from the CICU in 40/87 (46%) hospital survivors.

Reoperations and Postoperative Morbidity	
Reoperations	
Exploration for bleeding	20
Diaphragmatic plication	6
Shunt revision	4
Wound debridement	4
Thoracic Duct Ligation	2
Other	4
Morbidity	
Postoperative Catheterization	17
Cardiopulmonary Resuscitation	14
Renal Dysfunction (Creatinine < 1.5)	13
Seizures	13
Thrombocytopenia (<20,000)	11
Vocal Cord Injury	8
ECMO	7

Conclusions: Mortality rates for stage I reconstruction continue to improve and approach 5% in standard risk patients. However, there is significant morbidity and prolonged LOS in many patients, associated with considerable resource utilization. Further studies are underway to determine the long-term implications of the post-operative morbidity in hospital survivors.

47
Early primary repair of Tetralogy of Fallot: low prevalence of early arrhythmias

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Background: Late repair of Tetralogy of Fallot (TOF) has been associated with significant early and late arrhythmias. Suspected possible mechanisms are prolonged hypertrophy and cyanosis as well as extensive muscle resection in the right ventricular outflow tract (RVOT). Early primary repair of TOF leads to normal RV pressures and saturations and limits the necessity of RVOT resection. The purpose of this study was to assess the prevalence of ECG changes and arrhythmias early after early primary repair of TOF.
Patients and Methods: Between 1995 and 2001 54 consecutive unselected patients with TOF (8 with pulmonary atresia) underwent early primary repair at our institution. A transannular patch was placed in 39 patients (resection of muscle in the RVOT was performed in only 4 of them), 4 patients had an outflowtract patch and in 8 patients a homograft was implanted. Median age at operation was 42 days (3–111 days), median weight was 3,5 kg (2–6,6 kg) Charts, ECGs and Holter recordings were retrospectively reviewed for residual hemodynamic problems, electrophysiologic abnormalities or presence of arrhythmias.
Results: Median follow-up was 27 months (6–84) months. There was 1 early death of a patient with pulmonary atresia (1,8%) and no late death. ECG was normal in 22/53 (42%), complete right bundle branch block was present in 26/53 (49%), signs of RV hypertrophy in 11/53 (21%) and premature atrial contractions (PACs) in 1/53 (2%). Holter recordings were available in 46 patients and were normal in 38/46 (83%). Premature ventricular contractions were present in 4 patients, intermittent junctional rhythm in 2 and frequent PACs, ectopic atrial rhythm and recurrent SVT in 1 patient respectively.
Conclusions: Early primary repair of TOF can be performed with low mortality and morbidity. It is associated with a low prevalence of early arrhythmias and a relatively high prevalence of normal ECG and Holter recordings. These findings give hope that early repair might be able to decrease the prevalence of late arrhythmias.

48
Ventricular assist devices in children

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Ventricular assist devices are successfully used in the treatment of end-stage cardiac failure in the pediatric population. To evaluate early and late results of the implantation of ventricular assist device systems in children as bridge to recovery or bridge to transplant we reviewed all pediatric patients supported at the author's institution. Assist devices were implanted in 23 patients (age 7 ± 7.8 years, weight 22.5 ± 37.2 kg, BSA 0.84 ± 0.72 m^2, 15 male, 8 female). Fourteen pts (age 3.5 ± 6.7 years, weight 12.9 ± 19.5 kg, BSA 0,55 ± 0,63 m^2) were supported with ECMO, 4 pts (age 7.5 ± 5.9 years, weight 26 ± 19.1 kg, BSA 0.67 ± 0.28 m^2) with Medos, 2 pts (age 13.7 ± 6 years, weight 50.5 ± 31.8 kg, BSA 1.5 ± 0.62 m^2) with Berlin Heart and 3 pts (age 17.8 ± 1.74 years, weight 65.5 ± 2.1 kg, BSA 1.86 ± 0.12 m^2) with Novacor. Diagnosis were in 14 pts congenital heart diseases (TGA, DORV, mitral or aortic valve stenosis, TOF, AVSD, univentricular heart syndrome) and in 9 pts acquired heart disease (DCM, Kawasaki-Disease, graft failure after heart transplantation). The indication for implantation of an ECMO was postcardiotomy heart failure in 13 of 14 pts, the indication for the use of a Medos, Berlin Heart and Novacor assist device was bridging to transplant in DCM in all pts. Patients were supported for 3.15 ± 1.2 days (ECMO), 15 ± 6.9 days (Medos), 45 ± 31.7 days (Berlin Heart) and 8 ± 11.8 days (Novacor). The most common complication was bleeding, necessitating rethoracotomy in 12 pts (52%), 2 pts with Medos assist device showed thromboembolic complications. Myocardial recovery was observed in 43% (6/14) of the ECMO pts. Overall, 14 pts were successfully weaned from the device. Eight pts underwent transplantation: 14% of the ECMO patients (2/14), 50% of the Medos pts. (2/4), all Berlin Heart pts. (2/2) and 67% of the Novacor pts. (2/3). Eleven pts (48%) were discharged and are long-term survivors (ECMO 43% (6/14), Medos 50% (2/4), Berlin Heart 100% (2/2) and Novacor 33% (1/3)). These results demonstrate the efficacy of the implantation of ECMO or other VAD systems in children with cardiac disease as bridge to recovery or bridge to transplant.

49
Heart transplantation in children following mechanical circulatory support with pulsatile pneumatic assist device

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Background: Mechanical support with a pulsatile pneumatic ventricular assist device is a complex rescue procedure performed in children with untreatable cardiogenic shock. Its impact on early and long-term survival after subsequent heart transplantation remains to be determined.
Methods: We reviewed retrospectively the course of 95 children (median age 8 years, range 8 days–17 years, body weight 24 kg, range 3–110 kg) with heart transplantation. The elective-HTx group (A) consists of 33 children who were treated as outpatients before transplantation. The emergency-HTx group (B) comprises

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44 children who were critically ill and in hospital before transplantation but without a ventricular assist device, whereas the VAD-HTx group (C) consists of 18 children resuscitated and supported with a pulsatile pneumatic ventricular assist device for a median time of 20 days.
Results: Overall actuarial survival after cardiac transplantation was 86% at 1 month, 82% at 1 year, and 78% at 5 years without significant differences between the three subgroups. Group A had the best long-term survival rate with 88/88/80%, B had a survival rate of 88/82/79% and C 72/72/72%. There were no differences in neurological outcome, acute cardiac rejections or transplant failure. The survival rate was significantly better in the children with cardiomyopathy compared to those with congenital heart defects ($p = 0.014$).
Conclusions: Bridging to heart transplantation by pulsatile pneumatic assist device is a safe procedure in pediatric patients. After heart transplantation overall survival of these children is almost similar to that of patients who were bridged with inotropes, or were electively awaiting heart transplantation.

50
Influence of balanced ultrafiltration on plasma and urinary cytokine homeostasis and renal dysfunction during cardiac surgery in newborns and infants

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Objective: To assess the impact of balanced ultrafiltration on plasma and urinary cytokines and renal dysfunction during cardiac surgery in newborns and infants.
Methods: Twelve patients were included into this study (age: 3–214, median 13 d). Repeated analyses (before CPB, at the end of CPB, and 6 h after the end of CPB) of IL6 and IL8 were performed in plasma, urine and ultrafiltrate during Norwood I Op (2x) or repair of TGA (3x), AVSD (2x), VSD, hypolastic aortic arch + VSD, TAPVD, TAC, ALCAPA. Urine measurements of albumin, transferrin, IgG, a1-microglobulin, or NAG were used to assess renal dysfunction. After the end of the aortic clamp a balanced ultrafiltration of 40–360, median 180 $mL\,kg^{-1}$ was performed during the CPB.
Results: Plasma IL6 and IL8 were elevated at the end of CPB to 93 ± 24 and $68 \pm 15\,pg\,mL^{-1}$, respectively. Renal excretion of the cytokines was low during CPB and the following 6 hours (IL6: 36 ± 9, IL8: $71 \pm 23\,pg\,kg^{-1}$). Ultrafiltration removes $3200 \pm 1100\,pg\,kg^{-1}$ of IL6, and $6700 \pm 2500\,pg\,kg^{-1}$ of IL8 from the blood circuit, which correspond approximately to 20% of the IL6 and 50% of the IL8 remaining in the plasma at the end of CPB. There was a negative correlation ($r > 0.5, p < 0.05$) between plasma cytokine values and renal cytokine clearances, while renal cytokine excretion was dependent on the renal clearance function. Urinary albumin, transferrin, IgG, and a1-microglobulin concentrations rose continously up to 6 hours after the end of CPB. Elevated plasma cytokine concentrations and abased renal cytokine clearances were correlated with the elevation of urinary proteins.
Conclusions: Balanced ultrafiltration is highly effective in removing pro-inflammatory cytokines from the blood circuit. The correlation of elevated plasma cytokines with reduced renal cytokine clearance and elevated urinary proteins rises the hypothesis, that pro-inflammatory cytokines damage the kidney itself and thus contribute to postoperative renal dysfunction. Thus, balanced ultrafiltration on CPB should act protectively on the kidney.

51
Hypoplastic aortic arch in newborns rapidly adapts to postcoarctectomy circulatory conditions

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Background: Transverse arch hypoplasia is a part of neonatal aortic coarctation with coexisting intracardiac left-to-right shunt situations and/or complex anomalies. Resection of all ductal tissue and extended end-to-end anastomosis has been advocated with sliding up descending aorta to the underside of the transverse arch. Most centers accept less aggressive approach when arch hypoplasia is moderate. This study focuses on how quickly hypoplastic transverse arch adapts to postcoarctectomy circulatory conditions.
Patients/methods: Thirty-four consecutive neonates (mean age: 13.1 ± 6.19 days, mean weight: 2.8 ± 0.53 kg) with primary pathology of aortic coarctation were assessed by echocardiography then underwent resection and end-to-end anastomosis. Patients were grouped into normal and hypoplastic (defined as arch diameter just distal to left carotid artery/descending aorta ratio <0.5) groups. Detailed echocardiographies were performed on first, third and fifth postoperative days. Ascending and descending aorta flow velocities (AAFV and DAFV) were plotted against left ventricle shortening fraction (SF) to make comparisons.
Results: Normal ($n = 20$, arch ratio: 0.88 ± 0.14) and hypoplastic arch ($n = 14$, arch ratio: 0.35 ± 0.06, $p < 0.001$) groups did not differ in demographics. Coexisting anomalies were more frequent with hypoplastic arch (7/20 vs. 10/14, $p = 0.03$). No morbidity/mortality but one early reoperation for residual coarctation and another balloon aortovalvuloplasty for valvar aortic stenosis occurred. No postoperative radial-to-femoral mean gradients more than 10 mmHg were observed.

		P/Oday1	P/Oday3	P/Oday5
Normal	SF	0.39 ± 0.08*	0.41 ± 0.11	0.45 ± 0.06
	AAFV	1.26 ± 0.22	1.8 ± 0.4	1.92 ± 0.67
	DAFV	2.24 ± 0.41	2.44 ± 0.25	2.48 ± 0.5
Hypopl	SF	0.32 ± 0.07*	0.4 ± 0.09	0.46 ± 0.11
	AAFV	1.23 ± 0.24	1.57 ± 0.44	1.59 ± 0.38
	DAFV	2.73 ± 0.33	2.55 ± 0.35	2.49 ± 0.29

*$p = 0.01$

Conclusions: Although surgery did not address directly tranverse arch hypoplasia no statistically significant AAFV and/or DAFV difference was noted between normal and hypoplastic arch groups. DAFV is result of different parameters e.g. vessel wall elasticity, diameters and cardiac output, etc. SF was used as surrogate of cardiac output to compare velocity change in postcoarctectomy circulatory conditions. A logarithmic correlation was observed between SF, AAFV and DAFV suggesting that the transverse arch is complient to entertain increasing cardiac output. We conclude that arch remodelling is fairly rapid, enlargement of the hypoplastic aortic arch is not usually necessary.

52
Self-expandable valved stent: off-bypass implant in pulmonary position

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Objective: Percutaneous pulmonary valve implantation has been introduced in the clinical practice with the following limits: (a) applicability limited to patients >20 kg of body weight; (b) largest

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implantable valve of 18 mm; (c) need for balloon dilatation of the valved stent. We evaluated the off-bypass implantation of a self-expandable valved stent of unlimited size in pulmonary position.
Methods: A glutaraldehyde preserved valved bovine jugular xenograft, mounted in two rings of non-thermosensitive nitinol "Z" stent, self-expandable from 7 to 24 mm of internal diameter, prepared with a Teflon sheath stent-graft delivery system, was acutely evaluated in 6 adult pigs, mean body weight 55.6 kg (range 47 to 67 kg). Through a stent-graft delivery system (overall diameter 8.0 mm = 24F) the self-expandable valved stent was implanted off-bypass in pulmonary valve position by transventricular approach through median sternotomy, under guidance by intra-vascular ultrasound.
Results: The mean diameter of the main pulmonary artery measured with intra-vascular ultrasound was 21.7 ± 1.6 mm. The mean length of the self expandable valved stent was 23.1 ± 0.7 mm, the mean internal diameter 21.6 ± 0.7 mm and the mean external diameter 26.3 ± 0.7 mm. The mean peak pressure gradient recorded across the valve was 6.33 ± 2.8 mmHg (range 4.5 to 9.6 mmHg) at Doppler echocardiography, and 4.5 ± 3.1 mmHg (range 0–7 mmHg) at invasive measurement, with a pulmonary blood flow of 3.03 ± 0.05 L/min. Intra-vascular ultrasound showed complete opening and closure of the valve (mean area reduction from 315.08 ± 54.13 to 0 mm^2). In all animals echocardiography confirmed the absence of any valve regurgitation as well as of paravalvular leak. Post-mortem examination confirmed the adequate position of the valved stent in pulmonary position, as well as ruled out any valve deformation or thrombus.
Conclusions: (a) off-bypass implantation of self-expandable valved stent is feasible in pulmonary position with minimally invasive approach; (b) ballon dilatation of the implanted valved stent is not required; (c) off-bypass surgical approach allows for valved stent implantation of any size, including the adult size with adequate hemodynamic functioning; (d) intra-vascular ultrasound makes implantation and evaluation easy and reproducible.

53
Early results of the bovine jugular vein for right ventricular outflow tract reconstruction

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Objective: To study the early function of bovine jugular vein (BJV) when used for right ventricular outflow tract (RVOT) reconstruction.
Materials and Methods: 62 consecutive patients operated at University Hospital Zurich from Jan 2001 to Dec 2002 (median age: 5 yrs 5 months, range 3 days–39 years), who received a BJV graft for RVOT reconstruction. The procedures included Fallot's Tetraology & Pulmonary atresia(32), Ross Procedure(15), Correction of Truncus arteriosus(6), Rastelli Procedure(4) and Others(5). 23 of these procedures were reoperations and 25 needed branch pulmonary artery reconstruction. The size of BJV ranged from 8–22 mm (median 16 mm). The median CPB time and X clamp time were 210 and 83 minutes respectively. Patients were followed up with regular echocardiographic control. Median followup was complete at 6.5 months.
Results: Three patients died of myocardial failure in early postoperative period. Mean Gradient across the BJV graft was less than 10 mm in 50 patients and a median of 23 mm in the remaining patients. Two BJV grafts needed dilatation and stenting while two patients needed graft replacement for an anastomotic site stenosis caused by a fibrotic membrane. None of the grafts had more than mild insufficiency and this remained so during followup.
Conclusion: BJV when used for RVOT reconstruction showed excellent function in the early phase. However fibrosis leading to anastomotic site stenosis should be watched for. The large size range and off the shelf availability of this graft are particularly attractive.

54
Performance of Contegra bovine valved jugular vein conduits for pediatric RVOT-reconstruction – 3.5 years experience with 108 implantations

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Background: Paediatric RVOT-Reconstruction with Contegra, a bovine jugular vein graft, is available for more than 3.5 years. Mean term durability and compatibility is investigated after this interval; durability is compared to alternative RVOT grafts.
Patients and Methods: Within an FDA-controlled study, we implanted 108 Contegra Pulmonary Valved Conduits (PVC) from 5/1999 to 1/2003 in 104 infants (m/f 47/57) aged 2 days–20.1 years, median 1.6 years. 33 were primary repairs, 37 had previous graft implantations, 38 other repairs/palliations. Preoperative diagnoses: TAC (25 patients), TOF (48), DORV (14), TGA (7), 2 rare complex malformations, 1 Ross procedure. Echocardiography is performed at 1, then every 3rd month postoperatively. Total follow-up: 189 years. Results were compared to our 52 homograft- and 30 porcine xenograft recipients.
Results: PVC tissue is very apt for suturing. There were 8 deaths (7 early, 1 late). PVC insufficiencies (trace or mild) are found in 50%, without clinical significance or increase tendency. Redos: 6 for peripheral pulmonary arteries (pPA), 4 explants for pPA stenoses; 3 of them involved the peripheral anastomosis. One mild non stenotic calcification after 3.5 years was observed. We saw no relevant conduit dilatation, one valve degeneration. Transvalvular peak gradients remained below 25 mmHg, average <10 mmHg. Freedom from explantation at 3 years is 96%, and 90% at 3.5 years. Contegra patients performed slightly better than homograft patients concerning survival and freedom from explantation; they were significantly better than xenografts.
Conclusion: After pediatric RVOT reconstruction, Contegra PVCs show no relevant dilatation or hemolysis signs. After 3.5 years, its durability is superior to porcine xenografts and seems at least equivalent to homografts. At least until up to 3.5 years, they can be considered a serious alternative to homografts.

Session 7: Imaging

55
Non-invasive diagnosis of congenital vascular anomalies in newborns and infants: multi-slice CT-angiography and partially parallel 3D-MR-angiography

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Introduction: Conventional catheter angiography (CA) remains the standard procedure for the diagnosis of congenital vascular anomalies (VA). Aim of the study was to assess the value of the cardiac-gated multi-slice CT angiography (MS-CTA) and of partially parallel

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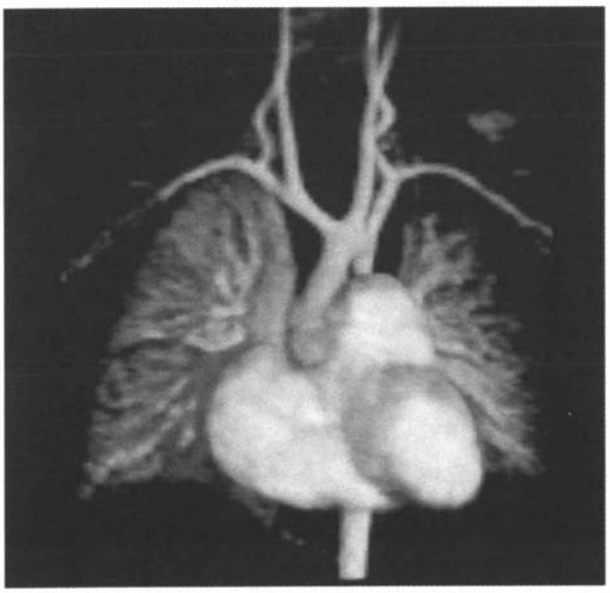
3D-MR-angiography (3D-MRA) and for preoperative evaluation of VA in newborns and infants in comparison to CA.
Method and Materials: Thirty newborns and infants with severe VA were evaluated by MS-CTA (n = 22) or SE-MRA (n = 8) and as well as CA. VA included pulmonary atresia (n = 5), double aortic arch (n = 3), interrupted/hypoplastic aortic arch (n = 4), aortic coarctation (n = 7), and to verify a vascular ring or sling (bronchoscopy revealed stenosis, n = 11). CTA was performed with a four-slice helical CT (scan-time = 15–30 s in apnea) using retrospective ECG-gating. MRA imaging protocol included ultra fast time-resolved 3D-MRA with partially parallel imaging techniques (scan time = 2 s) which was performed during free breathing. Image data were post-processed with volume-rendering. The extent and the type of VA as well as their relationship to esophagus, trachea or bronchi was assessed and correlated to CA.
Results: High quality MS-CTA and 3D-MRA data both were almost free of cardiac and respiratory motion could be acquired in all patients. In all cases, the extent and type of VA could be detected. Due to the thin collimation, the volume-rendered models provided excellent visualization of the vessels (graphic). Due to the tissue contrast the relationship between the VA and surrounding structures was substantially better appreciated than on CA images. In 8/30 patients a vascular ring or sling could be excluded non-invasively. In 7/30 MS-CTA and SE-MRA were the only diagnostic tool for surgical planning. In these 15/30 no further CA was necessary.
Conclusions: MS-CTA and 3D-MRA are non-invasive, accurate and robust techniques for detecting VA appears equivalent to CA while they are more accurate in delineating potential life-threatening complications such as compression of the surrounding structures. At the moment there is a small advantage of MS-CTA because of the better spatial resolution (1,25 mm vs. 2 mm). In near future 3D-MRA may becomes the modality of choice as a non-ionizing and free breathing technique for the diagnosis of VA.



56
Pulmonary blood flow pattern in patients with Fontan circulation depicted by cine MRI

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Background: After palliation of univentricular heart with a Fontan modification there is no subpulmonary ventricle modifying pulmonary blood flow. The aim of the study was to test the influence of cardiac cycle on pulmonary blood flow pattern in those patients and to compare it with that in normal subjects.
Patients and Methods: Blood flow pattern was investigated with cine MRI in the left pulmonary artery of 18 patients (20.6 ± 7.4 years old, 7 females) with Fontan circulation (4 RA-RV connections, 9 RA-PA connections, 5 TCPC) and compared with that in the superior caval vein and main pulmonary artery of 12 healthy volunteers (26.3 ± 6.0 years old, 10 females). Measurements were sampled over a period of about 2 minutes to distinguish respiratory effects. Blood flow pattern was depicted by interpolating the variable number of measured phases to 100 phases and normalizing flow to mean blood flow in that vessel. Then, average flow pattern could be calculated and compared between the studied groups.
Results: Fontan patients had an almost constant flow pattern in their left pulmonary artery. There was only a slight flow acceleration at the end of diastole and, depending on the type of Fontan modification, an even minor one in systole. This blood flow pattern did not resemble that in the caval vein of healthy volunteers, where a clear systolic flow peak and an only small flow acceleration in the beginning of the diastole was depicted. Nor it resembled that in the normal pulmonary artery, where blood flow is almost confined to a single systolic peak.
Conclusion: Pulmonary blood flow pattern in patients with Fontan circulation is almost independent from cardiac cycle. Further studies should focus on the influence of respiration on pulmonary blood flow.

57
Hearts late after Fontan – operation have normal mass, normal volume and reduced systolic function – a magnetic resonance imaging study

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Aims: To assess ventricular mass, volume and systolic function in patients late after Fontan – operation by cardiac magnetic resonance imaging.
Methods and Results: Twenty-four unselected patients (female 9, male 15) at a mean age of 19.1 ± 6.2 years, 10.2 ± 4.1 years after Fontan – operation were studied. A standard 1.5 T scanner (Gyroscan ACS NT, Philips) was used and analysis was performed using dedicated software (MASS®; Medis, Leiden, the Netherlands). 10 healthy volunteers (age 28 ± 6.3 years) served as control group. Mean endsystolic mass index was 74.7 ± 23.7 g/m^2 and 86.6 ± 18.0 g/m^2 in the controls (n.s.). Mean enddiastolic ventricular volume was 63.2 ± 18.7 ml/m^2 compared to 67.4 ± 4.7 ml/m^2 in the control group (n.s.). Mean ejection fraction was 47.5 ± 11.1% compared to 62.7 ± 18.8% in normals ($p < 0.01$).
Conclusions: We conclude that hearts late after Fontan – operation have normal mass, normal volume and reduced systolic function. Our findings do not support the discussion that decreased ventricular function in patients later after Fontan – operation is due to increased ventricular mass and decreased diastolic function.

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58
Myocardial perfusion of the anatomic right systemic ventricle in patients with congenitally corrected transposition

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Objectives: Myocardial blood flow (MBF) in unoperated patients with congenitally corrected transposition of the great arteries (CCTGA) was quantitatively investigated by positron emission tomography (PET).
Background: In CCTGA the systemic ventricle is of a right morphology and patients are at high risk of developing systemic ventricular dysfunction; inadaequate myocardial perfusion of the hypertrophied systemic ventricle may cause or accelerate a decline in ventricular function.
Methods: Fifteen patients with CCTGA were investigated by PET with nitrogen-13 ammonia at rest and during adenosine vasodilatation. A subgroup of 7 patients had isolated CCTGA (group A: 30.3 SD 11.9 years), while the remaining 8 patients had complex CCTGA associated with subpulmonary stenosis in all, and ventricular septal defect in four (group B: 30.6 SD 16.4 years). Eleven healthy adults (26.2 SD 5.1 years) served as the control group.
Results: Resting MBF was not different between both groups of patients with CCTGA and healthy young adults. Hyperemic blood flows were significantly lower in both groups of CCTGA compared to normals (195 SD 21 ml/100 g/min. for group A, 201 SD 27 for group B, 258 SD 22 for normals; $p < 0.001$); thus, coronary flow reserve was significantly lower in both groups of CCTGA compared to healthy adults (2.5 SD 0.28 for group A, 2.6 SD 0.48 for group B, 4.0 SD 0.43 for normals; $p < 0.001$).
Conclusion: Blood flow measurements suggest decreased coronary reserve in the absence of ischemic symptoms in patients with isolated and complex CCTGA. The global impairment of stress flow dynamics may indicate altered global vasoreactivity and quantitative changes in microcirculation suggest an important role in the pathogenesis of sytemic RV dysfunction.

59
The systemic right ventricle in patients after atrial switch repair for TGA compared to congenitally corrected TGA (ccTGA) – a study by magnetic resonance imaging (MRI)

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Objective: Dimesions and right ventricular systolic function were assessed in patients after atrial switch repair for TGA using MRI. Data were compared to corresponding values in patients with ccTGA and to those with normal left ventricles (LV).
Patients and Methods: 21 patients after atrial switch repair ($n = 11$, mean age 21,2 ± 4,3 yrs) or with ccTGA ($n = 10$, mean age 33,6 ± 16,9 yrs.) without associated lesions were studied by MRI. Enddiastolic and endsystolic volumes, stroke volume, ejection fraction (EF) and myocardial mass of the systemic right ventricle were assessed. Volume calculations were performed using short axis cuts of multiphased, balanced FFE sequences (Philips Gyroscan NT 1,5 T). Data were compared to corresponding values of the LV in 10 healthy volunteers (mean age 28,0 ± 6,3 yrs.). The presence of myocardial scars was tested by MR late enhancement technique after injecting 0.2 mmol Gadolinium DTPA i.v.
Results: In none of the patients a late enhancement signal could be detected within the myocardium.

Systemic	Ventricle	Atr. switch	ccTGA	Control
Enddiast.	Volume (ml)	188,1 ± 88*	188,5 ± 66*	128,2 ± 13
EF (%)		52,6 ± 14,8	47, ± 13,4*	62,7 ± 8,1
Stroke	Volume (ml)	89,1 ± 19,7	88,0 ± 34,0	80,3 ± 12,2
Myocardial	Mass (g)	201,5 ± 71*	162,8 ± 44,0	129,9 ± 32

*$p < 0.05$, compared to control

Conclusion: Patients after atrial switch operation or with ccTGA show systemic right ventricular dimensions that are larger and myocardial mass which is strongly increased compared to those in normal LV. In these patients systemic RV EF is reduced under rest, compared to that in normal LV, whereas stroke volume is preserved. By late enhancement technique, there is no evidence for myocardial scarring in these patients.

60
Regional left ventricular function in patients with ALCAPA late post repair: an ultrasound-based regional strain and strain rate imaging study

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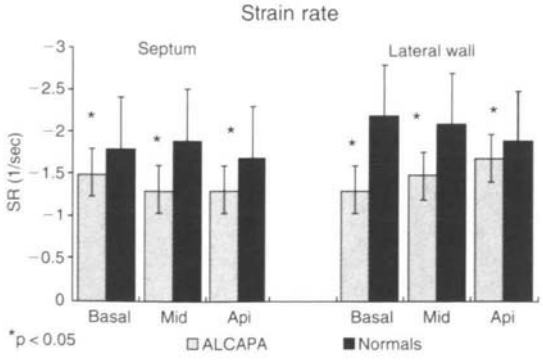
Background: After coronary reimplantation for ALCAPA recovery of function is described. Few data are available on residual regional dysfunction in these patients.
Aim: To evaluate right ventricular (RV) and LV longitudinal and radial deformation in 13 ALCAPA pts late after repair (>1year) by ultrasound-based Strain (S) (%) and Strain Rate (SR) (1/sec) imaging and to compare these data with normal controls.
Results: Patients were comparable to controls for LV and RV dimensions and for LV fractional shortening. There was a significant reduction in atrio-ventricular ring displacement for both the lateral and the interventricular septum observed in these patients ($p < 0.001$), while the tricuspid annulus at the RV free wall was similar to normals. Radial function in ALCAPA patients was normal as assessed by S/SR imaging (Patients: S = 50 ± 12; SR = 3.4 ± 1.6 vs Normals: S = 55 ± 12; SR = 3.4 ± 1.6, p = NS). On the other hand, regional longitudinal function, assessed by S/SR imaging, was significantly reduced in ALCAPA patients. This reduction was homogeneous in each studied wall (Figure 1). RV regional function assessed from RV free wall was similar to that of normals.
Conclusions: Late after apparently successful repair, LV regional normal radial function reduced longitudinal function was observed. This may be related to residual subendocardial fibrosis impairing long axis.



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61
The impact of cardiac MRI in the optimal timing for pulmonary valve replacement in severe pulmonary regurgitation after repair for Tetralogy of Fallot

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Background: Pulmonary regurgitation (PR) is a common residual lesion after surgical repair for Tetralogy of Fallot (TOF) and leads to right ventricular (RV) dilatation, arrhythmias and possibly sudden death. Thus a correct timing for pulmonary valve (PV) replacement is crucial to the management of these patients.
Aim: To evaluate the use of cardiac magnetic resonance imaging (MRI) in the haemodynamic assessment and planning of pulmonary valve replacement in children with repaired TOF.
Methods: Fourteen children with severe PR diagnosed at echocardiography were prospectively assessed by cardiac MRI. Their mean age was 13 ± 2.6 y, mean weight 38.6 ± 13.2 kg. Surgical repair was performed with homograft-monocusp in 7, RVOT patch in 4 and transannular patch in 3 patients. Time since surgical repair was 11.3 ± 2.6 y. Gradient-echo cine sequences were used to calculate the RV enddiastolic volume (RVEDV) and RV ejection fraction (EF). PR and differential lung perfusion were quantified by using phase-contrast sequences. A contrast-enhanced MR angiography was performed for anatomical evaluation of the pulmonary arteries.
Results: MRI confirmed severe PR in all patients, with a mean regurgitation fraction of 50.7 ± 17%. In 13 patients RVEDV measured 182.7 ± 33.8 ml/m^2 and the RV EF 46 ± 5%. No significant correlation was found between regurgitation fraction and RVEDV or RV EF respectively. MR angiography depicted a stenosis of the left pulmonary artery in 4 cases. Phase-contrast measurements demonstrated a decreased left pulmonary blood flow in all 4, with a mean of 28.5 ± 8.5% vs 71.5 ± 8.5% on the right. Applying the proposed cut-off of 150 ml/m^2 for RVEDV, 12 of 14 patients qualified for PV replacement without additional exams, such as invasive catheterization.
Conclusions: Cardiac MRI allows an accurate and non-invasive anatomical and hemodynamical evaluation of PR in children after TOF repair. RV size is the most important parameter for an optimal timing for PV replacement, as in presence of severe PR, regurgitation fraction do not correlate with RVEDV. Thus RVEDV may be influenced by additional factors. Since MRI is the technique of choice for quantification of RV size, assessment with cardiac MRI should be considered in every patient with severe PR.

62
Surgical correction of HOCM in children by a new approach

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Objectives: Treatment of children with HOCM is complicated by several factors including noncompliance medications and an increased risk of sudden death. The classic Morrow technique is not effective for HOCM children with midventricular and RVOT obstruction and extreme left ventricular hypertrophy.
Methods: The presented excision of the asymmetrical hypertrophied area of the interventricular septum causing obstruction of LVOT and RVOT simultaneously and midventricular obstruction is made from conal part of right ventricle transversely and anteriorly of the Lancisi muscle and moderator band but not through the whole thickness of IVS, that is, without penetration into the left ventricular cavity. 7 pediatric patients underwent this procedure. Ages ranged from 10 to 15 years (mean, 12,5). The midventricular obstruction was noted in 3 children, isolated RV obstruction in 1 patient. In 3 operated children the obstruction of LVOT and RVOT was noted simultaneously. The follow-up period was 16 ± 7 months.
Results: The mean echocardiographic intraventricular gradient in LV decreased from 78,9 ± 5,9 to 12,7 ± 5,2 mmHg, the mean value of gradient in RVOT also reduced. In patient with isolated RVOT obstruction gradient decreased from 60 to 8,7 mmHg. Echocardiographically determined septal thickness was reduced 31,7 ± 6,5 versus 16,1 ± 4,6 mm. Follow-up echocardiography showed reduction of left atrial size from 46,7 ± 7,1 to 38,5 ± 6,2 mm. Magnetic resonance imaging showed an increase of the diastolic volume of RV and stroke volume.
Conclusions: This method is a safe and effective technique for surgical treatment of pediatric patients with severe hypertrophic obstructive cardiomyopathy unresponsive to medical management.

63
Non-invasive assessment of stent lumen patency using MRI in swine

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Purpose: Endovascular stents are increasingly employed for the treatment of vascular obstruction. However, the application of MRI for the non-invasive assessment of stent patency is limited because susceptibility artifacts and radio frequency shielding effects can be misinterpreted as stent obstruction. Purpose of this study was to assess weather stent lumen patency of a stent with a new design concept can be determined using MRI.
Materials and Methods: Commercial available selfexpanding stents (diameter 8–10 mm), including Dynerlink (Guidant), Smart (Cordis) and Flexx (Angiomed) were examined with MRI and compared to a selfexpanding nitinol stent with a new design concept. Stents were tested in a well controlled flow phantom and in-vivo (swine, n = 2), whereas stents were placed in the aorta and iliac arteries (Dynerlink, Smart, Flexx each n = 2; new designed stent n = 6). MRI was done with bFFE, 3D angiogaphy and VEC-MRI. The following characteristics were investigated: (1) width of susceptibility artifacts at the stent wall (2) signal intensity inside the lumen of the stent versus next to the stent (3) quantitative flow through the stent versus next to the stent.
Results: The Cordis stent showed broad susceptibility artifacts and was therefore excluded from further investigation. All other stents showed only negligible susceptibility artifacts (1.3 ± 0.5 pixels, in-vitro measurements). In the new stent concept there was no significant decrease in signal intensity within the stent lumen. All other stents showed significant decrease in signal intensity of −27 ± 8% to −55 ± 9% ($p < .001$, in-vitro) and −33 ± 11% to 54 ± 7% ($p < 0.001$, in-vivo). Quantification of blood flow through the stent lumen was accurate in all investigated stents. There was no significant difference between measurements performed within the stent lumen when compared to measurements next to the stent lumen. However, only the lumen of the new stent concept was not obscured by significantly decreased signal intensity on the magnitude images. Thus tracing stent boarders for flow analysis was difficult in all other stent types and can potentially induce significant errors in quantitative flow.

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Conclusion: The results of this study imply that the new design concept for a nitinol stent is well suited for non-invasive assessment of lumen patency using MRI.

Session 8: Interventions 2

64

Transcatheter closure of perimembranous ventricular septal defects with the Amplatzer asymmetric ventricular septal defect occluder in children. Further experience and follow-up

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Background: The design of previously used devices for transcatheter closure (TC) of perimembranous ventricular septal defects (PMVSDs) is not ideal for this purpose and their use has been associated with several drawbacks. The aim of this study was to present further experience with TC of PMVSDs in children using a new device the Amplatzer asymmetric ventricular septal defect occluder (AAVSDO).
Methods: Seventeen patients, aged 1.5 to 14 years, with PMVSDs underwent transcatheter closure using the AAVSDO. The device consists of two low profile discs made of Nitinol wire mesh with a 1.5 mm connecting waist. The left-sided disc is 5 mm towards the apex and only ½ mm towards the aortic valve. The right-sided disc is 4 mm larger than the waist. The prosthesis size (waist diameter) was chosen to be 1–2 mm larger than the PMVSD diameter. A 7F or an 8F sheath was used for the delivery of the AAVSDO. Fluoroscopy and transesophageal echocardiography were used for the guidance of the procedure.
Results: The PMVSD diameter ranged from 2 to 8 mm. The device diameter ranged from 4 to 10 mm. Device placement was successful and associated with complete closure in 15/17 patients (88%). In two patients there was a trivial residual shunt after the procedure that had disappeared in one of them at the 3-month follow-up (94%). The main complication was embolization of the in 1/17 patients. No other complications occurred during a mean follow-up of 6.5 months (range 1–13 months).
Conclusions: The AAVSDO appears to be a promising device for the transcatheter closure of PMVSDs in children. Further studies are required to document its efficacy, safety and long-term results in a larger number of patients.

65

Perimembranous ventricular septal defect occlusion in children, using the transcatheter patch

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Perimembranous ventricular septal defect (VSD) occlusion is a challenging application for metal disk devices, since there is risk of aortic or tricuspid insufficiency (deficient rim); furthermore malalignment VSDs are excluded. The transcatheter patch (TP) is wireless, requires minimal rim and has 3-dimentional shape; therefore valve interference and aortic malalignment should not be an issue. Ten consecutive surgical candidates for perimembranous VSD correction were occluded by TP. Defect size varied from 5–16 mm (med.8) with a Qp:Qs larger than 2:1 in all cases. Most defects (8) had deficient sub-aortic rims; one Fallot Tetralogy case had significant aortic malalignment. Patient age varied from 2–10 years (med.7) and patient weight from 10–30 Kg (med.21). All VSDs were corrected transvenously over arteriovenous wires, using 9–12F long sheaths. TPs were released in 48 hours in 8 patients and in 24 hours in 2. All procedures resulted in effective occlusions (9 full occlusions, 1 trivial residual shunt). One TP was pulled out by mistake and was retrieved through the introducing sheath. There were no embolizations or cases of aortic insufficiency. There were no problems on follow-up except for a brief hemolysis episode in the Fallot Tetralogy case. All patients are doing well. In conclusion, TP occlusion of perimembranous VSDs appears effective and safe, even in cases with deficient subaortic rim or aortic malalignment.

66

Percutaneous closure of perimembranous septal defects

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Background: With the introduction of the Amplatzer VSD occluder (AVO), selected patients with perimembranous ventricular septal defect (pmVSD) can be suitable to transcatheter closure.
Aim: To report our experience in pmVSD closure with various AVO devices.
Patients and Methods: Between Jan 2000 and October 2002, 16 patients underwent percutaneous closure of pmVSD. Thirteen had perimembranous congenital VSD while 3 patients had residual post-surgical subaortic VSD.Two different Amplatzer devices were used: the muscular VSD occluder and the eccentric VSD occluder. In patients in whom the mVSD occluder was used the transaortic approach has been used without need for an artero-venous circuit, while in subjects in whom the eVSD occluder was employed and artero-venous circuit was needed. The mVSD device was used before the availability of the eVSD occluder in subjects with pmVSD but with a portion of tissue between the defect and the aortic valve.
Results: Median age at intervention was 9 years (range 2.5–62 years). Median weight was 30 kgs (range 11–80 kgs). Mean QP/QS was 2.1 ± 0.6. Mean procedure and fluoroscopy times were 172 ± 70 min and 33 ± 13 min, respectively. One patient had dilation and stent implantation of native coarctation of the aorta in the same session. The mVSD occluder was used in 10 patients while the eVSD devide was used in 5 subjects. The median size of device was 8 mm (range 6–16 mm). In one patient a 6 mm eVSD device embolized in the right pulmonary artery soon after implantation. The device was retrieved percutaneously and a 10 mm eVSD device was successfully implanted. All other patients experienced no procedural complication. A mild to trivial introprotesic residual shunt was showed in 4 subjects at discharge. All patients but one were discharged home 48 hours after procedure. One subject due to post-procedural fever was discharged 96 hours after procedure.
Follow-up: Median follow-up was 9 months (range 2–24 months). Complete occlusion was shown in all subjects except one. No late complication occurred. One subject showed trivial to mild aortic insufficiency.
Conclusion: In the current era and in experienced hands, pm VSD closure can be performed safely and successfully.

67

Transcatheter closure of congenital ventricular septal defects by the amplatzer VSD occluder device: mid-term follow-up

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Surgical closure of congenital ventricular septal defects (VSDs), is still associated with significant morbidity and mortality. Encouraging results of transcatheter closure of VSDs with Amplatzer VSD occluder device, have been recently described. The aim of this study is to report our experience in percutaneous closure of congenital VSDs. Between August 1998 and December 2002, 27 patients (pts) aged 5 months to 62 years (mean 17.9 ± 18.9 years) underwent transcatheter closure of a VSD. All patients, intubated and under general anesthesia, underwent right and left heart catheterization. The location of the defect was defined by angled angiographic views (8 musculars, 2 multiple musculars, 12 perimembranous, 5 residuals post/op). The deployment of the device has been guided by fluoroscopy and transoesophageal echocardiography. After release of the device a color-Doppler echocardiographic study, a left ventriculography, and an aortography were performed to detect a residual shunt or an aortic valve regurgitation. All patients received heparin (100 units/Kg) during the procedure and ASA (5 mg/Kg daily) after the procedure for the following 6 months. All patients had a chest-X-ray and a transthoracic color-Doppler echocardiographic study 24 hours after the procedure and at the follow-up in the outpatient clinic. The VSD diameters ranged from 4 to 14 mm. Pulmonary/systemic flow ratio (Qp/Qs) varied from 1.7 to 3 (mean 2.1 ± 0.9). The device was successfully delivered in all patients. Immediate complete closure was obtained in 18 pts; a tiny early residual shunt was detected in 9 pts. A suboptimal device position with substantial residual shunt occured in 1 pt; the device was surgically removed and the defect closed. There was no mortality. Follow-up echocardiographic data were available in all pts. The mean F-up is 17.5 ± 14.3 months. The device is in an appropriate position, not interfering with the adjacent cardiac structures in all pts but one with tricuspid regurgitation; no evidence of residual shunt in all pts. A residual VSD was observed in one pt with multiple muscular VSDs.
Conclusion: The transcatheter VSDs closure using an Amplatzer VSD occluder appears to be a promising technique; but before it enters routine clinical practice, studies documenting the long term safety are required.

68
Percutaneous closure of perimembranous ventricular septal defect – initial experience in the British Isles

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Background: Significant perimembranous ventricular septal defects (VSD) have previously required surgical closure. Advances in materials science and catheter technology have brought device closure of these defects within reach of interventional cardiac catheterisation. We present the results of the first 13 patients undergoing attempted device closure of perimembranous VSD in three cardiac centres in the British Isles.
Patients: 12 children (median age 5.75 y, median weight 19.9 kg) and one adult (aged 75, weight 70 kg) were considered for device closure. All patients were symptomatic, having significant shunts confirmed clinically and on transthoracic echocardiography.
Technique: Transoesophageal echocardiography and diagnostic cardiac catheterisation was performed under general anaesthesia. VSD size, morphology and adjacent structures were assessed. Doppler studies of aortic, tricuspid valve and mitral valve were recorded before and after attempted VSD closure. The Amplatzer membranous ventricular septal occluder (mVSDO), soft exchange wire and purpose designed delivery system were used in all patients. The wire was introduced retrogradely across the VSD, snared in the pulmonary artery and brought out of the femoral vein to form a loop. The delivery sheath was advanced antegrade over the wire into the aorta. The delivery sheath was withdrawn from the aorta and manipulated into a satisfactory position in the left ventricle, aided by a catheter advanced retrograde over the wire. Devices were presoaked in blood, and deployed through the delivery sheath to occlude the VSD using echocardiographic, fluoroscopic and angiographic guidance.
Results: Median VSD dimension was 8 mm. No patients were unsuitable for attempted closure. Successful device closure was achieved in all patients (median fluoroscopy time 22 min). There were no other important complications, and no patient had aortic incompetence after device deployment. Transient right bundle branch block was seen in one patient, trivial residual VSD flow in 3 patients immediately after the release, and mild tricuspid incompetence in 2 patients. In 2 patients the right ventricular component of the device was constrained and deformed by aneurysmal tissue, but complete occlusion was achieved. No patient developed haemolysis.
Conclusion: Transcatheter occlusion of restrictive perimembranous VSD has developed as a true alternative to surgery. Long-term results will allow for comparison between the two techniques.

69
Transcatheter therapy of vascular occlusions affecting the venous or arterial circulation

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Purpose: To report our experience with interventions performed for relief of acute arterial or venous obstructions in pediatric patients.
Patients: A total of 31 patients were treated. Nine patients (age 3 months to 15 years) had venous thromboses after insertion of central-venous catheters (n = 8) or after surgery with immobilization (n = 1), one patient had a co-existing pulmonary arterial embolism. 21 patients had arterial thromboses after retrograde cardiac catheterisation (n = 18) or placement of arterial lines (n = 3) for invasive monitoring. One patient had obstruction of the hemodialysis access site. All patients had signs of either venous congestion or insufficient arterial perfusion.
Methods: The venous thrombotic occlusions were recanalized by use of 0.035-in guidewires and clots were macerated by use of the Amplatz thrombectomy device (Helix, Bard). Angioplasty was performed after clot maceration to compress residual thrombus to the vessel wall. Local thrombolytic therapy was administered after recanalization in 2 patients with rt-PA and abciximab. The arterial thomboses were treated with anterograde (n = 17) or retrograde (n = 5) balloon angioplasty after recanalization of the occluded vessel by use of floppy guidewires (ACS) or fixed wire coronary angioplasty catheters (ACE, Scimed).
Results: All venous thromboses were successfully recanalized, in the patient with pulmonary arterial embolism, the pulmonary perfusion was effectively restored. In one patient, re-occlusion of the recanalized vein occurred since the central venous (Hickman) catheter was not removed. 19 of 22 arterial obstructions were successfully recanalized. Duplex-sonography showed a patent femoral artery in 19/22 patients, in 2/22 patients, there was a residual stenosis of the femoral artery with a luminal narrowing of 50% and 60%, respectively. In one patient, restoration of femoral arterial blood flow was impossible. The hemodialysis access was effectively recanalized.

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Conclusion: Interventional techniques provide a safe and effective therapy to restore venous and arterial blood flow in pediatric patients with no morbidity and no mortality and low re-occlusion rates. Transcatheter therapy may be superior to fibrinolytic therapy.

70
Percutaneous closure of patent foramen ovale for paradoxical embolism in patients with and without a thrombophylic condition

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Background: Several treatments have been reported for pts with a patent foramen ovale (PFO) and paradoxical cerebral embolism but no comparative data are available on the efficacy in pts with or without a thrombophylic condition (TC).
Methods: 35 consecutive pts were evaluated from 1999 to 2002 for percutaneous PFO closure because of a cerebral ischemic event of unknown origin. A complete hematologic survey evidenced at least a TC in 9 pts (27%) (Table). Contrast TEE showed a PFO with large right-to-left shunt (>20 bubbles) in 20 (57%) pts and an aneurysm of interatrial septum (ASA) in 18 (51%). Twenty pts (57%) had a stroke as first manifestation while 15 (43%) experienced a TIA. Patient history was retrospectively reconstructed from the first event to percutaneous closure for all. After the first event 7 pts (77%) with an associated TC received oral anticoagulation, while the others 325 mg/d of aspirin. Mean age at intervention was 43.3 ± 13.8 yrs.

Patient	MTHFR mutation + Hyperhomocysteinemia	Protein C deficiency	Prothrombin gene mutation	Lupus anticoagulant	Activated protein C resistance
1	+				
2	+		+		+
3					+
4	+		+	+	
5	+				
6	+				
7	+		+		
8	+				
9		+			

Results: Prior to percutaneous closure, pts with a TC had a significantly higher risk of recurrences than pts without ($p = 0.0013$) despite a similar follow-up ($p = 0.16$), a more aggressive medical treatment and regardless of shunt entity ($p = 0.45$), presence of an ASA ($p = 0.7$) and incidence of risk factors for stroke ($p = 0.9$). PFO closure was successful in all pts (29 Cardioseal and 6 Amplatzer PFO devices) without any complication. A residual shunt was immediately noted in 6 pts (29%) and in 2 pts (7%) at six months and were left on aspirin. After a mean of 16 ± 9.4 months from PFO closure no recurrent event occurred, despite a similar follow-up between the 2 groups ($p = 0.4$). Pts with a TC received warfarin for 6 months after closure while the others aspirin for 6 months + ticlopidine for 3 months.
Conclusions: The association of a PFO with a TC significantly increases the risk of recurrences in pts with cryptogenic stroke, despite oral anticoagulation. Transcatheter PFO closure was effective in both groups, so that the former seem to benefit even more. Detection of a TC in pts with a PFO should suggest transcatheter closure. Optimal medical therapy after closure still remains to be established.

71
Do we have to expect atrial fibrillation after transcatheter closure of interatrial communications? Single center experience in 1184 patients

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Background: In many centers transcatheter closure of secundum atrial septal defects (ASD) and patent foramen ovale (PFO) is performed as a routine procedure. One complication which can be detected more frequently is atrial fibrillation (AF). The purpose of this study was to investigate the incidence and clinical consequences of AF after catheter device closure.
Methods: In our center 1184 patients (pts) underwent transcatheter closure of interatrial communications (ASD n = 445, PFO n = 739) and were enrolled in one of several prospective studies. We used 8 different devices. ECG was performed after 4 weeks, 6 months and annually.
Results: In 37 pts (3.1%) the follow up ECG showed a new AF after closure (ASD: 4.7%, PFO 2.1%). The mean age was 53 ± 12 years (AF pts) vs. 47 ± 15 years (patients without AF) ($p < 0.01$). The median duration until onset of the AF was 1 ± 14 months (range 0.03–60 months) (ASD: 1 ± 0.4 months, PFO: 6 ± 21 months ($p < 0.01$)). AF was observed with different incidences depending on the devices used for closure: Sideris 7.7% (n = 4), Amplatzer: 5.5% (n = 18), ASDOS 4.6% (n = 2), Starflex 5.2% (n = 9), Cardioseal 3.7% (n = 1), Helex 1.6% (n = 3). The incidence of AF in the Helex subgroup was significantly lower compared to the other devices ($p < 0.01$). Cardioversion was successful in 30 pts. In the last follow up 7 pts still had AF. They were treated with anticoagulants. Patients with persistent AF had had a significant later onset of their AF compared to pts who changed into sinusrhythm (24 ± 25 months (persistent) vs. 1 ± 2 months (successful cardioversion) ($p < 0.01$)). No thrombembolic complications occurred. The following risk factors for AF could be found: CHD, Diabetes, arterial hypertension, elevated cardiothoracal quotient ($p < 0.01$), device size ($p < 0.05$). No relevance was found for defect diameter, QRS duration, RVD diameter, presence of an atrial septal aneurysm and size of the atrium.
Conclusions: Atrial fibrillation is not a rare complication after interventional ASD and PFO closure. It seems to be less frequent with the Helex occluder, which is very soft and which has a round design Delayed onset of the AF seems to be associated with lower success rates of cardioversion.

72
Transcatheter closure of paravalvar leaks using the Amplatzer muscular VSD occluder and gianturco coils

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Paravalvar leaks are rare, but significant complications after placement of artificial valves in aortic or mitral position. Surgical repair is difficult, particularly after double valve replacement. In order to determine the feasibility, safety and efficacy of transcatheter closure of these paravalvar leaks we reviewed our two-year single center experience using the Amplatzer Muscular VSD Occluder or Gianturco coils. From 1/01 until 12/02, transcatheter closure was attempted in five consecutive procedures on four patients, aged 9 to 31 years (median 23.6 years). Patient weight ranged from 31.2 to 94.0 kg (median 53.4 kg). Para-aortic leaks were present in 2 and

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para-mitral leaks in 3 procedures. Hemolysis was present in 3. Regurgitation ranged from angiographic grade 2.5–3.5 (median 3.0). Left ventricular end diastolic pressure was elevated in all patients ranging from 14 to 45 mmHg (median 23.5 mmHg). There was a single leak in 3 patients and 3 leaks in one patient. The leak diameter ranged from 2.0 to 6.4 mm (mean 4.4 mm). All attempts of transcatheter device or coil placement were successful. Four Amplatzer Muscular VSD Occluders were implanted in 3 patients (device diameter 6–10 mm). Ten 5 mm × 3 cm 0.038 inch Gianturco coils were implanted in 2 paravalvar leaks in 1 patient. Procedure time ranged from 107 to 275 minutes (median 143 minutes). Fluoroscopy time ranged from 17.6 to 85.8 minutes (median 28.8 minutes). There were no major complications. Minor complications were increased hemolysis after 1 procedure and transient hemolysis for 2 days after 1 procedure. There was no significant interference with artificial valve function. Complete closure was achieved in 2 patients. A trivial residual leak measuring 0.8 mm and a small leak measuring 2 mm was present at 1 month in 2 patients. Transcatheter closure of paravalvar leaks appears feasible, safe and relatively effective, although crescent shaped leaks may require placement of at least 2 devices.

Session 9: General

73

CardioPat – a comprehensive computer system which uses the European Paediatric Cardiology Code for documenting medical data in a clinical environment

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Introduction: The coding committee of the AEPC published the European Paediatric Cardiology Code (EPCC) in January 2000 (updated October 2002). We regard it as an important tool for comparing diagnoses on a larger scale e.g. throughout Europe. We also appreciate its accuracy. Therefore we decided to use the EPCC (long list) routinely for documenting diagnoses and procedures of our heart patients (900 in-patients, 400 open-heart-operations, 6,000 out-patients per year).
Purpose: The development of a computer system which enables doctors to use the EPCC with a minimum of additional work.
Methods: In 6 years of research and programming the author developed a computer application that allows the input of almost all medical data: patient's history, findings, diagnoses, cath lab reports, examination results, daily notes and the discharge letter. The programme is written in Borland Delphi and uses an Oracle database. CardioPat offers an efficient diagnosis platform which is open to the EPCC and the ICD10. In this way, the EPCC can be used for the cardiological and the ICD10 for non-cardiological diagnoses of heart patients. Because of the ICD10 mapping of the EPCC and the concept of single input and multiple use, the burden of coding and administrative work is reduced markedly. Finding the correct EPCC out of 4430 different codes is done easily, because standard abbreviations or keywords are related to these codes. Fulltext search is also possible. CardioPat uses Client/Server technology. Every doctor can use any PC in our hospital. The hospital's main computer delivers the case number, patient ID and other personal data of the patient. The laboratory results reach CardioPat through a HL7 messaging interface. At the moment the database holds more then 65,000 cases, 660,000 laboratory results and 20.000 diagnoses and procedure codes. CardioPat can be used in German or English.
Operating system: Microsoft Windows '95, '98, NT4, 2000 or XP.
Conclusion: The development of a comprehensive computer system (CardioPat) for paediatric cardiology enables us to reach relevant data in seconds. CardioPat is an important tool for research, quality assurance and administration. Optimal computerized support keeps the administrative work of the physician in tolerable limits.

74

Complete atrioventricular septal defect: surgical outcome of biventricular repair in relation to preoperative echocardiographic measurements

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Objective: To assess the relationship between preoperative echocardiografic measurements and operative outcome following biventricular repair of CAVSD.
Methods: Since 1992, 105 patients have undergone biventricular repair of CAVSD. A single observer (G.C.) undertook retrospective review of the preoperative echocardiograms of all patients. In 61(58%) patients it was possible to make measurements of the LAVV and RAVV diameter; LV and RV inflow length, LV and RV area in 4 apical chamber view. All measurements were expressed as Z-scores. 4 patients had associated TOF, 2 CoAo and 1 had LVOTO.
Results: Median Z-score of LAVV diameter was −1.3 (range: −5.7 to 3.5), of RAVV diameter was 0.7 (range: −6.5 to 6), of LV inflow length was −0.23 (range: −2.4 to 3), of RV inflow length was 0.2 (range: −2.1 to 2.9), of LV area −0.5 (range: −3.4 to 2.4), of RV area 1 (range: −2.1 to 4.3). Median follow-up was 6.2 years. There was 1 early death (1.6%), 3 late death (5%) and 3 patients subsequently underwent replacement of LAVV (5.2%). The population was divided in 2 groups on the basis of their surgical outcome. 54 patients (group 1) had no significant postoperative complications. 7 patients (group 2) had the above reported major postoperative complications. The association of small LAVV (Z-score < -3) and shortened LV length (Z-score < -1) was significantly more common in group 2.
Conclusions: Early and medium term results following biventricular repair of CAVSD are good even with small AV valve dimensions. The association of a small LAVV and shortened LV length is a significant negative prognostic factor. Precise echocardiografic definition of AVV and ventricles morphometric parameters is therefore important in preoperative assessment of this complex congenital heart disease.

75

Systemic embolism in children with myocarditis

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Background: Myocarditis continues to be an important cause of hospital admission in our part of the world. Systemic embolisation due to thrombi in left ventricle (LV) is a rare complication of these patients (pts).
Objective: To investigate the incidence, course and outcome of thrombo-embolism in children with acute myocarditis.
Design: A 3-year (Dec 99–Nov 02) analysis of all children <12 yrs admitted and diagnosed as having acute myocarditis in a single tertiary referral center.

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Patients and Methods: The charts and echocardiographic records of all pts with acute myocarditis were reviewed. Data was reviewed for mode of presentation, age, hospital course, and outcome. Echocardiography (echo) data was analysed with special reference to the size, function and presence of thrombus in LV.
Results: Of 2716 admissions to the paediatric cardiology unit, myocarditis was the underlying lesion in 405 pts (14.9%). The mean age was 2.1 + 4 yrs. Systemic embolisation was the presenting feature in 28 (6.9%) pts (Group A), while another 17 (4.2%) developed it during the hospital stay (Group B). Another 5 pts had LV thrombus on echo but did not develop embolisation. All 50 pts showed seriously impaired systolic function of the LV with fractional shortening (FS) of 10 ± 3%, range 5%–20%, as compared to those without thrombo-embolism with FS of 17% ± 6%, range 5%–26% ($P < 0.0001$). In Group A, 26/28 pts presented with stroke. Only 6 had thrombus in LV on echo at presentation. All pts were treated with heparin and oral anticoagulants. There were two deaths. In Group B, 3/17 pts had thrombus in LV on echo at presentation. They were started with heparin but still went on to develop stroke. There were two deaths in this group as well.
Conclusion: Myocarditis is an important cause of hospital admission in our part of the world accounting for 15% of all admissions to a paediatric cardiology unit. These pts are at risk of developing systemic embolisation, which could be the first presentation. The risk is higher in pts with lower FS of the LV. All pts with FS below 17% should be treated with prophylactic anticoagulative agents.

76
Carvedilol in pediatric patients with myocardial failure

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The objective was to evaluate the effect of the beta-adrenoceptor blocker carvedilol (C) in pediatric patients with congestive heart failure failing to improve on standard therapy (digoxin, ACE-inhibitors, diuretics). Due to the maturation of the myocardium, the autonomic nervous system and the systems involved in drug metabolism and elimination, children with heart failure may show a different response to carvedilol compared to adults. In a prospective open study, 30 patients (3 weeks to 19 years, 8 females, 22 males) with congestive heart failure due to dilated cardiomyopathy ($n = 15$) or congenital heart disease ($n = 15$) received increasing doses of oral C (initially 0.09 followed by 0.18, 0.35 and 0.70 mg/(kg day)) in addition to standard therapy. Ejection fraction (EF) was determined monthly up to 6 months and analyzed with the investigator blinded towards the patient and the measurement time point. Overall, EF improved from 38 ± 18% to 50 ± 19% after 6 months ($p < 0.05$) and this was accompanied by a decrease in Nt-proBNP plasma concentrations from 1090 ± 758 to 443 ± 268 fmol/ml ($p < 0.05$). Substantial age-dependent differences were noted. In young children (YC; 3 weeks to 1.5 years), EF increased from baseline immediately within one month after onset of C therapy, whereas in older children/adolescents (A; 3.5 to 19 years) ventricular function improved with a delay reaching values over baseline 3 months after initiation of therapy. C reduced heart rate earlier in young pediatric patients. An analysis of this important prognostic parameter using pharmacokinetic/pharmacodynamic modeling revealed an increased potency for heart rate reduction (EC50; YC: 2.8 ± 2.0; A: 10.6 ± 2.9 ng/ml C; $p < 0.05$). These results suggest that the early improvement of ventricular function in young children is associated with an early onset of heart rate reduction in young children compared to older children/adolescents. Young children benefit from C therapy mostly because they experienced early ventricular improvement.

77
Modulation of neurohormonal activity after carvedilol therapy in pediatric patients with heart failure

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Background: In adults with heart failure, neurohormonal overstimulation is related to the progression of the disease and influences prognosis. Beta-blockers, which act modulating neurohormonal activation, play nowadays an essential role in the pharmacological management of heart failure in adults, but its use in paediatric patients is very limited.
Patients and Methods: To investigate the effects of carvedilol administration on neurohormonal activation and left ventricular function, carvedilol was added to standard heart failure therapy in 9 pediatric patients with dilated cardiomyopathy due to heart muscle disease who were receiving standard therapy for at least 1 month. Study protocol consisted in a baseline evaluation to assess neurohormonal activation and echocardiographic left ventricular function. This was followed by a final evaluation at 12 months from carvedilol loading. Carvdilol was started at 0.05 mg/Kg/day and up-titrated every two week until the target dose of 0.8 mg/Kg/day was reached.
Results: Carvedilol administration was associated with a significant reduction in plasma norepinephrine, dopamine, aldosterone and renin-angiotensin system activation (Table). Similar reductions in vanilmandelic ($p = 0.0007$) and homovanillic acid ($p = 0.0001$) were noted. After 12 months a positive remodeling took place with significant reductions in end-diastolic and end-systolic diameters and increase in left ventricular ejection fraction. No adverse effects needing drug down-titration or interruption were noted in the run-in phase or in the maintenance period.

	Baseline	Carvedilol	p
LVEDD	6.7 ± 2.2	5.5 ± 2.0	0.004
LVDSD	4.6 ± 1.9	3.5 ± 1.6	0.009
LVEF	28 ± 6	47 ± 14	0.001
NE	914 ± 55	681 ± 150	0.00001
Ald	1242 ± 190	329 ± 151	0.00001
RA	31.5 ± 7.2	21.5 ± 5.7	0.0006
Dopamine	1239 ± 128	412 ± 116	0.0001

Ald indicates plasmatic aldosterone (pg/ml); Dopamine, plasmatic dopamine (mcg/g cretainine); LVEF, left ventricular ejection fraction (%); LVEDD, left ventricular end-diastolic diameter (cm/m^2); LVESD, left ventricular end-systolic diameter (cm/m^2); NE, plasmatic norepinephrine (ng/l); RA, plasmatic renin-angiotensin activation (ng/ml/h).

Conclusion: Carvedilol was a safe complement to standard therapy for heart failure in paediatric patients, allowing a significant reduction of neurohormonal activation with evident benefits on ventricular function and clinical condition.

78
Clinical feature of Kawasaki Disease in infants younger than 3 months of age

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Background: Previous studies have demonstrated that Kawasaki disease (KD) in infants (<3 months) is rare condition, however it usually takes severe and atypical clinical course with high incidence of coronary artery aneurysms. After the introduction of intravenous gamma globulin (IVGG) treatment for the acute KD, the incidence of coronary lesions was significantly decreased, however little information was available about the clinical features in this patient population after the introduction of IVGG treatment.
Method: From January 1995 through March 2001, 218 patients with KD were experienced in our hospital, of which 12 infants (5.5%) suffered from KD < 3 months of age. Their clinical feature was retrospectively analyzed.
Results: Patients consisted of 7 boys and 5 girls. Median onset of disease was 72 (ranged from 37 to 86) days after birth. Patients were admitted on 2.8 ± 1.3 days of illness. Only 3 patients were suspected of KD at the admission. Diagnosis was completed on 4.8 ± 1.7 days of illness, and finally they were satisfied with 5.4 ± 0.7 items of diagnostic criteria of KD. The lacking items of diagnostic criteria were cervical lymphadenopathy (n = 5), change of extremities (n = 3) and skin rash (n = 1). IVGG was indicated in 11 infants (2 g/kg/day in 9,400 mg/kg/day for 5 days in 2) and initial treatment was started on 4.4 ± 1.1 days of illness. Aspirin was given to all 12 patients. Initial IVGG treatment was failed in 4 patients, in such cases methylprednisolone pulse therapy was indicated. As a result, 6 (50%) of 12 patients were complicated with coronary arterial lesions, including 4 patients with coronary aneurysm and 2 patients with transient dilatation. Non-cardiovascular complications, such as aseptic pyuria (n = 5), disseminated intravascular coagulation (n = 2), aseptic meningitis (n = 2), hypotension or apnea during IVGG infusion (n = 2), convulsion (n = 1) or jaundice (n = 1) were mostly found within 5 days of illness.
Conclusion: In spite of the optimal IVGG treatment was used in these infants, the incidence of coronary artery aneurysm was still high. Early diagnosis and other therapeutic strategies should be required for the improvement of cardiovascular outcome in these high-risk patients.

79
Identification of high-risk status among patients with childhood hypertrophic cardiomyopathy

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Background: Hypertrophic cardiomyopathy is the commonest medical cause of sudden death in older children and teenagers.
Methods: In a retrospective cohortstudie from six paediatric cardiology centers 129 patients with hypertrophic cardiomyopathy (HCM) diagnosed before the age of 19 yrs, and with an average follow-up of 9.7 yrs, were identified. Multi-variable correlation analysis were used to identify EKG and echocardiographic variables that correlated with increased risk of sudden death or heart-failure-related death.
Results: There were 30 disease-related deaths, out of which 16 were sudden deaths. Deaths related to heart failure most commonly occurred during first two years of life, and then reappear after 12 years of age. Most sudden deaths occurred between 8 and 16 yrs of age, with a peak around 10–12 yrs. The risk factors for sudden death are different to the risk factors for heart failure-related deaths. There is a positive correlation between sudden death and septal and LV-wall thickness ($p < 0.0001$ and $p < 0.03$), ECG-amplitudes measured as sum of R and S-waves in all six limb leads ($p > 0.0001$), Sokolow-Lyon index ($p < 0.0001$) and frequent ventricular extra-systoles ($p < 0.004$). Heart failure-related death does not correlate with ECG-amplitudes, but does correlate most strongly to increased relative wall thickness expressed as wall-to-cavity ratio, particularly posterior LV wall-to-cavity ratio ($p < 0.0001$). The presence of a co-existing Noonan's syndrome or therapy with calcium-blockers increases risk of heart-failure related death, odds ratios 11.0 (95%CI 2.9–41) and 4.4 (1.1–17.3) respectively. Analysis of frequency distributions suggest the following measures as useful cut-offs between patients at high or low risk of sudden death: RS-sum in limb leads >11 mV, odds ratio 146 (8.4–2500); septum >2.0 cm, odds ratio 9.7 (2.0–47.8); frequent ventricular ectopics, odds ratio 15.6 (2.6–92.7). The patient at high risk of heart-failure related death is identified by an LV wall-to-cavity ratio >0.40, odds ratio 42.2 (2.0–885). MANOVA shows that therapy with high-dose propranolol reduces risk for all disease-related death independent of riskfactors ($p = 0.004$), with effects both on sudden death ($p = 0.03$) and heart-failure-related death ($p = 0.008$).
Conclusions: High-risk childhood HCM patients can be prospectively identified, and medical therapy which reduces the risk is available.

80
Management of children with severe pulmonary hypertension

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Introduction: Pulmonary vascular disease increases pulmonary vascular resistance (PVR), leading to right ventricular failure and death. Pulmonary hypertension (PH) may be primary (PPH) or secondary to several disorders, particularly congenital heart disease or lung disease. Untreated, the prognosis is poor, irrespective of etiology. Specific treatments of proven use in adults with PPH are now being given to children. The aim of the study was to review retrospectively the patient population and management of children with PH referred to our tertiary centre. Characteristics of patients referred: 52 severely symptomatic children aged between 2 weeks and 17 years, presented at a mean age of 9 years. 24 patients had primary and 28 secondary PH. Haemodynamic studies in 47 children: Pulmonary artery pressure^3 systemic in 26 patients. Pulmonary vascular resistance (PVR) was >10 μm^2 in 25, and 5–10 μm^2 in 12 children. PVR was unresponsive to acute vasodilator testing, including nitric oxide, in 30 patients.
Outcome: Comparison of treatment before and after referral showed an increase in the use of expensive specific therapies (continuous intravenous epoprostenol and oral bosentan which is an ET-A and ET- B receptor antagonist) after referral (Fig). 14 children aged 1–16 years have received epoprostenol for 8 months – 5 years (mean 1.7 years). 16 children aged 1–16 years have received bosentan for 1–10 months (mean 4.2 months). 5 of these take both drugs. 95% of children have been stabilised and show clinical improvement, and a beneficial shift in NYHA classification (Table). Hickman line needed resiting in 6 patients because of a local line infection. Liver dysfunction has not been a feature of treatment with bosentan in these children, although previously reported in adults.

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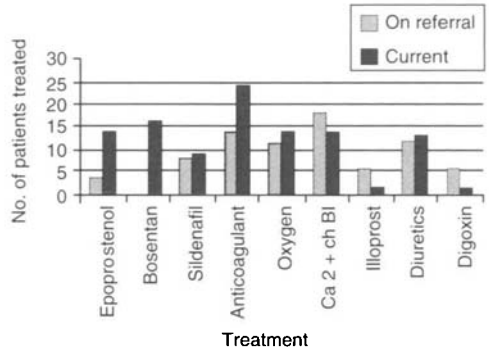
	Class I	Class II	Class III	Class IV
On referral	0	16	26	10
Current	2	22	22	3
Deceased				3



Conclusion: New but complex therapies, given singly and in combination can improve quality of life and survival. Most children improved significantly, older ones returning to school.Early diagnosis, prompt referral to a specialist PH team giving comprehensive paediatric care and prompt use of specific therapies is crucial. Response to treatment needs constant review and frequent revision, with respect to both type of drug and dosage.

81

Levosimendan is a pulmonary vasodilator in children after cardiac surgery

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Background: Increased pulmonary vascular resistance (PVR) in children with congenital heart disease remains an issue in postoperative intensive care management. The role of K+-channels, known to be involved in pulmonary hypoxic vasoconstriction, has not been elucidated in this situation. We compared intravenous Levosimendan (Simdax®), an inodilator acting on pulmonary KATP-channels, with the pulmonary vasodilating effects of inhaled nitric oxide (NO).
Methods: 5 infants (age 1.6 ± 2.6 years, weight 7.9 ± 5.6 kg) with intracardiac shunting defects and postoperatively increased PVR (6.2 ± 2.3 WU m^2) were included. All were examined in the early postoperative period while they were still sedated and paralysed. PVR was measured using the direct Fick principle with measurement of oxygen consumption by respiratory mass spectrometry. Measurements were performed at baseline, with FiO_2 of 0.65 combined with NO (20 ppm) before and after a Levosimendan bolus (24 μg/kg).
Results: Intravenous Levosimendan (LS) alone decreased postoperative PVR-Index with 41% efficacy of NO alone (i.e. PVRI fell 9.5% with LS vs 23.1% with NO, $p = 0.04$). The combination of LS with NO produced no further pulmonary vasodilation. Intrapulmonary shunting increased in LS alone (fall of arterial pO_2 from 163 ± 0.51 to 137 ± 0.58 mmHg), but not in NO.
Conclusion: Levosimendan lowered PVR with almost half the potency of inhaled NO. The associated increase in intrapulmonary shunting remained clinically acceptable. Thus, intravenous LS may be useful as an additional substance to reduce right ventricular afterload.

Session 10: Interventions 3/General

82

Left ventricular pressure–volume relationship in patients with atrial septal defect: acute influence of the balloon occlusion of atrial septal defect

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Background: Catheter interventional treatment of atrial septal defect (ASD) can cause acute hemodynamic changes under the physiological condition without the influence of cardiopulmonary bypass. These hemodynamic changes probably influence on systolic or diastolic parameters of left ventricle. However, the acute hemodynamic changes before and after transcatheter closure of ASD have not been evaluated completely. We previously reported that significant elevation of brain natriuretic peptide transiently 24 hours after the transcatheter closure of ASD.
Purpose: The purpose of study was to evaluate the acute hemodynamic changes of left ventricle before and after transcatheter closure of ASD using pressure–volume curve.
Methods: Sixteen patients with transcatheter closure of ASD were enrolled in this study. Their ages ranged from 5 to 20 years (median 11.4 years). Diameter of ASD by transesophageal echocardiography ranged from 9 to 22 mm (median 15 mm). Occlusion balloon diameter of defects ranged from 10 to 33 mm (median 15.9 mm). Their Qp/Qs ranged from 1.2 to 4.0 (median 2.2). The pressure volume relationship data were obtained before and after balloon occlusion of ASD using an integrated conductance and micromanometer-tipped catheter placed in the left ventricle.
Results: Due to balloon occlusion of ASD, end-diastolic volume (EDV) was significantly increased (89 ± 38 ml vs. 101 ± 42 ml, $p < 0.001$), and ejection fraction (EF) was significantly decreased (65 ± 9% vs. 62 ± 8%, $p = 0.029$). Stroke volume (62 ± 29 ml vs. 63 ± 27 ml) and stroke work (4590 ± 2494 mmHg ml vs. 4802 ± 2062 mmHg ml) were not significantly changed. Although peak filling rate did not change (617 ± 314 ml/s vs. 630 ± 343 ml/s), end-diastolic pressure (EDP) was significantly increased after balloon occlusion (7.6 ± 4.0 mmHg vs.13.7 ± 5.0 mmHg, $p < 0.05$). There was significant positive correlation between the rate of change of EDV and that of EDP ($Y = 0.32 + 4.56X$, $R^2 = 0.464$, $p = 0.0073$).
Conclusion: These data suggested that the left ventricle in this setting were exposed to the acute volume over load with the elevation of EDP after transcatheter closure. Although left ventricle systolic and diastolic functions were well tolerated in our patient population, attention should be paid in patients with decreased LV function, such as patients with elder population or other cardiac complex.

83

Catheter therapy of pulmonary valve atresia with intact ventricular septum in neonates

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Background: Pulmonary valve atresia with intact ventricular septum (PAIVS) has a high early mortality of >35% in the first year of life. Establishing continuity between right ventricle and pulmonary artery and antegrade flow, either alone or combined with

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a systemic-pulmonary artery shunt, may promote growth of the right ventricle.
Patients and Methods: Catheter treatment with either laser or radiofrequency wires was attempted in 24 neonates. Their weight varied between 0.820 and 4.1 kg. The valve annulus was usually dilated to 100–120% of its size.
Results: There were 7 (29%) deaths, 5 of which occurred in the first 12 patients and 2 in the remaining 12 patients. There were 4 catheter related deaths, and 3 late deaths, of which 2 were considered to be due to overcirculation. Of the 17 survivors, 8 required additional arterial duct-related procedures up to 63 days after the initial procedure. Two of these had balloon dilation of the duct and the remaining 6 had stenting of the duct, 4 at the time of catheter valvotomy. During mean follow up of 3.6 years, range 3 months–11.7 years, 14 have achieved two-ventricle circulation and the remaining 3 have achieved 1½ ventricle circulation. Z-values for the tricuspid, pulmonary and mitral annulus diameters were measured from the echocardiograms. The pre-procedure tricuspid valve diameter was between 5 and 13 mm, mean 9.6 mm, the z-value between <7 and +2, and the pulmonary valve diameter was between 3 and 9 mm, mean 6.3 mm, z-value −5 and +0.8, mean −2.3. At the latest follow up, the diameter of the tricuspid valve was between 12.7 and 24.6, mean 18.6, and the z-value of tricuspid valve was −5.5 and +1.8, mean −3.1.
Conclusion: The results of catheter valvotomy for neonatal pulmonary valve atresia with intact ventricular septum have continued to improve over the years. With appropriate selection, two-ventricle circulation can be achieved in a majority of patients despite small right ventricles initially. Some patients require temporary additional pulmonary blood flow after the procedure. A small number of patients need 1½ ventricle approach.

84
Percutaneous pulmonary valve replacement in large right ventricular outflow tract: an experimental study

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Background: Percutaneous pulmonary valve replacement has recently opened new perspectives to transcatheter replacement of cardiac valves, but to date, this technique is limited to the small number of patients who have right ventricular outflow tract smaller than 22-mm of diameter. We report our initial experience of percutaneous pulmonary valve replacement in animals with large pulmonary trunk.
Methods and Results: Ewes with pulmonary trunk larger than 25-mm of diameter were divided in 2 groups. In the first, a newly designed self expandable stent containing an 18-mm bovine valve was implanted in pulmonary position. In the second, a non-valved stent was firstly placed into the pulmonary artery. An 18-mm valve mounted in a balloon expandable stent was delivered two months later into the previously placed device. Devices were successfully delivered and were perfectly functioning during the study. The downsize mechanism allowed the reduction of pulmonary diameter to 18-mm without major repercussion on the right ventricular function in any animals.
Conclusion: Non-surgical implantation of a pulmonary valve is possible in animals in all type of pulmonary trunk without regarding to its size. A downsize stent is obviously needed to allow implantation in large trunk. This technique should be feasible in humans in a very near future.

85
Percutaneous pulmonary valve implantation in humans – initial experience

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Objectives: Residual obstruction with or without regurgitation is the commonest indication for reoperation after congenital heart surgery involving the right ventricular outflow tract (RVOT). We report our experience of percutaneous valve implantation in pulmonary position in humans.
Background: Conventional treatment of residual lesions of the RVOT includes balloon dilatation with or without stenting for stenotic lesions, and surgical placement of a valved conduit for regurgitant or mixed lesions. Surgical treament is also used for failed catheter intervention for stenotic lesions or development or exacerbation of pulmonary regurgitation after balloon dilatation or stenting. The risk of re-operation and failure of growth in pediatric population makes surgical intervention unattractive.
Methods: 12 children and 3 adults with stenosis and/or insufficiency of the pulmonary graft underwent percutaneous implantation of a bovine jugular venous valve in pulmonary position. All patients had clear indications for surgery of the right ventricular outflow tract. 8 patients were in New York Heart Association (NYHA) functional class 3, and 4 patients in class 4. The mean age of children in the study group was 13.2 ± 2.4 years(mean ± SD).
Results: Percutaneous pulmonary valve (PPV) replacement was successful in all patients. The mean hospital stay was 36 hours. The peak systolic gradient 70 ± 18 mmHg and reduced to 37 ± 7 mmHg after the valve implantation. However, angiography and echocardiography after the procedure showed no significant regurgitation of the implanted valve. 8 patients had a residual gradient soon after procedure which decreased by a further 30% during the first follow-up visit, 2 weeks later. There were 3 adverse events. One patient required implantation of a second valved stent within the first to relieve significant residual obstruction during the same procedure, with a successful result. One patient had significant residual gradient requiring explantation of the valved stent with placement of a homograft. One patient had a late explantation for infective endocarditis resulting from unprotected dental procedure. All patients showed remarkable improvement in their effort tolerance and NYHA functional class.
Conclusions: Percutaneous pulmonary valve implantation is a safe and effective procedure to treat residual RVOT lesions. Furthermore, this new technique may have an important role in delaying or avoiding surgery.

86
New percutaneous technology of vascular shunting

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Purpose: To investigate the anatomical feasibility of creating percutaneous Vascular connections in an animal model.
Material and Methods: Connections between several medium size vessels were attempted in eleven Beagle dogs. Different approaches and devices were tested to best bridge the path between both vessels. In Bakuolev Scientific Center of Cardio-Vascular Surgery the catheter system for magnetomechanic positioning of a zone vascular bypass has been developed. The flexible kinematic needle located

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in catheter with a magnetic tip has a high degree of accuracy of prompting both magnetic catheters, and can be entered into nearby vessels (5–30 mm). The effective emission of a needle from a gleam of one vessel in a gleam adjusts another by the special terminator located on proximal end of a needle. Capture of a core of a needle allows to use it for the delivery of a stent graft by a coaxial system, and its accurate placement between connected vessels. The vascular connection was established with a prosthesis specially designed for this study.
Results: Among the 11 dogs used in this study, it was possible to establish an aorto-caval shunt in 2 dogs. Right atrium-right pulmonary artery shunt – 2 dogs and a superior vena cava-right pulmonary artery shunt were attempted in 3 dogs. Ascending aorta-truncus pulmonalis shunt were attempted in 2 dogs. Glenn shunt was performed in 1. In the last experiment percutaneous Fontan was attempted in one dog. The tract was stabilized with a polyurethane cone-shaped covered prosthesis. This prosthesis had a tubular configuration in its mid portion and flared ends which allowed an effective anchoring between two vessels without leaks.
Conclusions: Preliminary studies carried out in dogs open a new perspective of establishing a direct connection with prosthesis between two vessels. Although the technique needs to be improved, the targeting with flexible kinematic needle, Dormia basket and the use of magnetic devices on both vessels are promising elements to achieve this ambitious step.

87
Mitral valve repair in children with rheumatic valve disease – long term results

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Background: Mitral valve replacement for rheumatic valve disease in children is associated with a high complication rate both at surgery and during follow up. Mitral valve repair is therefore a preferred surgical modality. Immediate results have been promising, but only few long-term studies have been reported.
Material: Between January 1994 and December 2001, 117 children, 54 boys and 63 girls, with a mean age of 12.5 ± 2.5 years (6–15 years), underwent mitral valve repair due to rheumatic mitral valve disease. 97 of the patients (83%) had predominant mitral insufficiency. Twenty five patients (21%) had associated aortic valve insufficiency and 14 (12%) tricuspid valve insufficiency addressed during the same surgery. The mitral valve disease was corrected using various techniques described by Carpentier. In addition there were 24 aortic valve repairs and 1 aortic valve replacement with homograft. Tricuspid insufficiency was corrected with de Vega's technique.
Results: There were no hospital deaths or severe postoperative morbidity. The children were examined every 6 months. There were no late deaths during a mean follow up of 61 months (12–96 months). In one patient the repair failed and mitral valve replacement was performed 2 days later. Six patients developed recurrent mitral regurgitation, 4 requiring re-operation. Two had renewed repair and 2 had mitral valve replacement using bioprothesis. The recurrent mitral valve insufficiency/stenosis free survival was 95.7% at 5 years and 84.2% at 8 years respectively.
Conclusions: Mitral valve repair in children with rheumatic valve disease has excellent immediate results with low operative risk and satisfactory long term results and should therefore be the preferred treatment of choice.

88
Impact of associated cardiac lesions on outcomes of parachute mitral valve in 84 patients

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Parachute mitral valve (PMV) is defined as a unifocal attachment of the chordae to a single or dominant papillary muscle and may cause subvalvar obstruction. We sought to determine factors associated with outcomes in patients with PMV. All patients assessed with PMV from 1977 to 2001 were identified from databases, and those with atrioventricular septal defect were excluded. Demographic, anatomic, management and outcome data were collected, as well as serial echocardiographic measurements. N = 84 patients (64% male) presented at a median age of 3 days (range, birth to 5.4 years). Associated cardiac anomalies in 99% included aortic coarctation in 65%, ASD 55%, bicuspid aortic valve 50%, PDA 50%, VSD 49%, aortic valve stenosis 32%, subaortic stenosis 21%, hypoplasia of the LV 20%, supravalve mitral stenosis 7% and supravalve aortic stenosis in 1%, with complex anomalies (TGA, DORV, DILV) in 14%. Noncardiac anomalies were noted in 26%. There were 17 total deaths, with Kaplan Meier survival estimates of 82% at one year, 79% at 10 years and 74% at 20 years. The presence of LV hypoplasia ($p < 0.001$) and ASD ($p < 0.003$) were independently associated with poorer survival. No interventions were performed in 14 patients (6 died). Ten patients (2 died) had procedures towards an eventual Fontan. Of the remaining 60 patients (9 died), 11 had surgical mitral valvotomy (1 died), with subsequent mitral valve replacement in 2. Kaplan Meier estimates of freedom from mitral valvotomy in non-single ventricle pathway patients were 95% at age 6 months, 80% at 10 years and 72% at 20 years, and risk was independently increased with the absence of aortic coarctation ($p < 0.02$) and the presence of subaortic stenosis ($p < 0.04$). Repeated measures analysis of serial echo measurements showed no significant increase in peak or mean gradient of the PMV over time, but higher mean gradient was independently associated with the presence of supravalve mitral stenosis ($p < 0.001$), absence of ASD ($p < 0.04$), presence of VSD ($p < 0.02$) and subsequent mitral valvotomy ($p < 0.01$).
Conclusions: Outcomes for patients with PMV are dependent on the spectrum of associated cardiac lesions. The degree of mitral valve obstruction remains stable, and the majority will not require valvotomy.

89
Long-term experience with heart transplantation in children

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Despite successful short-term results, long-term prognosis of pediatric heart transplantation (HT) is still to be determined. This study was to review a fifty-year experience with pediatric HT, to assess current results and future outcome.
Methods: Data was collected from two French centers. Immunosuppression was achieved by ATG induction therapy associated with ciclosporine-based regimen. All patients underwent monthly non-invasive monitoring, including clinical examination, ECG and echocardiography and doppler measurements. Patients more than 10 years old underwent routine invasive endomyocardial biopsy. Yearly dobutamine-stress echocardiography and/or coronary angiography were performed for detection of graft coronary disease.

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Results: From 1987 to 2002, 82 patients aged 2 days to 18 years (median 10 years), underwent 83 HT; indications included 41 congenital diseases, 38 cardiomyopathies, 1 iterative HT and 3 miscellaneous. Follow-up ranged 9 months to 14.5 years (median 6 years). Survival was 80%, 75% and 70% at respectively 1, 5 and 10 years post-transplant. Survival rates increased up to 95% (5th year) and 80% (10th year) for patients who survived more than 1 year post-transplant. Twenty-nine deaths occurred: 13 early cases were due to pre-transplant visceral failure, 7 first-year deaths were related to acute rejection (AR) or infection and 9 late cases occurred from graft vasculopathy, AR or infection. Mean incidence of AR was 1.8 episode per patient, 50% occurring during the first 3 months; 42% were late AR due to non-compliance and commoner in adolescents; 93% of the AR episodes resolved. Eight graft coronary diseases were diagnosed (9.6%) and 2 led to death. Dobutamine-stress echocardiography was a sensitive non-invasive technique for detection of myocardial ischemia. Ten patients (12%) had significant renal function impairement (no dialysis). Creatinine clearance was lower in younger (<2 years old at HT) transplanted patients (65 ml/mn versus 85 ml/mn). Antihypertensive therapy was needed in 6 cases. Linear growth was significantly impaired in 2 patients with severe renal failure. Psychological difficulties occurred in 9 patients leading to non-compliance-related AR (11%). In summary, our long-term results of pediatric HT show good survival rates and quality of life; bad compliance, renal failure and graft coronary disease seem to be the main causes of long-term prognosis impairement.

90

The prevalence and manifestation of post transplantation lymphoproliferative disorder (PTLD) in paediatric heart transplant recipients

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Introduction: The reported prevalence of post-transplantation lymphoproliferative disorder (PTLD) in paediatric organ transplant recipients varied between 6% and 16%. Therefore, we evaluate the institutional incidence of PTLD in paediatric heart transplant recipients.
Patients and Methods: The records of 110 paediatric patients who underwent heart transplantation at a young age between 1990 and 2002 were reviewed with respect to the occurrence of PTLD and EBV status. Immunosuppressive therapy consisted of triple-drug immunosuppresion with cyclosporin or FK506, steroids, and azathioprine or mycophenolate mofetil (MMF). Specific anti-viral prophylaxis was performed in all EBV-sero-negative children.
Results: PTLD was diagnosed in 4 (3.6%) children with median age of 4 (range 1.3–13.3) years, after median post-transplant period of 3 years (range 4 months to 4.5 years). All of these patients received steroids, 3 set on cyclosporine and 1 on FK506. At the time of PTLD diagnosis 2 children received Azathioprine the other 2 received MMF. PTLD manifested predominantly as EBV-associated lymphoma: multiple abdominal nodules (n = 2), femoral lymphoma (n = 1), buccal lymphoma (n = 1). The initial imaging findings of these episodes included bowel wall thickening, abdominal lymphadenopathy, and focal tumour masses. Only one patient suffered one recurrent episode of the lymphoma in orbital and buccal region. All patients are now in full remission without death after treatment including reduction of the immunosuppressive, anti-viral and chemo-therapy. Surgical procedures to remove tumours were necessary in all 4 patients.
Conclusion: The manifestation of PTLD in young heart transplant recipients is predominately associated with EBV-infection. Close specific follow up and consequent anti-viral prophylaxis in EBV-sero-negative patients contributes to the relatively lower incidence of PTLD in our patients.

Session 11: Short Slide Presentations 1

91

Changing patterns of infective endocarditis in congenital heart diseases over the past decade

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The aim of the study was to assess changing patterns of infective endocarditis (IE) in children and adults with congenital heart diseases (CHD) over the past decade. From 1966 to 2001, 153 IE occurred: 81 (2.5 cases per year) in period I: <1990 and 72 (6.5 cases per year) in period II: >1990. TOF incidence decreased, unoperated VSD, Rastelli repair and palliated cyanotic CHD increased. Age was significantly higher in period II.Twenty-eight percent of the cases in group I and 20% in group II did receive appropriate prophylaxis. Non-compliance with protocols was more frequent in group II (15% versus 4%). Source of infection was more often identified in period II (73% versus 65%). Postoperative cases were more frequent in period I; dental cause was the second most frequent event in period I (20%) and the most frequent in period II (34%). Cutaneous causative infections increased from 5% to 17%. The proportion of negative blood culture lessened since the past decade, from 21% to 8% ($p = 0.026$). Streptococcus was the commonest causative organism in both periods. The incidence of severe heart failure decreased (20% to 4%) and cardiac complications lessened (31% to 18%). Early surgery was more frequently performed in period II (35% versus 28%) Overall mortality was 25% in period I and 8% in period II ($p = 0.064$). The incidence of IE-related deaths was higher in period I (11% versus 3% in period II, $p = 0.046$). Survival rates were similar (90% at 1 year, 80% and 90% at 10 years, $p = 0.33$). In summary, improvement of microbiological diagnosis and early surgical treatment allowed a significant decrease of mortality over the past decade. However, the incidence of IE did not decrease and the proportion of adults, of complex cyanotic diseases and of non-compliance to prophylaxis increased. Primary prophylaxis of IE should be improved.

92

Percutaneous pulmonary valve implantation for right ventricular outflow tract lesions after congenital heart surgery

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Objectives: Residual obstruction with or without regurgitation is the commonest indication for reoperation after congenital heart surgery involving the right ventricular outflow tract (RVOT). We report our experience of percutaneous pulmonary valve implantation in humans.
Background: Conventional treatment of residual lesions of the RVOT includes balloon dilatation with or without stenting for

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stenotic lesions, and surgical placement of a valved conduit for regurgitant or mixed lesions. Surgical treament is also used for failed catheter intervention for stenotic lesions or development or exacerbation of pulmonary regurgitation after balloon dilatation or stenting. The risk of re-operation and failure of growth in pediatric population makes surgical intervention unattractive.
Methods: 12 children and 3 adults with stenosis and/or insufficiency of the pulmonary graft underwent percutaneous implantation of a bovine jugular venous valve in pulmonary position. All patients had clear indications for surgery of the right ventricular outflow tract. 8 patients were in New York Heart Association (NYHA) functional class 3, and 4 patients in class 4.
Results: Percutaneous pulmonary valve (PPV) replacement was successful in all patients. The mean hospital stay was 36 hours. Angiography and echocardiography after the procedure showed no significant regurgitation of the implanted valve. 8 patients had a residual gradient soon after procedure which decreased by 30% during the first follow-up visit, 2 weeks later. There were 3 adverse events. One patient required implantation of a second valved stent within the first to relieve significant residual obstruction during the same procedure, with a successful result. One patient had significant residual gradient requiring explantation of the valved stent with placement of a homograft. One patient had a late explantation for infective endocarditis resulting from unprotected dental procedure. All patients showed remarkable improvement in their effort tolerance and NYHA functional class.
Conclusions: Percutaneous pulmonary valve implantation is a safe and effective procedure to treat residual RVOT lesions. Furthermore, this new technique may have an important role in delaying or avoiding surgery.

93
Enhanced visualisation of the right ventricle by contrast echocardiography in congenital heart disease

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Background: Contrast echocardiography improves endocardial border detection of the left ventricle. Whether this is true for the highly trabecularized right ventricle (RV) is unknown. Specific problems are poor visualisation of RV anterior wall mainly due to poor near field resolution and the complex geometry of the RV, which makes echocardiographic assessment of RV function hazardous. The aim of this study was to assess whether the use of contrast (SonovueTM) echocardiography has additional value in RV endocardial border visualisation (EBV), and whether it has impact on the echocardiographic judgement of the RV function.
Methods: Twenty adult patients, 10 with transposition of the great arteries and 10 with corrected tetralogy of Fallot, were imaged using two-dimensional echocardiography. Second harmonic imaging was compared with contrast echocardiography. Two independent observers analysed endocardial border visualisation (EBV) of 13 RV wall segments in each patient. EBV was graded for each wall segment from 0 to 3 (0 =not visible, 3 =optimal visualisation).
Results: Endocardial border visualisation improved in all patients with contrast echocardiography (mean EBV 2,13 ±0,75) compared to second harmonic imaging (mean EBV 1,00 ±0,77, $p < 0,0001$) (Figure 1). The benefit was most evident in the near field images, the apex in apical 4-chamber view and RV anterior wall in parasternal short-axis view. The advantage of contrast echocardiography was greatest in patients with poor imaging quality. In 55% of the patients visual estimation of RV function changed with use of contrast echocardiography.

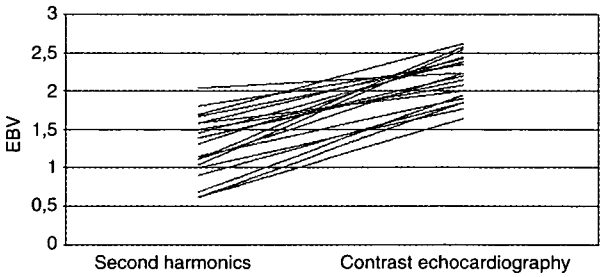

Figure 1. Endocardial border visualisation (EBV) for all patients. Mean endocardial border visualisation score for all RV segments for each patient.

Conclusion: The use of contrast echocardiography is superior to second harmonic imaging for endocardial border visualisation of the RV. Improved endocardial border visualisation may allow more accurate assessment of RV dimensions and function.

94
Left ventricular structure and function in adolescent swimmers

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Heart structure and function in adolescent swimmers could be affected by their long-term endurance training since young age.
Purpose: To evaluate left ventricular structure and function in adolescent swimmers.
Methods: Twenty eight swimmers between 14 and 17 years old (15 boys and 13 girls), training at least 12 hours/week for the last three years and 28 nontraining control subjects (14 boys and 14 girls) matched for age, weight and height were studied. Two-dimensional, M-mode and Doppler echocardiography were performed. Left ventricular internal diameters in diastole and systole (LVIDd and LVIDs, respectively), interventricular septum (IVS), left ventricular posterior wall (LVPW) were measured from M-mode echocardiography. Left ventricular mass (LVM), hypertrophy index (HI), shortening fraction (SF) and ejection fraction (EF) were calculated. Diastolic parameters including mitral valve inflow velocities and pulmonary vein flow were measured: maximal early (peak E wave), and late (peak A wave) mitral velocities, E/A ratio, isovolumic relaxation time (IVRT) and deceleration time (DT), pulmonary systolic (S), diastolic (D) and A wave velocities. Comparisons between the two groups were made using the independent student t-test.
Results: There were no significant anthropometric differences between the two groups. Compared with controls, male swimmers showed a significantly greater LVIDd (5.36 ± 0.43 vs 4.91 ± 0.43 cm, $p < 0,009$), a thicker IVS (0.79 ± 0.10 vs 0.65 ± 0,09 cm, $p < 0,0001$) and LVPW (0.79 ± 0.13 vs 0.64 ± 0.008, $p < 0,001$), an increased LVM (192,7 ± 54,7 g vs 128,7 ± 28,6 g, $p < 0,001$). Female swimmers had significant increased in LVIDd (4,8 ± 0,37 vs 4,5 ± 0,24 cm, $p < 0,007$), but no increase in IVS or LVPW thickness, neither LVM. SF and EF was similar in all groups. Transmitral inflow parameters were similar in both groups, except for a prolonged TD in the swimmers group (220 ± 47 vs 181 ± 20 ms, $p < 0,008$ in males and 227 ± 50 vs 177 ± 31 ms, $p < 0,004$ in females).
Conclusions: These findings suggest that long-term swimming in adolescents promotes increase in left ventricular size and mass with

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normal systolic and diastolic function. A prolonged decelaration time is characteristic of physiological left ventricular hypertrophy.

95

Value of the novel isovolumetric acceleration index in the evaluation of ventricular systolic function in patients following corrective surgery of TOF

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Objective: Following corrective surgery of tetralogy of Fallot evaluation of systolic function is difficult due disturbed geometry of both right and left ventricle. Recently, Vogel et al., described in experimental pigs a tissue Doppler derived myocardial acceleration index which is load independent and can assess systolic function of the ventricle without any geometrical assumption. Our study was carried out to evaluate the right and left ventricular systolic function in postoperative TOF using tissue Doppler and applying this novel parameter.
Material and Method: In 10 asymptomatic postoperative TOF patients and in 10 aged matched normal subjects, myocardial velocity of both ventricular free wall in the basal and middle regions were examined in the apical four chamber view. The beginning of the isovolumetric contraction time was determined from the myocardial velocity curve (nadir preceding the systolic wave) and confirmed by atrioventricular valve closure in the simultaneously taken two dimensional (2-D) loop. The end of this time was recognised by the small notch on the ascending limb of the acceleration velocity curve. The isovolumetric acceleration index was calculated via dividing the myocardial velocity by the isovolumetric contraction time.
Result: The index of both ventricles was age and heart rate independent. In the TOF group no significant correlation was found between the index and the gradient across the RVOT as well as the grade of pulmonary regurgitation. Regarding the RV the index was lower in the TOF group compared to normal in the basal region ($p = 0,038$) and in the basal segment ($p = 0,07$). The TOF group had significantly lower index in both LV basal ($p = 0,001$) and LV middle segments ($p = 0,0009$) compared to the normal group.
Conclusion: The tissue Doppler derived isovolumetric acceleration index is an easy, reproducible and bedside parameter to evaluate systolic ventricular function. In TOF both ventricles have altered systolic function. The clinical value of this parameter needs further comparative studies.

96

Outcome of transcatheter closure of muscular ventricular septal defects using the Amplatzer ventricular septal defect occluder

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Background: There are few reports in the literature of transcatheter closure of muscular ventricular septal defects (MVSDs) using the Amplatzer ventricular septal defect occluder (AVSDO) with encouraging initial results. In this report, we present further experience and intermediate- term outcome in 30 patients with single MVSDs who underwent transcatheter closure with the AVSDO.
Methods: Thirty patients, aged 4 months to 16 years, with MVSDs underwent transcatheter closure using the AVSDO. The device consists of two low profile disks made of Nitinol wire mesh with a 7 mm connecting waist. The prosthesis size (waist diameter) was selected to be equal to the balloon "stretched" diameter of the defect. A 7F to 8F sheath was used for the delivery of the AVSDO. Fluoroscopy and transesophageal echocardiography were used for the guidance of the procedure.
Results: The "stretched" diameter of the defect ranged from 6 to 14 mm. Complete occlusion of the communication occurred in 28/30 patients (93% closure rate). One patient (an infant 4-month-old) with sustained complete left bundle branch block after the procedure went on to developed complete heart block one year later. No other complications were observed during a mean follow-up of 2.2 years (0.25 to 4.5 years).
Conclusions: The AVSDO is an efficient prosthesis that can be safely used in the majority of patients with a single MVSD. Further studies and long-term follow-up are required before this technique enters routine clinical practice.

97

Early cardiopulmonary functional improvement following transcatheter atrial septal defect closure

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Background: Conflicting reports exist on short-term cardiopulmonary functional recovery in asymptomatic or mildly symptomatic patients after transcatheter closure of secundum atrial septal defect (ASD).
Methods: 21 consecutive patients (9 males; mean age 42.3 years, range 8–70) underwent spirometric and maximal cardiopulmonary testing the day before and at 6 months from transcatheter ASD closure. The cardiopulmonary test consisted in ergometer cycle exercise testing with breath-by-breath measurement of peak oxygen consumption (VO_2), carbon dioxide production and minute ventilation. Before closure, measurements were made of mean pulmonary artery pressure (mean 15.6 mmHg, range 9–25), pulmonary-to systemic flow ratio (QP/QS; mean 1.99, range 1.4–3.5) and ASD stretched diameter (mean 25 mm, range 18–31).
Results: Overall, peak VO_2 improved at 6 months from percutaneous ASD closure ($p < 0.0001$). The improvement was significant both in patients aged <40 years ($p = 0.023$) and in those aged >40 years ($p = 0.0017$); it was also significant both in patients with a QP/QS <2 ($p = 0.0064$) and those with QP/QS >2 ($p = 0.022$). No significant correlation was found between peak VO_2 variation and age at closure or QP/QS entity. Improvements in vital capacity ($p = 0.043$) and peak oxygen pulse ($p = 0.0032$) were also noted at 6 months. No significant change was noted in peak heart rate at 6 months from closure.
Conclusions: This study strongly suggests that transcatheter closure of ASD leads to a significant improvement in cardiopulmonary function within 6 months in most patients, irrespective of age at closure and shunt entity.

98

Comparison of surgical repair with balloon angioplasty for native coarctation in infants over 3 months of age

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Objectives: Comparison of surgical repair and balloon coarctation for native coarctation of a localized morphology is performed retrospectively.

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Background: Surgery and balloon angioplasty for coarctation of the aorta have shown comparable short-term results, but long-term follow-up remains unclear. To allow a valid comparison between both techniques, identical inclusion criteria were applied.
Methods: Results of surgery (group A, 90 patients, age 6.1 ± 5.2 years) and balloon angioplasty (group B, 28 patients, (age 3.9 ± 3.4 years) for isolated, native coarctation in children >3 months, performed in a 28-year-period, were compared. Additionally, resection and end-to-end repair (50) and patch angioplasty (36) were compared. Kaplan-Meier analysis was performed in all groups. Mean follow-up was 12 ± 7.0 years.
Results: Immediate success was 100% following surgery and 96% following balloon angioplasty. Mortality was 1%, following surgery. In group A, recoarctation occurred in 8 patients (9%), 6 following balloon angioplasty, 2 following patch angioplasty and in 2 (7%) following resection and end-to-end anastomosis in group B. Log rank analysis reveals a difference in favor of surgery ($p = 0.02$). No statistical difference was found for different types of surgery in respect to reinterventions ($p > 0,05$). Aneurysm formation was encountered once, following patch angioplasty.
Conclusions: Surgical repair of native coarctation yields less reinterventions than balloon angioplasty in comparable patients. End-to-end anastomosis and patch angioplasty yield comparable reintervention rates.

99
Accelerated transcatheter patch occlusion of large patent ductus arteriosus

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Large patent ductus arteriosus (PDA), is a challenging lesion to occlude by any device. The Transcatheter Patch (TP) can have a predictable full occlusion result (previous test balloon occlusion) and minimal risk of embolization (continuous out side control); furthermore a single device can accomodate various shapes and sizes of PDA by volume adjustment. Accelerated transcatheter patch application was used in the occlusion of 9 large PDAs. Preformed clot from the same patient was applied on the patch carrying balloon and no heparin was used during the procedure. In two of the cases there was history of failed attempts by other devices. PDA size varied from 7–21 mm (med.15), patient age from 3–19 years (med.13) and patient weight from 13–58 Kg (med.40). Seven of the PDAs were conical and two were short. The patches were introduced transvenously through 9–12F long sheaths and were released in 23 hours. All cases were successfully fully occluded. There were no cases of embolization, coarctation, left pulmonary artery stenosis or other complications either acutely or on follow-up. In conclusion accelerated TP occlusion of large PDAs appears effective and safe, with the patch released in 23 hours. Larger trials are justified.

100
Stent implantation into the arterial duct in lambs

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Stenting the arterial duct may realise a non-surgical, artificial Blalock-Taussig shunt. The purpose of this study was to establish the feasibility of this method using standard coronary stent in an animal model.

Methods: Twelve newborn lambs (weight: 1.8–3.5 kg) underwent stent implantation into the arterial duct. Three of them had previously undergone antenatal banding of pulmonary trunk. Implantation was realised via a femoral artery approach using a 5–6F sheath. Initial lateral aortography was performed to assess duct morphology and patency and duct was completely occluded in 10 lambs. Duct was initially crossed by a Radifocus guidewire replaced by 0.018 inch guidewire to place the stent mounted on a balloon catheter. Stent diameter was 3.5 to 6 mm, and length 12 to 18 mm. Two of them were covered stents (Jostent, Jomed). Implantation succeeded in 10. Failure occurred in 2 lambs: one haemopericardium and one embolisation due to a large duct. These 2 lambs and another one with a patent stented duct died. Autopsy of the 9 remaining lambs was performed 1 to 2 months after implantation. Duct patency was noticed in 7 with development of neointima within the stent. Metal strands not in contact with the duct wall were incompletely covered by neointima. Two complete ductal occlusions on the aortic side (unstented part of the duct) were observed. Use of standard coronary stents allows correct duct stenting in the neonatal period. This technique is effective and simple using a femoral artery approach. Major risks at implantation are duct perforation, embolisation and/or malposition of the stent. Need of covering the whole length of the duct is necessary to avoid future occlusion and repeat angiography 3 months after implantation is necessary to control stent patency and intimal proliferation. Duct stenting may be thus an alternative to surgical Blalock-Taussig shunt in future. Best stent type (balloon expandable, heat expandable, covered stent) and associated treatment (antiplatelet agent or anticoagulation treatment...) remain to be stated in clinical practice. This work was supported by a grant from the French Society of Cardiology and SESERAC.

101
Use of covered Cheatham-Platinum stents in aortic coarctation and recoarctation

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Covered stents have not been used routinely in congenital heart disease and their role is still to be defined. Theoretically they could be used for the purposes of baling out in patients developed aneurysms or dissections following an interventional procedure. The aim of this study is to report our initial experience with 4 patients (age range 12–19 yrs, weight range 45–94 kg), in whom custom-made covered Cheatham-Platinum (CP) stents were used. All the patients had aortic coarctation and were on antihypertensive treatment. Two pts had balloon dilation previously and both developed aortic aneurysms immediately after balloon dilation, 1 pt developed aortic recoarctation and recanalisation of the arterial duct after surgical treatment, 1 pt had coarctation with long segment hypoplasia of transverse arch. The covered CP stents (length 34–45 mm, diameters 14–22 mm) were deployed in the correct position in all pts. After implantation gradients decreased from 10–40 mmHg to 0–5 mm Hg. No complications were encountered. Angiography confirmed an excellent result with no opacification of the arterial duct and exclusion of the aneurysms. The antihypertensive treatment was continued in all the patients, but the dosage could be reduced with improved control of the blood pressure during follow up. CT angiography performed between 3 and 8 months after stent implantation showed good result (no aneurysm

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formation, wide patency of stents) in all pts. All the patients are well 4–16 mths after stenting. Covered CP stents may have an important role in dealing with aneurysm formation after previous surgery or balloon dilation of aortic coarctation. Furthermore in combination of coarctation of aorta patent ductus arteriosus can be treated successfully with covered CP stents. Careful follow up, especially with regard to neointimal formation, is needed to determine the role of covered CP stents in treatment of congenital heart disease.

102

Transcatheter treatment of aortic coarctation. Fifteen years experience

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From 1987 to 2002, 79 pts with aortic isthmus coarctation received transcatheter treatment. In 40 pts with post-operative recurrent coarctation (mean age 5 ± 6 years and weight 22 ± 12 Kg) and 27 pts with native coarctation (mean age 9 ± 8 years and weight 29 ± 7 Kg) balloon dilation was performed. In 15 pts (mean age 19 ± 17 and weight 49 ± 16), 6 with recoarctation and 9 with native coarctation, stents were implanted. Balloon angioplasty in recoarctation (40 pts): the mean peak pressure gradient (PG) changed from 38 ± 16 to 13 ± 11 mmHg ($p < 0.001$). The stenosis diameter (SD) and its ratio to the diameter of diaphragmatic aorta (SD/DA) increased from 5 ± 3 and 46 ± 17 to 8 ± 3 mm and 65 ± 19 respectively ($p < 0.001$). Seven procedures (17%) were unsuccessful: 2 pts underwent surgery, 1 stent implant and 4 are waiting for stent. Of 6 pts (15%) with restenosis, 1 underwent surgery, 5 repeated angioplasty. The mean follow-up period is 47 ± 45 months. The aortic isthmus doppler PG is 14 ± 12 mmHg. Balloon angioplasty in native coarctation (27 pts): the PG across the stenosis changed from 34 ± 13 to 15 ± 15 mmHg ($p < 0.001$). The SD and SD/DA increased from 6 ± 3 and 42 ± 13 to 10 ± 4 mm and 66 ± 16 ($p < 0.001$). Nine procedures (33%) were unsuccessful: 5 pts underwent surgery, 1 stent implant and 3 are waiting for stent. There were 3 restenosis (11%): 2 treated with balloon dilation and 1 with stent. The mean follow-up period is 21 ± 25 months: the aortic isthmus doppler PG is 12 ± 12 mmHg. Stents implant (15 pts): 10 Palmaz-Shatz and 5 CP stents were implanted. The PG changed from 36 ± 24 to 2 ± 4 mmHg. The SD and SD/DA increased from 8 ± 3 and 43 ± 15 to 15 ± 3 mm and 78 ± 19 respectevely ($p < 0.001$). The implant was successful in all cases. The mean follow-up period is 18 ± 20 months: in 6 pts hemodynamic study 1 year after demonstrated persistent good result. The Doppler PG in 15 pts is 8 ± 5 mmHg; In conclusion 61 pts (77%), 35 with recoarctation and 26 with native coarctation, were successfully treated with angioplasty and/or stent. Surgery was necessary in 8 pts (10%). Transcatheter treatment is an effective alternative to surgery both in recoarctation and in native coarctation.

103

Problems and complications during transcatheter closure of the moderate and large patent ductus arteriosus utilizing Amplatzer duct occluder in 22 patients

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Background: The Amplatzer duct occluder (ADO) was introduced for transcatheter closure of the large PDA as an alternative to surgical ligation.

Aim of the Study: Our aim was to report complications and problems which occured while using this device, especially in the young age group.

Methods and Patients: Between July 2001 and September 2002 at total of 22 patients (14 females, 8 males) underwent attempt of closing their PDA using an ADO. The median age of these patients was 2.2 years and the mean weight was 12.1 kg. Eleven patients (50%) weighed less than 10 kg. The only patient who failed the procedure weighed 8 kg.

Results: The narrowest diameter of the PDA, obtained from the aortogram was a mean of 5.12 ± 2 mm (range 3.4–11.1 mm). The procedure was successful in all but one patient (95%). A total of 16/21 (76%) who had successful implantation left the hospital with complete occlusion.

Issues Pertinent to Procedure: Mean number of angiograms performed to visualize the PDA before implantation was 1.8 (range 1–7). Fluoroscopy time was a mean of 17 minutes and procedure time a mean of 104 minutes.

Problems and Complications: On seven occasions 7/22 (32%) problems were encountered. The device was wasted in two patients. Pull through of device into the pulmonary artery occurred in three patients. Kinking of the Mullins sheath and inability to retrieve a device in one, excessive bleeding requiring transfusion in another. All but one patient with complications weighed less than 10 kg.

Follow-up: At mid term follow up, 95% had eventually complete occlusion. In the remaining patient ($n = 1$) there was only an insignificant residual shunt. There was no increase in left pulmonary artery or descending aortic flow at short-term follow up.

Conclusions: Transcatheter occlusion of PDA by ADO has a high complete occlusion rate and is effective in PDA up to a narrowest diameter of 11 mm. Especially in the young age group (<10 kg), a complication rate of over 30% can be encountered in the learning phase. Poor visualization of the large PDA can explain some of the potential problems.

104

Left-sided atrioventricular valve insufficiency after AVSD correction

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Introduction: Left-sided atrioventricular valve regurgitation (LAVVR) is the main indication for reoperation in patients after repair of atrioventricular septal defect (AVSD). We sought to determine the risk factors for the development of severe LAVVR and for reoperation, and tried to identify the most appropriate strategy for patients with severe LAVVR.

Methods: Retrospective review of clinical, operative, and echocardiographic data were performed. From 1990 until 2001 164 patients, aged less than 16 years, underwent correction of their AVSD. Six Patients died in the immediate postoperative period, and 2 patients were lost to follow-up. Of the remaining 156 patients 110 patients (71%) had a complete AVSD, and 46 patients (29%) had a partial AVSD. Ninety-four patients (60%) had Down syndrome.

Results: During follow-up (9 months to 12 years; median 6 years), 30 patients (19%) developed severe LAVVR. Sixteen of these patients had severe LAVVR in the immediate postoperative period. Of these 16 patients 4 patients showed spontaneous regression to near-normal valve function during follow-up. The other 14 patients developed severe LAVVR during further follow-up. Of these 14 patients, 8 remained stable with medication only. Sixteen out of 30 patients with severe LAVVR were reoperated. Of these

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16 patients 11 underwent valvuloplasty of the mitral valve once, in 2 patients valvuloplasty was necessary twice, in 2 patients valvuloplasty was followed by mitral valve replacement, and one patient underwent primary valve replacement. Most reoperations (94%) were performed within 3 years after AVSD correction. After reoperation 3/16 (19%) patients died, all 3 because of severe congestive heart failure related to persistent mitral insufficiency. Risk factors for the development of severe LAVVR and reoperation after the primary operation were severe preoperative LAVVR and when no valvuloplasty was performed.
Conclusions: Severe LAVVR develops in a significant number of patients (19%) after correction of AVSD. The main risk factor is the presence of preoperative severe LAVVR. Although reoperation can be performed with an acceptable risk and usually results in good valve function, spontaneous regression after the immediate postoperative period should be waited for. In a considerable number of patients (8/14; 57%) reoperation can be postponed by medical treatment.

105

Pediatric cardiac surgery risk stratification: The Pediatric Cardiac Care Consortium (PCCC) categories reflect mortality and length of stay in a large German unit

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Background: A procedure and age based risk classification in pediatric cardiac surgery has been developed by the JCCC (4370 operations) and was described by C. Jenkins et al in the JTCVS 1/2002. We compare their classification results to our patients and investigate length of stay in the different groups.
Patients and Methods: 2111 procedures for correction of congenital cardiac malformations in patients under 16 years were performed consecutively between 7/96 and 10/02 in our center. They were classified by the described method in 6 risk categories (1 = e.g. ASDII, PDA > 30 days, 6 = DKS or Norwood Stage I operation). Risk group distribution in our/the PCCC population was 1:318/964, 2:733/1433, 3:659/1523, 4:267/276, 4/4, 130/168. In our patients/the PCCC group, 18.8%/19.2% were under 1 month, 37.5%/31.6% 1–12 months of age, respectively.
Results: Hospital mortality (%) in our population/the PCCC Group was 0.3/0.4, 4.2/3.8, 5.2/8.5, 10.1/19.4, 50.0/0, 40.8/47.7% in risk Group 1 to 6, respectively. Mean pre- and postoperative duration of stay was 14.2, 25.0, 29.6, 33.5, 35.8, 41.6 days. The average postoperative length of stay in surviving patients was 9.5 days longer than in non-survivers. This is mostly due to patients of class 3–6.
Conclusion: The classification proposed by C. Jenkins et al might be applicable to European pediatric populations, too. The applied stratification yields very similar results in the US and in our large center concerning distribution and outcome. The higher mortality risk coincides with a higher average length of hospital stay.

106

Postoperative course and growth after extracardiac Fontan operation in children under 4 years of age

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Objectives: It is hypothesized that the early removal of cyanosis and volume overload in functionally single ventricle could be beneficial for the postoperative course. We analyzed our early and medium-term experience with the extracardiac Fontan operation (ECFO) in children under 4 years of age regarding the incidence of postoperative complications and somatomotoric development after the early palliation.
Methods: Between 11/95 and 2/02 of the total of 56 ECFOs, 30 were performed in children aged 1.5 to 4.0 (median 3.0) years. The median body weight at birth was 3.2 kg (30th percentile) and at the time of the bi-directional cavo-pulmonary shunt 8.7 kg (10th percentile, n = 28). Twelve patients retained an aorto-pulmonary shunt additionally to the cavo-pulmonary shunt until ECFO. The body weight with median of 13.5 kg (range: 8.8–17.5 kg) was preoperatively in the 25th percentile. ECFO was established using Gore-Tex® conduit with median diameter of 20 (range: 16–24) mm in twenty-four children on the beating heart with normothermic cardiopulmonary bypass, and in 6 on cardioplegia and mild hypothermia.
Results: There were no differences in the preoperative body weight in patients who had staged aortopulmonary and cavopulmonary shunt or both the shunts until Fontan procedure. After the Fontan operation there was no perioperative death. The median duration of mechanical ventilation was 17 hours, median hospital stay 19 days. Eight patients (26%) developed pleural effusions lasting longer than 10 days. Of 28 patients with preoperative sinus rhythm 27 retained it postoperatively. No new onset arrhythmias and no late deaths were observed during the median follow up of 4.0 years. At the end of the follow up the median body weight was in the 50th percentile.
Conclusions: The extracardiac Fontan operation could be performed in small children with low mortality and morbidity and especially with low incidence of postoperative arrhythmias. In nearly all patients a conduit with the diameter sufficient for adults can be implanted. An accelerated growth of the children is to be observed after the completion of the Fontan circulation.

107

A new clip device for construction of vascular interrupted anastomosis in congenital cardiac surgery

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The U-clips, a new mechanical vascular anastomotic device developed for CABG surgery, allow precise quick construction of any vascular interrupted anastomosis, preserving pulsatility and growth potential. We report the first use worldwide of U-clips in 10 pediatric patients (mean age 2.4 ± 1.7 years) between July 2001 and July 2002 for coarctation repair (3), bi-directional Glenn shunt (5), Blalock-Taussig Shunt (1) and arterial switch (1). Median operation time was 207 ± 26 (105–365) min, CPB time 83 ± 19 (0–135) min, aortic cross-clamp (AXC) time 40 ± 10 (0–90) min and postoperative length of stay 8 ± 6 (6–61) days. For coarctation repair (2 end-to-end and 1 extended end-to-end anastomosis) U-clip size 50 was used and AXC was 31, 20 and 32 min, respectively. U-clip size 35 was used for the Glenn anastomosis. Device handling, primary hemostasis and patency were excellent and no device-related complication occurred. During a follow-up time of 11 ± 4.4 months the child with PAIVS and BTS died, all others are doing well. Echocardiographic assessments demonstrate excellent conditions of all anastomoses without growth restriction. We conclude, that U-clips may be a valuable alternative to

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conventional suturing in the construction of vascular anastomoses in congenital cardiac surgery.

108
Blood cardioplegia protects myocardial metabolism and function better than crystalloid cardioplegia in pediatric heart surgery

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Objective: We hypothesised that cold blood cardioplegia protects myocardial metabolism and function better than is achieved with St Thomas' crystalloid cardioplegia.
Patients and Methods: 30 consecutive children with complete AV-defects were randomly allocated to either blood or crystalloid intermittent cold (6 C) cardioplegia. The coronary sinus was cannulated for blood sampling of lactate and oxygen. LV ejection fraction was determined by echocardiography (single-plane Simpson). Measurements were performed after weaning off bypass.
Results: A significant release of lactate in the coronary sinus was observed after crystalloid cardioplegia but not after blood cp (arterial – coronary sinus lactate concentration -0.55 ± 0.15 vs -0.03 ± 0.04 mM/L, $p = 0.001$). Coronary sinus blood oxygen saturation did not differ significantly, 43 ± 4 vs $52 \pm 4\%$, $p = 0.100$). LV ejection fraction was lower after crystalloid than after blood cardioplegia (0.45 ± 0.05 vs. 0.57 ± 0.03, $p = 0.036$). A negative correlation was found between oxygen saturation of coronary sinus blood and lactate release, indicating that low oxygen saturation was associated with a high release and vice versa ($r = 0.63$, $p = 0.001$). Lactate release and impairment of LV ejection fraction were related ($r = 0.54$, $p = 0.004$).
Conclusions: This study indicates that blood cardioplegia protects myocardial metabolism and function better than crystalloid cardioplegia. The correlation between metabolic and functional parameters suggests a causal relationship, possibly linked to aerobic metabolism.

109
One stage correction for aortic arch obstruction and ventricular septal defect

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Objective: To evaluate current outcome and reoperation rate after one-staged correction strategy for ventricular septal defect (VSD) combined with interrupted aortic arch (IAA) or aortic coarctation (CoA) adopted since 11/1999.
Methods: From 11/99 until 08/02 22 consecutive patients underwent definitive repair of aortic arch obstruction and VSD. There were 11 patients with CoA and 11 with IAA. Mean age at operation was 12 days (3–92d), mean weight 3.6 kg (2,1–7.3). Body weight of 8 patients was below 2,5 kg. Associated anomalies in the IAA group included transposition of the great arteries (d-TGA), aorto-pulmonary window, and persistent truncus arteriosus, one patient each. Three patients with IAA and 2 with CoA had additional left ventricular outflow tract obstruction (LVOTO). Subaortic muscle resection was performed in two patients. A selective brain perfusion via the innominate artery was used and an extended aortic arch anastomosis established in all.
Results: Mean cross-clamp time was $72 + 32$ min, bypass time $161 + 47$ min, lowest temperature $22,8 + 4,3$°C, circulatory arrest with selective brain perfusion $34 + 13$ min. Fourteen patients required delayed sternal closure, which was performed at median 1.5 days (1–5 days) postoperatively. There were no early deaths. Early neurological complications were not noted. Follow up is complete, mean time is $1.5 + 0.8$ years. There is no late mortality. There are no residual VSDs. The aortic arch anastomosis remained unobstructed in 18 (82%) patients. Four patients (18%) (all IAA group) developed recoarctation. One patient required stent implantation, balloon dilatation was successful in the other three. The pressure gradients across the anastomosis are now less than 10 mmHg in all. The patient with d-TGA developed progressive right ventricular outflow tract obstruction requiring surgical reconstruction seven months later. All 22 patients are asymptomatic and develop normally.
Conclusion: One stage complete correction is feasible with good surgical and functional outcome in newborns with VSD and aortic arch obstruction. Recoarctation occurred in about 20% but can be treated interventionally. Selective brain perfusion during the aortic arch reconstruction reduces the early postoperative morbidity and seems to avoid neurological complications, but dedicated and comparative neurological follow up is necessary to investigate the long term outcome.

110
Long-term mortality and complication rate in children with mechanical prosthesis: a 30 years single center experience

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Background: The aim of this study is to assess the long-term mortality and the late outcome after cardiac valves replacement by mechanical prosthesis in children.
Methods: Retrospective study; setting: tertiary referral center. Patients: Between 1972 and 2002, 92 young patients, 46,7% female, mean age $8,25 \pm 4,9$ years (range 8 months to 16 years) underwent mechanical valve replacement in our institution. Abnormal preoperative findings included congenital (72%), degenerative (8%), rheumatic (14%), infective (5%) and iatrogenic (1%) valvular diseases. Surgery was proposed for mitral disease in absence of atrioventricular septal defect (AVSD) (group A, $n = 37$), mitral disease in relation with AVSD (group B, $n = 27$) or aortic disease (group C, $n = 28$). Mean follow-up was $9.94 \pm 5,4$ years (range 3 months to 22 years).
Results: Long-term mortality was 9% in group A, 38% in group B ($p < 0,001$ vs group A) with a strong hospital death (70%) and 8% in group C ($p =$ ns versus group A, $p < 0,001$ vs group B). Re-intervention was necessary in 10% of group A, 19% of group B and 5% of group C ($p < 0,05$ vs group A and B). A patient/prosthesis mismatch was responsible of 50% of re-intervention in group A and B but none in group C. In aortic position adult size prosthesis could always be implanted. Other severe complications (complete heart block, transient stroke, valve thrombosis without re-intervention) were observed in 7,2% of group A, 7,8% of group B and 3,2% of group C patients ($p < 0,05$ vs group A and B). No infective endocarditis or major haemorrhagic events were observed.
Conclusion: In young patients with mechanical prosthesis, the prognosis and the long-term mortality depend more on the presence of an extra-valvular disease (ASVD) than on the localisation of the valve replaced. Nevertheless, the re-intervention and complications rates are more important in mitral replacement. The low rate of mortality, re-intervention and complication for aortic replacement by mechanical prosthesis make it a reliable technical for aortic dysfunction in children.

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111

Transvenous pacemaker implantation for sinus node dysfunction and arrhythmias after the Fontan operation

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After a Fontan operation, sinus node dysfunction may occur as a result of extensive atrial surgery and right atrial distension. Loss of sinus rhythm with an inadequate nodal escape rhythm leads to poor exercise tolerance. When associated with recurrent intra-atrial re-entry tachycardias (IART) this leads to poor ventricular function and thrombus formation. Epicardial pacemaker implantation requires repeat thoracotomy and scarring from previous surgery leads to high thresholds. Steroid eluting transvenous leads allow low thresholds but there are concerns about thrombus formation. Transvenous pacemakers were used in 11 Fontan patients aged 3.6–33.7 years median 11 (double inlet left ventricle 5, tricuspid atresia 4, hypoplastic left heart syndrome 1 and cTGA with pulmonary atresia 1). 3 patients were limited by a poor chronotropic response and had AAIR pacemakers. 8 patients had tachyarrhythmias (IART in 6 and atrial fibrillation in 2) and were on anti-arrhythmic drugs that were not controlling their arrhythmia satisfactorily or producing excessive bradycardias: 3 patients had AAIR pacemakers for drug induced bradycardias, 1 had an AOOR pacemaker to underdrive terminate drug refractory IART, 1 had a BiAtrial AAIR pacemaker for paroxysmal atrial fibrillation and 3 had antitachycardia DDDR pacemakers. In the latter 3 patients an endocardial ventricular lead was placed via the coronary sinus in 1 and via perforation of the atrial baffle in 2. In two patients a large atrial clot had developed and persisted despite anticoagulation. Cardioversion to sinus rhythm followed by pacemaker implantation resulted in resolution of the thrombus. Cerebral embolism complicated a paroxysm of atrial fibrillation in 1 child prior to pacemaker implantation. Follow up ranges from 3 months to 6 years. All patients with a systemic ventricular lead or previous thrombus or poor ventricular function were maintained on Warfarin but in 4 patients only aspirin was used. Exercise tolerance improved in those with chronotropic incompetence and arrhythmia control was excellent in all but one in whom the antitachycardia algorithm was ineffective. Transvenous pacemaker implantation using various pacing modalities is able to deliver considerable benefit in the setting of sinus node dysfunction and arrhythmias after the Fontan operation.

Session 12: Short Slide Presentations 2

112

Magnetic Resonance Imaging in children with ventricular arrhythmias

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The aim of the study was to: 1. Evaluate results of magnetic resonance imaging (MRI) of the heart in 52 children with ventricular arrhythmias (VA). 2. Compare clinical characteristics of VA in pts with normal and abnormal MRI. All 52 pts underwent complex cardiological examination: routine, signal-averaged (SAECG), exercise and 24-hour Holter ecg, echocardiography, MRI, radionuclide evaluation of right (RV) and left ventricular (LV) ejecton fraction (EF). In echocardiographic examination 46 pts had structurally normal heart, in 8 pts mitral valve prolapse (MVP) was diagnosed. Material was divided into 2 groups: MRI(−) −40 pts (77%) with normal MRI; MRI(+) −12 pts (23%) with small single (9pts) or diffuse focuses of T1- hyperintensive signal which may represent fatty tissue replacement (FTR) in RV wall. Endomyocardial biopsy (EMB) was performed in 5 pts from MRI(+) nad 12 from MRI(−) pts. Only in 3 pts (3/12 =25%) from MRI(+) group with diffuse FTR fulfilled clinical criteria for arrhythmogenic right ventricular cardiomyopathy (ARVC) and FTR was confirmed by EMB. Biopsy proven myocarditis had 2 pts from MRI(+) and 13 from MRI(−) group. There was no statistically significant difference between the MRI(+) and MRI(−) pts in age at control examinations (14,5 vs 14,4 yrs), follow-up (6,4 vs 5,8 yrs), incidence of VT (58 vs 52%), symptoms (50 vs 29,5%), MVP (17 vs 15%), QTd time (28 vs 17 ms), abnormal SAECG (33 vs 25%), LV EF (54,5 vs 56%), RV EF (53,2 vs 53%), VA as % of total beats at first (10 vs 28,8%) and last (9,7 vs 10,6, p =0,09) Holter ecg, VA morphology (mono vs polymorphic, LBBB vs RBBB). In MRI(+) pts VA were rarely stopped during exercise test (2,5 vs 18%, $p > 0,05$) and antiarrhythmic drugs were more often used to control VA (50 vs 20%, $p < 0,05$). During observation there was no death and VA disappeared in 25% of pts from MRI(+) and 30% from MRI(−) group ($p < 0,05$).
Conclusions: 1. MRI suggested focuses of FTR in RV wall in 23% of children with VA, but only 25% of these pts fulfilled criteria for ARVC diagnosis. 2. Prognosis in VA with small single focuses of FTR in MRI seems to be good.

113

Pharmacological treatment of young children with permanent junctional reciprocating tachycardia: follow-up data of tachycardia and cardiac function

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The objective of the study was to assess the efficacy of pharmacological treatment, mainly with propafenone, to reduce the incidence of PJRT or to bring the 24 hours mean heart rate to a normal level in young children with PJRT. In this retrospective study 21 children with PJRT were enrolled (1990–2001). Follow-up visits were made at least every 6 months. Presence of PJRT per 24 hours and mean heart rate over 24 hours including PJRT on the Holter recording were registered. Mean heart rates over 24 hours of 1004 otherwise healthy children were used as reference values. Cardiac function was assessed by echocardiography. Pharmacological treatment was started with propafenone alone or in combination with digoxin as the first choice. Pharmacological treatment was considered effective if the outcome was either complete disappearance of PJRT after discontinuation of medication, or sinus rhythm with medication; partially effective if the 24 hours mean heart rate on the last Holter recording was <1SD above normal; and not effective in the remaining cases. At diagnosis children were 0.05 (0–0.5) years of age; two children had abnormal LV function. All 21 children had a normal LV function at the end of follow-up. Overall duration of follow-up was 2.4 (0.9–4.9) years. PJRT had disappeared spontaneously in 7 of 21 (33%) children: 5 of them were <1 year old. Propafenone alone (n =3) or in combination with digoxin (n = 13) was used most frequently (n = 16; 76%). In 12 (75%), 2 (13%) and 2 (13%) cases this treatment was effective, partially effective and not effective, respectively. Efficacy for all drugs was 67% (n = 14), 20% (n = 4) and 14% (n = 3), respectively. Adverse effects (n = 5) were mild or asymptomatic. No signs of proarrhythmia were registered. Pharmacological

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treatment with propafenone either alone or in combination with digoxin is safe and effective in children with PJRT. The mean heart rate is brought to a normal level and cardiac function is restored and preserved. RFCA may be delayed to a safer age and spontaneous disappearance (33%) may be awaited.

114
Tissue factor expression is promoted by thrombin via Rac and the p21-activated kinase PAK in pulmonary artery smooth muscle cells

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A prothrombotic state is frequently associated with pulmonary hypertension and may promote 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 are not well understood. Here we investigated the role of the GTPase Rac and its downstream target p21-activated kinase (PAK) in the regulation of TF expression by thrombin in human pulmonary artery smooth muscle cells (PASMC). Thrombin increased TF mRNA expression peaking within 1 to 4 h and TF protein expression peaking at 16 h. Thrombin stimulation also resulted in a rapid, but transient phosphorylation of PAK, which preceded the activation of protein kinase B (PKB), p38 MAP kinase (p38MAPK) and ERK1/2. Overexpression of a hyperactive PAK increased thrombin-induced TF expression whereas expression of a kinase-deficient PAK abrogated this response. Furthermore, activation of PKB, p38MAPK and ERK1/2 by thrombin was abolished in cells expressing the kinase-deficient PAK. These PAK-regulated pathways were dependent on Rac, since expression of the constitutively active RacG12V increased activation of PAK, PKB, p38MAPK and ERK1/2 and the upregulation of TF by thrombin, whereas these responses were attenuated in cells expressing dominant-negative RacT17N. Overexpression of a constitutively active PKB enhanced thrombin-dependent TF expression whereas TF levels were abrogated in cells expressing a kinase-deficient PKB. Treatment with the p38MAPK inhibitor SB203580 diminished thrombin-stimulated TF expression, whereas the inhibitor of the ERK1/2 pathway, PD98059, was not effective. These findings show that PAK is a downstream target of Rac in PASMC and controls thrombin-induced TF expression via two pathways involving either the activation of PKB or the activation of p38MAPK. Thus, PAK may play an important role in promoting the prothrombotic state and vascular remodeling in pulmonary hypertension.

115
Differential proteomic profiling of right ventricle after 3 weeks of RV pressure overload in young rats

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Background: RV failure is an important problem in congenital heart disease. To understand the mechanisms involved in the development of RV hypertrophy and failure, we performed differential protein profiling in rats with RV pressure overload. We report the results after 3 weeks of pulmonary artery banding (PAB).

Methods: Male Wistar rats (age 8.1 ± 0.4 wk) underwent PAB (n = 8) or sham operation (n = 6). For proteomic analysis, homogenates of RV myocardium containing equal amounts of protein from each rat with similar treatment (ie PAB or sham) were mixed. Next, these mixtures were subfractionated in cytoplasmic and myofilament preparations and separated by two-dimensional electroforesis (2-DE, pH 3–10 and 10–250 kDa). PDQUEST was used for image-analysis of the Coomassie blue-stained gels. For quantitative comparison of the measured intensities of protein species in two corresponding gels, we used about 250 proteins that were matched. Spots that differed in their relative intensity more than 2-fold were excised and digested by trypsin for analysis by MALDI/TOF mass spectrometry. The tryptic masses were then evaluated and identified using the peptide-mass fingerprinting search engine Mascot.
Results: After 3 wk, RV pressure (under anaesthesia) in PAB was increased (38 ± 5 vs 18 ± 1 mmHg, $p < 0.01$), without alterations in right atrial (RA) and aortic blood pressures, and heart rate. Body weight (BW) and weight gain were similar in PAB and sham. In PAB rats the ratio of cardiac weight to BW was increased for RA/BW (0.25 ± 0.02 vs 0.10 ± 0.01, $p < 0.01$), RV/BW (1.29 ± 0.05 vs 0.7 ± 0.04, $p < 0.01$) and for LV/BW (1.58 ± 0.05 vs 1.32 ± 0.03, $p < 0.05$). In PAB rats, we identified altered expression of cytoskeletal proteins (increase of skeletal a-actin, decrease of Cypher-2), decreased expression of three β-oxidation enzymes, increased expression of two glycolytic enzymes (energy metabolism) and decreased expression of stress-proteins (HSP27, dna K-type chaperone).
Conclusions: Three weeks PAB induces increased RV pressures and accordingly RV and RA hypertrophy. The RV hypertrophy could be associated with alterations in cytoskeletal, metabolic and stress-related protein expression in RV myocardium. Delineation of alterations in protein expression in relation to time, severity of overload and myocardial performance, is subject for further investigation and may identify future therapeutic targets.

116
N-terminal pro-B-type natriuretic peptide. Normal plasma levels from birth to adolescence. Elevated levels at birth, and in patients with heart disease

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Background: The natriuretic peptides are secreted from the heart in response to volume load and cardiac dysfunction. N-terminal pro-B-type natriuretic peptide (NBNP) is secreted along with BNP. In adults, NBNP is a marker for cardiac dysfunction. Little is known on NBNP levels in the pediatric age group.
Aim: To determine plasma levels of NBNP in newborns, infants and children in health, and to evaluate NBNP levels in heart disease.
Methods: Plasma NBNP levels were measured in 45 healthy infants and children and in umbilical cord blood of 31 newborns. NBNP was also measured in children with heart diseases, including 5 with left to right shunt, 4 with right ventricular volume overload and 6 with dilated cardiomyopathy.
Results: NBNP levels (median, pg/ml) were significantly elevated in cord blood (453) and in the first days of life (614) but did not change significantly with age from 4 months to 15 years (78). NBNP levels were significantly elevated in patients with left to right shunt (1143), right ventricular volume overload (429) and dilated cardiomyopathy (3138).

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Conclusions: NBNP levels are elevated in the first days of life and are stable from age 4 months to adolescence. NBNP levels reflect cardiac volume overload and dysfunction in infants and children.

117
Doppler evaluation of cardiac function in trisomy 21 and normal fetuses prior to 14 weeks of gestation

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Ultrasound measurement of nuchal translucency thickness (NT) at 11–14 weeks of gestation identifies fetuses at increased risk for both chromosomal abnormality and isolated heart defects. We hypothesised that subtle abnormalities in cardiac function and loading, might account for the increased NT in both. Fetuses underwent initial echocardiography at the time of NT evaluation, prior to karyotyping. Doppler studies were attempted in 163 fetuses subsequently shown to have trisomy 21(mean NT 6.0 mm, crown rump length (CRL) 64.9 mm). These were compared with one control group consisting of 104 fetuses with normal NT (NT 1.6, CRL 64.4 mm) and another of 264 fetuses with increased NT (NT 5.1, CRL 62.5 mm) but no chromosomal or other abnormality. Left and right atrioventricular valve E/A ratios and left and right heart Tei indices were calculated. The presence or absence of significant atrioventricular valve regurgitation was noted. The ductus venosus (DV) a-wave was classified as positive, absent or reversed. As shown in the table, there was no significant difference in left Tei index in trisomy 21(T21) compared with either control group. However, the right Tei index was significantly lower T21 fetuses compared with each control group. E/A ratios for both atrioventricular valves were significantly decreased in T21 fetuses compared with both control groups. Significant tricuspid regurgitation occurred in 57% of the trisomy 21 fetuses but was not found in normal fetuses. The prevalence of tricuspid regurgitation in T21 was 13 times greater than with increased NT alone and reduced only to 46% when 34 T21 fetuses with atrioventricular septal defect were excluded from the analysis. An absent or reversed DV a-wave was 2.7 times more likely in T21 fetuses than otherwise normal fetuses with increased NT and 4.9 times more likely than in normal fetuses. In fetuses with T21, tricuspid regurgitation was unrelated to abnormal DV flow. Changes in Tei index and E/A ratio in T21 were in the opposite direction to that expected with impaired myocardial function and reduced ventricular compliance. The findings in T21 fetuses would be better explained by increased intravascular volume and preload than by impaired cardiac function.

	Lt Tei ind	Rt Tei ind	Lt E/A	Rt E/A	Tric Reg.	Abn DV a
Trisomy 21	0.361	0.321	0.583	0.610	57%	74%
	n = 53	n = 51	n = 114	n = 118	(78/136)	(10/68)
Normal	0.365	0.382*	0.549*	0.583*	0%*	15%*
	n = 63	n = 51	n = 83	n = 80	(0/78)	(10/68)
"Normal"	0.384	0.364*	0.560*	0.577*	4%*	27%
inc NT	n = 115	n = 92	n = 198	n = 193	(9/211)	(164/45)

118
Prenatal closure of the fetal ductus arteriosus – result of thoughtless pain medication?

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Introduction: Physiological fetal circulation requires patency of the ductus arteriosus. As gestation proceeds, the sensitivity of the ductus to dilating prostaglandins diminishes. The sensitivity to constricting agents like PGE-synthetase inhibitors, present in many analgetics, however, increases. Fetuses affected by an antenatal closure of the ductus arteriosus (DC) may present with different signs of cardiac failure including dilated right ventricle, tricuspid regurgitation and abnormal venous Doppler.
Patients and Methods: We report on three cases with prenatal DC, presenting at 34 + 0 (case 1), 35 + 3 (case 2) and 36 + 6 weeks of gestation (case 3). Case 1 was medicated with metamizol due to implantation of a renal double-J drainage. Five days later, fetal right heart dilatation was noted on routine scan. Fetal echocardiography revealed tricuspid insufficiency (III–IV), a dilated right ventricle with impaired contractility, and a small arterial duct with increased velocities on pulsed Doppler examination. Case 2 was referred because of fetal right heart dominance; echocardiography demonstrated variable aliasing in the isthmic region. One week later, the arterial duct was small with increased Doppler velocities. There was massive tricuspid and pulmonary insufficiency as well as a pathological Doppler of the ductus venosus. Ibuprofen was taken a couple of days before. Case 3 had received prophylactic low-dose-aspirin until 36 weeks of gestation due to lupus erythematodes. Routine fetal scan showed right heart dilatation and a small sized ductus arteriosus. In all cases, immediate delivery was performed. The neonates were in good condition; echocardiography showed no residual ductus arteriosus and different degrees of right heart hypertrophy. The latter disappeared in all infants until the age of 3 months except in case 2.
Discussion: Unexplained fetal right heart decompensation requires detailed echocardiografic evaluation of the ductus arteriosus and a sophisticated medical history with regard to analgetics. In contrast to ibuprofen and high dose aspirin, metamizol and low dose aspirin have not yet been reported as possible agents constricting the fetal arterial duct. In any suspected context, early delivery as in our cases may save the babies life. Any application of non-steroidal anti-inflammatory drugs in pregnancy requires close fetal follow up due to their potentially life threatening effect.

119
Ectopia cordis in prenatal life

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In a series of 7,810 patients referred for fetal echocardiography in 9 year period, 9 fetuses were found to have ectopia cordis (ec). Association with cardiac and extra-cardiac malformation, chromosomal anomalies and pre/postnatal outcome was analysed. There are 7 cases of complete ectopia cordis with all the heart outside of the chest and 2 with the heart partially exposed. Cardiac anomalies were detected in 8 fetuses, in 3 double outlet right ventricle, 2 ventricular septal defect, 1 tetralogy of Fallot, 1 truncus arteriosus, 1 atrioventricular septal defect. The remaining 11 week old fetus, the cardiac anatomy was not adequately visualized with further termination of pregnancy. Exomphalos was detected in 7 cases and gastrochisis (2). Bilateral cystic hygroma and single umbilical artery was found in 1 fetus. Autopsy performed in 5 patients showed midline thoraco-abdominal wall defect with extrophia of liver, spleen and bowels. Also absence of sternum, diaphragm and pericardium. Fetal karyotype was performed in 6 patients, normal in 5 and trisomy 18 (1 case). First trimester diagnosis was accomplished in 3 fetuses.

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Intrauterine death ocurred in 2 fetuses, neonatal in 5 patients, termination of pregnancy (1) ec alive (1). Placement of the heart into the chest and closure of thoracic-abdominal wall was carried out in 2 patients. Both had a muscular vsd. The one with complete ectopia cordis had abnormal ventricular filling leading to postoperative death. Further neonate with partial ec is alive and has a mild secundum asd. In conclusion, it is important to distinguish cases of partial ectopia cordis from those with protusion of all heart outside the chest and significant strutural cardiac malformation. The prognosis will depend on the severity of the heart disease and associated extracardiac anomalies but usually has a poor outcome. A multidisciplinary approach to diagnosis and management of these pregnancies is recommended.

120
Amiodarone: efficacious treatment of refractory fetal tachycardia

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Background: Persistent fetal tachycardia (FT) associated with hydrops fetalis or ventricular dysfunction, is associated with mortality in 5–30% and risk of permanent neurologic sequelae. Several tachycardia mechanisms, most commonly reentrant supraventricular tachycardia (SVT), atrial flutter (Afl), ventricular tachycardia (VT), and junctional ectopic tachycardia (JET) have been described. Treatment response may vary depending on the mechanism of the tachycardia, antiarrhythmic and dose, and the route of administration. Proarrhythmia has been reported, but it is unclear whether more proarrhythmic antiarrhythmic agents confer a greater mortality risk.
Methods: We report results utilizing amiodarone (amio) at 6 Midwest USA Hospitals (n = 26), and University Hospital, Utrecht (n = 1), over a 12 year period in 27 fetuses 19–36 weeks (wks) gestational age (GA). Prior to amio, patients had failed digoxin (n = 24), flecainide (n = 5), sotalol (n = 2), and other agents (n = 6). Amiodarone was administered orally to the mother, 1800–2400 mg/day in 3–4 divided doses for 2–5 days, followed in 24 patients by maintenance of 200–800 mg/day for 1–15 wks. Three were delivered urgently. Digoxin was continued in most circumstances. A second antiarrhythmic agent (AA) (verapamil, flecainide) was utilized after 8–15 days if sinus rhythm (SR) was not restored. Amio was discontinued after 3 wks of FT quiescence.
Results: The table shows efficacy by diagnosis for those treated more than 6 days (n = 24), the mean time to conversion. Mortality or serious neurologic sequelae did not occur. 5 fetuses had transient biochemical hypothyroidism not requiring treatment unless post-natal amio was continued (n = 1). Maternal side effects were rash (n = 1), minor conduction changes (n = 6), and thrombocytopenia (n = 1).
Conclusions: Amiodarone is safe and effective for conversion of fetal SVT, VT (in the absence of QT prolongation), and JET. Sotalol appears more efficacious for Afl.

121
Cardiac disturbances in insulin-dependent diabetes mellitus in child

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The aim of the study was to present the main cardiac anomalies, which occur in type I insulin dependent diabetes mellitus (IDDM), in child.

Methods: The authors followed up for at least 24 months 34 children with IDDM (aged 5–18 years); all patients underwent complete clinical examination, ECG, chest X-ray and echocardiography (echo). The autonomic neuropathy (AN) was proved by 2/5 positive results in Ewing cardiovascular standard reflex tests. The hemoglobin A1c levels estimated the diabetic control.
Results: All patients were normotensive and free of signs of cardiovascular suffering. ECG and chest X-ray were generally normal. Echo showed cardiac abnormalities suggesting diabetic cardiomyopathy (29% of cases), but only in children with AN: thickening of the interventricular septum and of the posterior wall of LV (6 cases) – mild dilatation of LV with a small decrease of shortening and ejection fraction of the LV (5 cases); decreased septal contractility(6 cases); increased isovolumic relaxation time and increased pre-ejection period/LV ejection time ratio (9 cases); diastolic dysfunction of LV in 11 cases. It was found out an important correlation between the cardiovascular disorders (especially diastolic function) and the IDDM evolution (especially the long duration of IDDM, the presence of a poor glycaemic control, microangiopathic complications, a great number of ketoacidosis). The normalization of the glycaemic control improved the diastolic function of the LV in 6 cases. Autonomic neuropathy (57% of cases) was present even in the first 1–2 years of diabetes evolution when the systolic and diastolic functions of the LV were still normal.
Conclusions: The precocity and high incidence of cardiac dysfunctions in children with IDDM and often lack of cardiovascular symptoms, impose a systematically and precocious complete cardiological investigation. It is also necessarry to monitoring for morphological aspects and function of the LV using echo, especially in diabetic children with a poor glycaemic control and autonomic neuropathy.

122
Prognostic value of balloon occlusion test (BOT) in candidates for various types of cavo-pulmonary anastomosis

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Purpose: Prognostic value of BOT for further treatment was estimated in patients (pts) with various types of functionally single ventricle.
Material and Methods: BOT was performed during catheterization in 19 pts and was followed by transcatheter intervention and/or surgery when necessary. The age of pts ranged from 1,4 to 11 (mean 6,25) years. In all of them at least two sources of pulmonary blood supply were present. In 8 pts BOT of the pulmonary artery (PA) (group I) – and in 11 pts BOT of B-T shunt (group II) was performed. During BOT pulmonary blood pressure and systemic saturation were measured.
Results: In 7 pts from group I mean pulmonary arterial pressure (MPAP) decreased after BOT from 20,7 to 14,2 mmHg and all were qualified for second stage of Fontan procedure. In 1 pt after PA occlusion MPAP decreased from 26 to 22 mmHg with simultaneous decrease of saturation and BT shunt was performed. In 10 pts of group II BOT of BT shunt resulted in decrease of MPAP from 23 to 14 mm. Four pts were qualified for bidirectional Glenn and 6 – for TCPC operation. In 1 pt MPAP after BOT remained elevated (decreased from 50 to 36 mmHg) and PA banding was done instead of previously planned Glenn anastomosis. Transcatheter intervention was performed in 15 pts. PA stenosis was

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treated by balloon angioplasty in 4 and stent implantation in 2 pts, BT shunt was closed with coils in 5 pts, azygos vein escape in another 2 and PA with Amplatzer device in 2 pts. Follow up was uneventful in all pts, but one.
Conclusion: In patients with previously palliated complex cardiac lesions temporary balloon occlusion tests may help to predict hemodynamic changes that can occur after planned transcatheter or surgical procedures.

123
Normal values of B type natriuretic peptide in infants, children, and adolescents

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Objective: To examine prospectively normal values of plasma B-type natriuretic peptide from infancy to adolescence using a commercially available rapid assay.
Methods: In 195 healthy infants, children, and adolescents at age 0 day to 17.6 years BNP plasma concentration was measured using the Triage BNP assay (fluorescence immunoassay). Statistical differences between the age groups, pubertal stages, and the two sexes were evaluated using rank score test to unpaired data (Mann-Whitney). Correlation and regression analyses were performed using natural log transformated BNP values to assess association to age, body mass index, creatinine, creatinine clearance (univariate regression analyses), pubertal and menarchal stage (Spearman rank correlation).
Results: During the first week of life, the mean (SD) plasma level of BNP in newborn infants decreased significantly from 231.6 (197.5) to 48.4 (49.1) pg/ml ($p = 0.001$). In all subjects older than 2 weeks BNP plasma concentration was lower than 32.7 pg/ml. There was no significant difference in mean BNP plasma levels measured in boys and girls younger than 10 years (8.3 (6.9) v 8.5 (7.5) pg/ml). In contrast, plasma concentration of BNP in girls aged 10 years or older was significantly higher than in boys of the same age group (12.1 (9.6) v 5.1 (3.5) pg/ml, $p < 0.001$). BNP plasma levels were significantly higher in pubertal than in prepubertal girls (14.4 (9.7) v 7.1 (6.6) pg/ml, $p < 0.001$) and were significantly correlated to the Tanner stage ($r = 0.41$, $p = 0.001$).
Conclusions: BNP plasma concentrations in newborns are relatively high, vary greatly, and decrease rapidly during the first week of life. In children older than 2 weeks, mean plasma level of BNP is lower than in adults. There is a sex-related difference in the second decade of life with higher BNP levels in girls. BNP levels in girls are related to pubertal stage.

124
Cardiac involvement and myocardial microcirculation disturbance in children with systemic sclerosis

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Background: Myocardial microcirculation disturbance and decreased coronary reserve contribute to cardiac dysfunction in patients with systemic sclerosis (SSc). Patients with skeletal muscle disease are at increased risk of cardiac death.
Objective: To assess cardiac function and perfusion in children with SSc and myositis.
Patients and Methods: 7 girls (mean age 9.2 ± 3.2 years) who met the American College of Rheumatology criteria for SSc and had features of polymyositis, as defined by the presence of proximal muscle weakness and elevated serum creatine phosphokinase or aldolase level, underwent cardiac evaluation with echocardiography, Thallium-201 myocardial perfusion imaging and selective coronarography in 2. All had multivisceral extracardiac involvement including lung restrictive syndrome, oesophagal and intestinal dysfunction.
Results: All patients had cardiac involvement with dilated cardiomyopathy in 5, patchy myocardial perfusion defects in 7, and severe ventricular arrhythmias in 1. Selective coronarography was normal when performed. Two patients died from endstage cardiac failure, one under mechanical circulatory support device. One underwent successful heart transplantation. Left ventricular function improved in one after allogeneic bone marrow transplantation. Myocardial perfusion improved in one after carvedilol and nifedipine therapy. Histological studies showed no inflammation nor microangiopathy but segmental fibrosis consistent with ischemically mediated injury.
Conclusion: Severe cardiac involvement is frequent in children with SSc. These patients should be screened for myocardial perfusion defects. A potentially reversible abnormality of coronary vasomotion could be responsible for ischemic dilated cardiomyopathy in this condition. Coronary microcirculation could be the target for therapeutic interventions preventing cardiac dysfunction in these children.

125
Myocardial performance, baroreceptor reflexes and coronary flows evaluation using the tilt table in preterm and term neonates

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Tilt table test is the preferred method of testing the baroreceptor response in children. The effect of short-term body tilting on myocardial performance, baroreceptor reflexes and coronary flows were studied in preterm and term neonates. Fifteen preterm infants with gestational age 32 ± 3 weeks, birth weight 1750 ± 245 g, and 15 term infants with 39 ± 1 weeks, 3450 ± 200 g were studied in supine, 30, and 60 head-up positions. No significant difference in heart rate, blood pressure, left ventricular cavity dimensions, ejection fractions, stroke volume, cardiac output, end systolic wall stress (ESWS) and velocity of circumferential fiber shortening (VcFs) were found between baseline and head-up tilt positions in preterm and term neonates. In coronary flow measurements, there is no significant difference in LAD flow volume and LAD flow volume/LV mass in term neonates in all head up positions. But in premature neonates a significant decrease in LAD flow volume/LV mass in 60 head up position was observed. LAD flow volume(ml/min)in supine, 30 and 60; 2,7 ± 0,8, 2,3 ± 0,4 and 1,9 ± 0,3 in prematures and 3 ± 1,3, 3,1 ± 1,2 and 3,2 ± 1,1 in matures were measured respectively. LAD flow volume/LV mass (ml/g) in supine, 30 and 60; 0,57 ± 0,16, 0,46 ± 0,14 and 0,38 ± 0,17 in prematures and 0,58 ± 0,17, 0,59 ± 0,20 and 0,61 ± 0,19 in matures were measured respectively (LAD: Left anterior descending coronary artery) As a conclusion; we did not find a significant change in contractility parameters with head-up tilt, which demonstrated the adaptability of the neonatal myocardium to changes in the loading conditions. But in premature neonates a significant decrease in LAD flow volume/LV mass in 60 head up position were observed. These findings can be important for evaluating the performance of myocardial functions in preterm and term neonates.

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126
Conversion from cyclosporine A to tacrolimus after pediatric heart and heart lung transplantation

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Side effects of immunosuppressive therapy gain more and more importance. Changing from cyclosporine (CsA) to tacrolimus as the basic immunosuppressive drug in heart transplanted children represents a great advance in reducing the side effects attributable to CsA. In a prospective study we investigated the effects of conversion from CsA to tacrolimus due to different side effects in pediatric recipients of cardiac allografts. Primary indications for the switch to tacrolimus were gingival hyperplasie in 4 pts, renal insufficiency in 2 pts, atopic eczemas in 2 pts and severe hypertrichosis in 3 pts. 10 patients (pts) with stable graft function and 1 pt with a rejection episode (planned for conversion due to renal insuffiency and gingival hyperplasie but representing with rejection) were assigned to conversion to tacrolimus. Demographic patient data: n = 11, female 5, male 6, HTx 10 pts, HLTx 1 pt, diagnosis: DCM n = 7, Kawasaki Disease n = 1, idiopathic pulmonary hypertension n = 1, congenital heart disease n = 2, weight 47.5 ± 20.9 kg, age 12.6 ± 5.7 years (yrs), follow-up time 4.8 ± 3.0 yrs. Renal insufficiency was evaluated in values of serum creatinine and creatinine clearance, atopic eczema was assessed by calculating the Scorad score, hypertrichosis by calculating the Ferriman-Galway-Index. Tacrolimus was introduced directly (0.15 mg/kg starting dose) with doses adjusted to a range of 8–10 ng/ml. Concomittant immunosuppressive therapy, consisting of mycophenolate mofetil and low dose steroids in the heart-lung-transplanted patient was maintained after the switch. In pts who were switched for the respective symptom the following improvements were achieved 3 months after the switch: gingival hyperplasia had strongly resolved in all pts 3 months after the switch. Renal function increased with mean serum creatinine of 1,5 mg/dl and 1,2 mg/dl 3 months after the switch. Atopic eczema disappeared in 1 pt and remained unchanged in the other one. Hypertrichosis had strongly resolved in all 3 pts. The Ferriman-Galway-index for hypertrichosis had decreased by 62%. Conversion from cyclosporine to tacrolimus in pediatric heart transplanted pts is effective. CsA related side effects improved within a few months, resulting in a better quality of life and an improved cardiovascular risk profile.

127
Double orifice mitral valve (DOMV) – clinical presentation, natural history and outcome in children and adolescence

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Background: Double orifice mitral valve (DOMV) is considered to be a rare congenital cardiac anomaly, but is an increasingly recognized incidental finding during echocardiography, heart surgery, or autopsy. Previous case reports and one pathologic series reveal DOMV to be either isolated or associated with AV Septal defect, and left sided obstructive lesions. The purpose of this study was to describe the clinical presentation, mode of diagnosis, associated anomalies, natural history and outcome in a large group of patients (pts) with DOMV, in order to clarify the nature of this anomaly.
Methods: We retrospectively reviewed the records of 46 children with DOMV aged 2days–16 years (median: 2.4 years) who were diagnosed and treated in our medical centers over a 22 year period (1980–2002). Clinical and surgical reports, echocardiographic examinations, cardiac catheterization data, and follow up outpatient visits were evaluated.
Results: There were 13(28%) male and 33(72%) female pts identified. Symptoms were described at presentation in 12 pts. Partial AV septal defect was the most common associated lesion (28%), followed by complete AV septal defect (23%), and aortic coarctation (19%). Mild to moderate mitral insufficiency was detected by color Doppler echocardiography in 43% of pts. Mitral inflow obstruction (peak and mean Doppler gradient 3.49 ± 5.01 and 2.07 ± 3.03 mmHg respectively; mean ± SD) was observed in 15% of pts. Seventeen pts underwent surgical repair of their associated anomalies with correction of the DOMV deemed necessary in only 6/17 pts.
Conclusion: DOMV is an uncommon congenital heart anomaly frequently associated with AV septal defect and/or left ventricular obstructive lesions, but it can occur in isolation. Symptoms are related to the degree of associated mitral insufficiency or stenosis. Surgical attempts to reconstruct the valve are usually not indicated, but might be of benefit when the DOMV is approached in the context of a wider operation such as for an AV septal defect.

128
Preoperative immunological and serological differences between children with and without post-surgical capillary leak syndrome

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Post operative capillary leak syndrome (CLS) and multi organ failure (MOF) can occur in children undergoing major cardiac surgery with cardiopulmonary bypass (CPB). Patients with postoperative complications differ in their preoperative complement concentration. Furthermore, in adults the severity of heart impairment correlates with serum cytokine levels. Own results in a small group of patients indicate that based on preoperative data a risk assessment of CLS could be performed. The aim of the present study was to investigate preoperative immunological differences in patients for their prognostic significance of CLS. 24 pediatric patients (3–16 yr.) undergoing cardiac surgery with CPB were investigated. Data out of routine laboratory, differential blood picture, serology, complement system and antigen expression was estimated. Only few differences were significantly increased (histamine, s ICAM) and only one parameter (IL-8) was decreased in CLS patients. On the basis of 24 h pre-operatve differences of the immunological status it is possible to estimate indicators for risk of CLS after cardiac surgery. Risk patients have a slightly activated immune sytem with increased complement consumption and activation and cytokine levels as well as increased activation of monocytes and neutrophils. An early risk assessment can be performed by a combination of parameters and could eventually lead to an improved therapy of risk patients.

129
Slow kinetics of oxygen uptake during exercise in patients with a Fontan-type circulation

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Peak oxygen consumption and anaerobic threshold are both decreased in patients with a Fontan-type circulation. The aim of this is to evaluate oxygen uptake kinetics at the onset and at the end

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of a steady-state low-level exercise. The delay in cardiorespiratory response was evaluated by calculating the oxygen deficit at the onset of exercise and the recovery half-time at the end. Twelve patients with Fontan circulation (aged 11.4 ± SD 5.1 year; 5.2 ± 1.9 year after surgery) and 26 normal controls of comparable age (11.3 ± 2.2 year) were submitted to a constant-load exercise test of six minutes on a treadmill (speed 5 km/h, inclination 4%). Gas exchange was measured using a breath-by-breath technique. The normalized oxygen deficit was calculated by subtracting the oxygen uptake (VO_2) values measured at the onset of exercise from the steady-state VO_2 obtained at the end of exercise. These differences were cumulated and expressed as a percentage of the cumulated oxygen cost for the 6 min exercise test. The half-time recovery time was defined as the time to reach 50% of the end exercise VO_2 value. The normalized oxygen deficit was significantly higher in Fontan-patients compared to the control group (10.2 ± 4.6% vs. 6.1 ± 1.3%; $p < 0.001$). Also the recovery half-time was significantly higher in the patient group compared to the control group (74.2 ± 25.6 s vs. 51.2 ± 10.8 s; $p < 0.05$). A blunted heart rate response was present in the patients during the first two minutes of exercise, indicating that a slowed cardiac output response could explain the decreased oxygen kinetics in Fontan-patients. It is concluded that the slower adaptation of the oxygen transport system the onset of exercise may explain the difficulty perceived by patients with Fontan-circulation to adapt to the onset of exercise.

POSTER PRESENTATIONS

P130

CONCOR: National registry and DNA-bank of adults with congenital heart disease in the Netherlands

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Introduction: Survival of patients with congenital heart disease has dramatically improved after surgical repair became available. However, long term complications are frequent and may require indefinite follow up. Exact data on longterm outcome are still lacking. Moreover, little is known about the underlying genetic cause of congenital heart disease.
Purpose: A national registry and DNA-bank of adults with congenital heart disease has been instigated in the Netherlands to investigate long term outcome in these patients and to allow investigation of the molecular basis of congenital heart defects.
Methods: All eight academic medical centers in the Netherlands are participating in the CONCOR-project (CONgenital CORvitia). After informed consent has been obtained clinical data of patients with congenital heart disease are collected using a web-based application. For each patient, all clinical events (diagnoses and interventions) are registered using the EPCC (European Pediatric Cardiac Code) coding scheme. Data are transferred via the Internet to the CONCOR server using a secure connection. Data are collected in accordance with the Netherlands Privacy Protection Laws using several Privacy Enhancing Technologies. Blood samples of participating patients are collected, and DNA is isolated and stored. Investigators with a request for data and DNA have to follow a stringent procedure before data and DNA are delivered to the investigator. At a later stage participation of the pediatric cardiologists and local hospitals will be pursued. The CONCOR-project is supported by a grant from the ICIN and the Netherlands Heart Foundation.
Results: Ten months after the start of the project 1700 adult patients on an estimated total of 25000 adults in the Netherlands, are included. More then 99% of the patients asked to participate gave their consent for both registration and storage of their DNA-samples.
Conclusions: Registration and collection of DNA of patients with congenital heart disease is feasible and important to facilitate research on long term outcome and to allow investigation of the molecular basis of congenital heart defects.

P131

Reduced expression of sarcoplasmic reticular Ca^{2+}-ATPase mRNA in volume overloaded atrial myocardium from children with congenital heart defects

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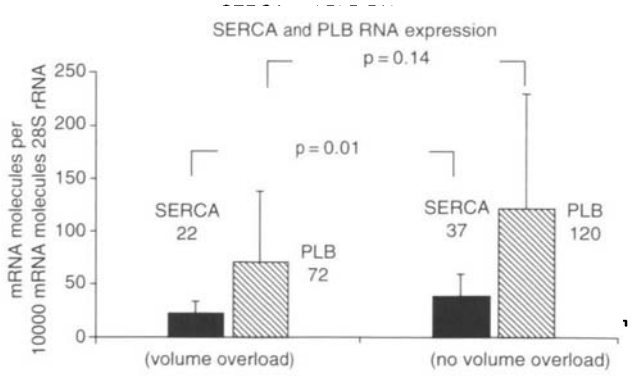
Background: The cardiac isoform of sarcoplasmic reticulum calcium-ATPase (SERCA2a) and its inhibitor phospholamban (PLB) are important calcium regulatory proteins. Differences in expression have been related to various states of cardiac overload, species, age and different cardiac regions. To date studies were performed mainly in animal models and adult humans. Therefore we studied atrial myocardium of children with congenital heart defects with volume overloaded compared to normal right atria.
Methods: Atrial myocardium was excised during corrective surgery. SERCA2a and PLB mRNA expression was measured by quantitative real-time reverse transcription polymerase chain reaction. Western blot analysis was used to study the protein expression. The study protocol has been approved by the ethics committee.
Results: Overall 30 patients were studied. 18 patients had a volume overloaded (VO) right atrium. 12 patients had congenital heart defects without volume overload (NO) of the right atrium. The age distribution (0.8 to 165 months, mean 44 months) was statistically not different between the groups. The amount of transcripts was expressed as mRNA molecules per 10000 28S rRNA (internal control) molecules. SERCA2a mRNA expression was lower in the VO group (22 ± 11) compared to the NO group (38 ± 23, $p = 0.01$). PLB mRNA expression showed no statistical difference between VO group (72 ± 53) and NO group (120 ± 123, $p = 0.14$). The ratio between SERCA2a and PLB in the VO group was 0.36 ± 0.14, in the group without volume overload 0.42 ± 0.18 ($p = 0.28$). The protein expression of SERCA2a and PLB, respectively, was investigated in 6 patients from each group and showed no statistical difference.
Conclusion: Expression of SERCA2a mRNA was significantly reduced in volume overloaded pediatric atrial myocardium, which is in line with results from studies with various atrial pathologies like atrial fibrillation. The lack of statistically significant difference in protein expression may be due to the low number of patients investigated on. Changes of calcium regulating components could explain functional consequences like impaired contractility and arrhythmias.



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P132
Specificity protein (SP) factors are required for basal and cAMP mediated cardiac troponin T (cTNT) gene expression in embryonic cardiac myocytes
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Developmental regulation the cTNT gene promoter is used as a model of cardiac-specific gene expression and myocardial differentiation. In addition to some well characterized binding sites (MCAT, AT-rich, GATA and MEF-3 elements) for trans-acting factors (TEF-1, MEF-2, GATA) the cTNT promoter contains a GC box (at −50 to −55) similar to an Sp-1 binding site. In this study we examine the contribution of the Sp factors and their putative GC-box binding site to regulation of the cTNT gene. Western blot analysis of the cardiac muscle nuclear extracts from embryonic day 12 chick embryos shows that Sp1, Sp2, Sp3, and Sp4 are all expressed in embryonic cardiomyocytes. DNA binding studies using GC-box probe and cardiac nuclear extracts also show the presence of 2 mobility-shift complexes that bind in a sequence-specific fashion when wild-type and mutant competitor are added to the binding reaction. Furthermore, supershift complexes are formed by antibodies to Sp1, Sp2, Sp3 and Sp4. Transient transfections of primary embryonic cardiomyocytes requires a full length promoter (−268 cTNT) for cTNT gene activity. The upstream "cardiac element" consisting of AT-rich, GATA and MEF-3 elements (at −201 to −268) is necessary for activity in cardiomyocytes. Artificial constructs containing the AT- rich element upstream of the minimal promoter (−129 cTNT) rencostitues 70% of the gene activity in vitro. Transient transfections of primary embryonic cardiomyocytes with full length promoter (−268 cTNT) containing a mutated GC-box results in a 10-fold reduction in gene activity. Likewise, cAMP mediated upregulation of cTNT gene expression in embryonic cardiomyocytes is abolished by mutation of the GC-box binding site. Sp factors are essential for basal and cAMP-mediated activation of the cTNT gene.

P133
Usefulness of Tc-99m HMPAO labeled WBC heart scan to predict the impaired ventricular function and coronary aneurysms in children with Kawasaki disease
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Forty-nine children with Kawasaki disease (KD) were included in the study. Based on the severity of carditis evaluated by Tc-99m HMPAO labeled WBC heart scan (WBC scan) findings, the children were separated into 2 groups. Group A of 24 children had significant carditis and group B of 25 children had not significant carditis. The left and right ventricular ejection fractions (LVEF and RVEF) were evaluated by equilibrium multigated blood pooling ventriculography (EMBPV). The diameters of left and right coronary arteries (LCA and RCA) were measured by two-dimensional echocardiograms (2D-Echo). The results showed that group A patients had lower LVEF and RVEF but larger diameters of LCA and RCA than those of group B patients. Our findings suggest that significant carditis found on WBC scan can predict impaired ventricular function and coronary aneurysm.

P134 (see Abstract 113)
Pharmacological treatment of young children with permanent junctional reciprocating tachycardia: follow-up data of tachycardia and cardiac function
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P135
Postnatal management of critical aortic stenosis in the fetus: an alternative to in utero treatment
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Backgrounds: Critical aortic stenosis (AS) prenatally detected is associated with significant mortality and termination is chosen in many cases with early diagnosis. To prevent hypoplastic left heart at term, prenatal aortic balloon valvuloplasty (ABV) has been advocated, but only one survivor is reported in the world experience.
Methods: Among 208 cases of structural heart disease referred to our fetal unit in a five years period, 6 (2.8%) had critical AS. The mean gestational age at referral was 27 wks (range 20 to 35). All cases had aortic valve (AV) forward flow, reversed flow at foramen ovale and distal aortic arch level, and LV dysfunction with endocardial fibroelastosis. Mitral regurgitation (MR) was mild-to-moderate in four cases and severe in two hydropic fetuses. Termination was chosen in 2 out of 3 cases with early presentation. To promote early decompression of LV with acceptable fetal maturity, delivery was planned at 34–35 wks in three patients. One patient was delivered at 29 wks because of maternal complications. Cesarean section was elected in all cases. All 4 newborns, mean weight 2.3 Kg (range 1.6 to 3.0), received mechanical ventilation, PGE1 and inotropic agents infusion, and all undewent ABV through a right carotid approach within 5 hrs of life.
Results: There was one early death and three survivors at a mean follow-up of 30 mths (range 7 to 70). LV volume, LV systolic function and MR improved in all survivors. Major complications included: significant aortic regurgitation in two cases, right diaphragmatic palsy and hydrocephalus in one case each. The mean ventilation time has been 33 days. All survivors had repeated AVB and/or surgical valvulotomy, one patient had a PDA ligation. All patients are well with no medication, two have significant AV gradient.
Conclusions: Prudent delivery anticipation and immediate neonatal ABV can be a promising alternative to in utero treatment of critical AS prenatally detected. Improvement of LV systolic function and of MR can occur, even in the hydropic fetuses. Prolonged ventilation time, high incidence of restenosis or insufficiency of the AV is anticipated.

P136 (see Abstract 96)
Outcome of transcatheter closure of muscular ventricular septal defects using the Amplatzer Ventricular Septal Defect Occluder
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P137

Use of 3F catheters for diagnostic and interventional procedures in newborns and small infants

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Miniaturisation of diagnostic and interventional tools is of main interest for cardiac catheterization of newborns and small infants, due to the limited vascular access of these patients. Since January 2001 to November 2002 we performed 10 diagnostic and 5 interventional procedures in 14 infants, utilising custom-made 3F sheaths and catheters. Median age and weight of the patients were 7 days (range 1–180) and 2.8 Kg (range 2–4), respectively. Diagnostic catheterization gave the required information in all cases. Angiography was performed following the results of in vitro pressure/volume tests. Blood sampling was possible by using gentle aspiration. Pressure measurement, although showing some dumping, allowed the evaluation of peak to peak gradient through right and left ventricular outflow. It was always possible to perform an interventional procedure, when required. No procedural complications occurred. The price of a diagnostic or interventional procedure performed with 3F sheath and catheters exceeded of 23 Euros that of a procedure performed with material measuring 4F or more. Patients underwent echographic examination of femoral vessels at a median follow-up of 6.3 months (2–10). All vessels of the examined patients were patent, without stenosis nor collateral circulation. In conclusion, preliminary results utilising 3F sheaths and catheters are encouraging, in terms of feasibility of diagnostic and interventional procedures, absence of procedural complications and preservation of vascular access. Financial implications are not of major concern.

P138

Psychosocial service provision within British Paediatric Cardiology Units

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The Bristol Royal Infirmary Inquiry and recent Department of Health Paediatric & Congenital Cardiac Services Review have highlighted the importance of appropriate psychosocial support services within Paediatric Cardiology. The recommendation that "National standards should be developed as a matter of priority, for all aspects of care and treatment of children with congenital heart disease" (Kennedy Report 2001) clearly encompass psychosocial support. To ensure psychosocial service development within the Yorkshire Heart Centre, Leeds, was based on current evidence and best practice, a survey to investigate the provision and configuration of psychosocial support available within the 19 Paediatric Cardiology centres in the UK and Ireland was carried out. A questionnaire was sent to the 19 centres which aimed to ascertain the type of psychosocial services, models of service delivery, gaps in service, obstacles to service delivery and development, and the type of psychosocial problems encountered within units A 100% response rate was obtained and basic statistical analysis of the quantitative data and descriptive analysis of the qualitative data was conducted. The survey revealed a significant variation between centres in numbers of staff, type of professionals delivering the service and the nature of their employment. The survey also highlighted the array of psychosocial problems encountered in the centres and consequently the clear psychosocial needs of children with CHD and their families. The survey revealed that current psychosocial services are not meeting the needs of children and their families. Only 2 of the centres felt they were delivering a service that met psychosocial need. A number of barriers to the development of psychosocial services within Paediatric Cardiology were identified, including funding difficulties and apathy towards the significance of psychosocial support within units. The development of appropriate psychosocial services, in line with report recommendations and patient need, will clearly have resource implications.

P139

Total autologous Ross procedure

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Ross operation is the procedure of choice for certain case groups with young age, active life style, endocarditis, and child expecting females. It is a technically demanding procedure with slightly higher procedural mortality, morbidity with additional risks as autograft failure and late conduit degeneration and stenosis but described advantages might overweigh the risks. Using autologous fresh pericardium for right ventricle outflow tract (RVOT) reconstruction may decrease the risk of future need for conduit replacement. With this idea we constructed fresh autologous pericardial monocusp and tube instead of biological RVOT conduits. Between 1995 and 2002, 23 patients underwent Ross procedure with overall mortality of 4.4% (1 patient). Due to unavailability of homografts porcine or bovine biological valved conduits were used for RVOT reconstruction (Medtronic Freestyle, Cryolife Ross, Medtronic Contegra). On midterm follow up no significant autograft dysfunction was detected but >25 mmHg gradient increase was observed in most of the patients. Although none of them needed reoperation for RVOT replacement we began to seek alternative methods in place of xenograft valved conduits. In the last 3 patients we used home-made RVOT conduits. One patient received bovine pericardial tube and Gore-Tex pericardial membrane monocusp and 2 patients received fresh autologous pericardial monocusp and tube conduit for RVOT reconstruction. 8 year old young female patient had active endocarditis of the native aortic valve with root abscess and the other 14 year old female patient had rheumatic aortic valve disease. Both patients recovered from the complex procedure smoothly and on early follow up no significant problems were detected on autograft or RVOT. We believe that if pulmonic homograft is not available, total reconstruction of RVOT with autologous fresh pericaridum may offer the best early and longterm results in the current era of Ross procedure.

P140

Extracardiac and lateral tunnel Fontan conversions for late problems after Fontan procedures

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Fontan procedure is an ever evolving orthoterminal surgical procedure. It offers a relatively normal life for most of the patients at mid and longterm. But some patients may have reduced exercise tolerance due to dilated atrial structures, arrhythmias and pulmonary arteriovenous fistula formation. Therefore in centers busy with the care of this difficult group of patients are increasingly dealing with revisions of the previous Fontan procedures. On long-term follow-up of our 114 patiens who underwent various modifications of Fontan procedure since 1987. Three needed reintervention because of decreasing functional status, atrial

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arrhythmia and cyanosis. First and the second patient were at ages 9, 10 and 3, 5 years after atriopulmonary Fontan procedure consecutively. Both had dilated right atrium and atrial arrhythmia (intra atrial reentrant tachycardia and atrial fibrillation). On lateral tunel conversion operation pulmonary artery plasty and closure of residuel Blalock Taussig shunt were needed in addition to reseptation and reduction plasty of atria. Postoperative course was uneventful and both patients are arrhythmia and symptom free on early follow-up. Third patient had bilateral bidirectional cavopulmonary anastomosis and had increasing cyanosis due to pulmonary arterio venous fistula formation. Connecting hepatic veins to pulmonary artery by a pericardial extracardiac tube was planned with the expectation that hepatic factor may reach the pulmonary system in order to regress the AV fistula formation and cyanosis. After the completion of the Fontan circulation, circulation cyanosis regressed gradually over 6 months as expected in this patient (O_2 saturation increased from 55% on room air to 85%). During the close follow-up of patients revision of previous Fontan modifications is sometimes inevitable. With proper work-up appropriate technique might be applied as lateral or extracardiac Fontan modifications with good results in these patients who will have a disappointing prognosis otherwise.

P141

Glutathione, Glutathione peroxidase and Glutathione reductase responses to hypoxia-reoxygenation in adult isolated rat hearts following neonatal hypoxia

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We have asked whether hypoxia neonatally (FiO_2 = 0.12, days 1–10 of life) has longlasting effects on the myocardial ability to deal with oxidative stress during acute hypoxia-reoxygenation. Left ventricles from 20 adult Sprague Dawley rats (439 + 12 g) were analyzed for reduced and oxidized glutathione (GSH and GSSG) by an automated glutathione reductase recycling method while glutathione peroxidase (GPx) and glutathione reductase (GR) activities were determined from the decrease in absorbance of NADPH. Ten rats experienced hypoxia neonatally (Neonatal Hypoxia) while 10 others did not (Control). The hearts were isolated and perfused in the constant pressure Langendorf mode and the left ventricles quickly frozen either following perfusion with i) oxygenated perfusate (25 minutes) (Baseline, n = 5 each group), or ii) oxygenated perfusate (25 minutes) – hypoxic perfusate (10 minutes) – oxygenated perfusate (5 minutes) (Reoxygenation, n = 5 each group). At Baseline there was no difference between Control and Neonatal Hypoxia in GSH (1.45 + 0.06(SEM) vs. 1.48 + 0.03 μmol/g), or GSSG (0.12 + 0.02 vs. 0.16 + 0.02 μmol/g). However, GPx was significantly greater at Baseline in Neonatal Hypoxia compared with Controls (0.90 + 0.02 vs. 0.78 + 0.03 U/mg protein P = 0.009) while GR tended to be greater (0.023 + 0.001 vs. 0.020 + 0.001 U/mg protein, P = 0.09). On Reoxygenation GSH, and GSSG decreased in Controls (1.45 + 0.06 vs. 1.07 + 0.06 μmol/g, P = 0.002; 0.12 + 0.02 vs. 0.07 + 0.004 μmol/g, P = 0.02). In contrast in Neonatal Hypoxia GSH did not change on Reoxygenation (1.48 + 0.03 vs. 1.39 + 0.07 μmol/g) although as in Controls GSSG decreased (0.16 + 0.02 vs. 0.10 + 0.01 μmol/g, P = 0.01). GR did not change on Reoxygenation in either group (Control 0.020 + 0.001 vs. 0.189 + 0.001 U/mg protein; Neonatal Hypoxia (0.023 + 0.001 vs. 0.022 + 0.001 U/mg protein). GPx increased on Reoxygenation in Controls (0.78 + 0.03 vs. 0.94 + 0.07 U/mg protein, P = 0.05) but did not change in Neonatal Hypoxia (0.90 + 0.02 vs. 0.91 + 0.02 U/mg protein). Our results suggest a greater activity/reserve of the glutathione redox system following neonatal hypoxia. This may provide increased protection of the myocardium from hypoxic damage. However, the increased oxidative reserve may also decrease coronary vasodilation with hypoxia-reoxygenation(1) and provide an explanation for our previous finding of impaired coronary vasodilation and delayed myocardial recovery following an acute hypoxic stress after neonatal hypoxia(2).

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2 R

P142

Symptomatic neonates with Ebstein's anomaly

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Background: Ebstein's anomaly is associated with a high mortality in symptomatic neonates. We report on a patient who was treated with closure of the tricuspid and pulmonary valve and placement of an AP-shunt (Starnes operation) and have a retrospective view on the newborns treated in our department.
Patients and Methods: Immediately after birth the term neonate presented with severe heart failure and desaturation. Mechanical ventilation and prostaglandin infusion were started. Echocardiography showed severe Ebstein's anomaly with massive tricuspid and pulmonary valve regurgitation, a PDA and right-to-left shunt at atrial level. For six weeks increased pulmonary bloodflow because of decreasing resistance was expected. However, pulmonary bloodflow continued to be scarce. Then a modified Starnes operation was performed. At 7 months the infant underwent a bidirectional Glenn procedure and is now scheduled for TCPC. Since 1992 we observed 15 symptomatic newborns with Ebstein's anomaly. Eleven needed prostaglandin, nine of whom were operated up to now; Starnes operation (first 6 weeks of life) n = 3 (dead 2, alive 1); corrective surgery (age 10 to 65 months) n = 6 (all alive).
Conclusion: Symptomatic newborns with Ebstein's anomaly benefit from declining pulmonary vascular resistance, which results in augmented pulmonary flow. Hence surgical intervention is rarely indicated in neonates with this anomaly and an expectant approach is justified. If the combination of severe tricuspid regurgitation and poor right ventricular function is present, the Starnes procedure will lead to a better outcome.

P143 (see Abstract 108)

Blood cardioplegia protects myocardial metabolism and function better than crystalloid cardioplegia in pediatric heart surgery

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P144

Percutaneous balloon pericardiotomy for the treatment of nonmalignant pericardial effusions in children: futher experience and long-term outcome

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Background: There is very limited preliminary experience with the use of percutaneous balloon pericardiotomy (PBP) for the

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treatment of children with nonmalignant pericardial effusions (PE). In this study, we report further experience and long-term outcome in 15 patients with large nonmalignant PE who underwent PBP.
Methods: Fifteen patients, aged 5 to 16 years, with nonmalignant PE underwent PBP at our institutions. The procedure was performed in the cardiac catheterization laboratory under fluoroscopic and echocardiographic guidance using standard subxiphoid approach. Dilations were performed using standard balloon catheters (18–25 mm wide, 3 cm long) that were advanced into the pericardial space over a stiff 0.035 inch guidewire.
Results: The mean volume of the pericardial fluid removed at the time of PBP was 430 ± 120 (range, 320–600 ml).Three patients, with rapid reaccumulation of PE after initial dilation was treated successfully with a repeat dilation using a larger balloon. The procedure was successful in 12/15 patients (80%). In three patients there was recurrence of PE 1 to 3 months after the procedure. In 2/3 patients with recurrent PE the balloon had ruptured during dilation and in one of them a part of the balloon remained entrapped within the pericardial space causing no symptoms. No recurrence of PE was observed in the remaining patients during a median follow-up of 33 months (range, 6–70 months).
Conclusions: PBP is an effective and safe procedure for creating a adequate pericardial window in children with nonmalignant PE. The pericardium in cases with nonmalignant PE appears to be more resistant to dilation, compared to those with malignant effusions, and this might have contributed to the relatively high rate of recurrences of PE in our study.

P145
Are young females with type 1 diabetes at risk for developing a diabetic cardiomyopathy?

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Background: Several studies in adults have established diabetes mellitus as a strong risk factor for cardiovascular morbidity and mortality, especially in women. This study was designed to assess whether children and adolescents with type 1 diabetes have early echocardiographic signs of systolic and or diastolic dysfunction and whether tissue Doppler provides additional information.
Methods and Results: Eighty unselected children and adolescents with stable type 1 diabetes and fifty two age and sex matched controls underwent a complete echocardiography, including tissue Doppler measurements of the septal mitral annulus. Diabetic children had significant higher body-mass index standard deviation scores (BMI-SDS) compared to controls (0.57 vs −0.28, $p < 0.0005$). Female diabetics showed significantly larger left ventricular wall dimensions ($p < 0.05$) and signs of significant diastolic filling abnormalities ($p < 0.05$) on conventional and tissue Doppler echocardiography (increased mitral and tricuspid peak A velocity with decreased E/A ratio, higher E/E′ ratio, longer isovolumic relaxation time-IVRT), suggesting delayed myocardial relaxation. Male diabetics did only differ significantly from their controls for IVRT ($p < 0.005$). Correlation analysis showed an important influence of age and BMI-SDS on these parameters in the control group, less present in the diabetic group; only a weak correlation was found for diabetes duration and glycosylated hemoglobin levels.
Conclusion: Young diabetics have already significant changes in left ventricular dimensions and myocardial relaxation, the girls clearly being more affected and at risk. Tissue Doppler proved to have additional value in the evaluation of ventricular filling in this population.

P146
Pulmonary venous flow dynamics and its association with vessel cross-sectional diameter in the fetus

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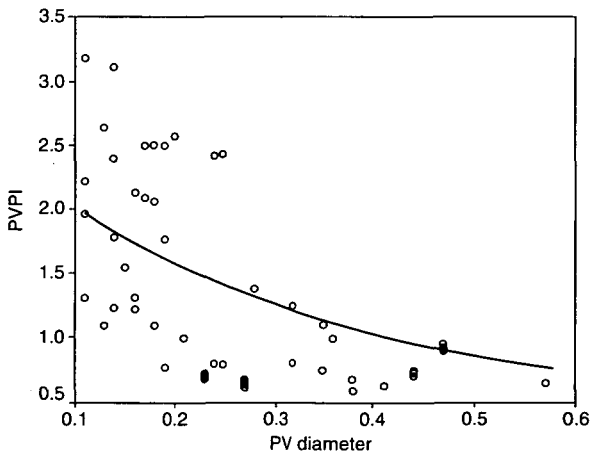
Background: Analysis of the pulmonary vein waveformas, especially its pulsatility index, has being used along with other parameters in the assessment of fetal diastolic function. The usual position of the Doppler sample volume is in the distal portion of the pulmonary vein, close to its opening to the left atrial chamber, where the vessel diameter is maximal. It seemed logical to suppose that if the Doppler study was performed more proximally, in a region where the pulmonary vein size was smaller, the results should be different, expressing an increased impedance to the forward flow where the vessel is narrower.
Objective: This study was carried out to test the association of the pulmonary vein pulsatility index (PVPI) with the vessel diameter in two different sites along the pulmonary vein tree.
Methods: Twenty-three normal fetuses, with a mean gestational age of 28.6 to ±5.3 weeks (20–36 weeks) were studied by cross-sectional (2-D) and Doppler echocardiography. Pulmonary venous flow was assessed in the upper right vein, in two different sites: adjacent to the opening to the left atrium ("distal" position) and in the middle of the vein ("proximal" position). The vessel diameter was measured in the two levels by 2-D echo enhanced with power Doppler and the PVPI was obtained by the pulsed Doppler ratio (systolic peak – presystolic peak)/mean velocity. Statistical analysis utilized t-test and exponential correlation studies, with a limit confidence of 99%.
Results: Mean "distal" internal diameter was 0.33 ± 0.10 cm (0.11–0.57 cm) and mean "proximal"diameter was 0.16 ± 0.08 cm (0.11–0.25 cm) ($p < 0.0001$). Mean "distal" PVPI was 0.84 ± 0.21 (0.59–1.38) and mean "proximal" PVPI was 2.09 ± 0.59 (1.23–3.11) ($p < 0.0001$). Exponential inverse correlation between pulmonary vein diameter and pulsatility index was highly significant ($p < 0.0001$), with a determination coefficient of 0.439.
Conclusion: This study demonstrates that, in the normal fetus, the pulmonary venous flow pulsatility decreases along the way from the lung to the heart, and that this parameter is inversely correlated to the cross-sectional diameter of the pulmonary vein, which increases from the proximal to the distal portion of the vessel.



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P147
Validation of 3d echocardiography to evaluate right ventricular volume and function in pediatric patients
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The right ventricle (RV) is the main pumping chamber in many congenital heart malformations. Unfortunately, its irregular crescent shape doesn't allow the use of any geometrical assumption to evaluate echocardiographically its volumes and function. The recent introduction of 3-D echocardiography allows to calculate volumes of the cardiac chambers without the use of geometrical formulas. Aim of our study is to validate a 3-D echo reconstruction of the RV volume in pediatrc age. We have measured by 3-D echo RVED volume in 18 patients with ASD II type, who were anesthetized and intubated, in the operating room, before surgical correction. We utilized a HP Sonos 5500 with a standard transthoracic 4 MHz rotating probe; the images were 3-D reconstructed by means of the disks summation method. The 3-D Echo RVEDV was compared to the volume measured intraoperatively by the surgeon, at the time of repair under induced ventricular fibrillation, injecting saline solution through the tricuspid valve, up to the complete distension of the RV and to the complete closure of the tricuspid valve, after MPA cross-clamp at its origin. Correlation between the two measurements was very good, with a correlation coefficient ρ of 0,995 and $p = 0.0001$. The distortion between the two observations (calculated as the mean of the differences between echocardiograpic and surgical volumes) was 5.01 ml/m^2, the echocardiographic measurements overestimating by 4–9% the surgical ones. This overestimation can be justified considering that 3D echocardiography measures the RVEDV, while the surgical volume is evaluated in a fibrillating heart, not completely relaxed. We conclude that 3-D reconstruction of the echocardiographic images is a reliable tool in evaluating RV volumes and pumping function in pediatric patients, and we propose this method for the clinical use.

P148
Myocardial perfusion abnormalities after surgical repair with the aid of cardiopulmonary by pass (CPBP) in infancy
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In order to evaluate the late results of cardiac surgical procedures carried out with the aid of CPBP in infancy, we have performed sestaMIBI myocardial perfusion scan after pharmacologically induced stress by adenosina and at rest in 33 children (M17, F16) after repair of: TGA (arterial switch) (22), ToF (7), VSD (3), AP window (1), at a mean follow-up period of 8 years. Coronary artery angiography was also performed in 4 patients after arterial switch operation (ASO). Presence of myocardial perfusion defects was related to the following variables: age and weight at surgery, duration of CPBP, duration of cross clamp, use of deep hypothermic circulatory arrest, minimal rectal temperature, doses of cardioplegic solution employed, identification of perfusionist and surgeon. Myocardial ischemia after pharmacologically induced stress was present in 14 patients (42%): 4/7 (57%) after ToF repair, 8/22 (36%) after ASO, 1/3 (33%) after VSD repair, 1/1 after AP window repair. It was not reversible at rest in 2 patients with TGA, 1 with ToF and 1 with aortopulmonary window. Coronary angiography in 4 patients with perfusion defects after ASO disclosed central coronary arteries and major branches within normal limits. Statistical analysis showed that the use of deep hypothermic circulatory arrest increases by 14 times the risk for myocardial ischemia at follow-up ($p = 0.021$, Ho 5%, OR 13.93). No other variables reached statistical significance. We conclude that: (1) myocardial perfusion abnormalities at myocardial scan are common after surgical correction with the aid of CPBP in infancy, (2) manipulation of the coronary arteries as it is in ASO does not seem to have any role in the genesis of such a finding, (3) among the considered variables deep hypothermic circulatory arrest only increased significantly the risk of myocardial ischemia at follow-up.

P149 (see Abstract 104)
Left-sided atrioventricular valve insufficiency after AVSD correction
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P150
Hand-carried echocardiography in pediatric cardiology
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Introduction: In clinical practice several questions can be answered without extensive echocardiography. Recently, hand-carried (HC) echocardiography devices have been introduced. These devices are attractive because of their size, portability and cost. The use of these devices has not been evaluated in young patients.
Methods: In the present study we compared a HC device (Optigo; Philips Medical Systems) with a standard echo (SE) machine. (Sonos 5500, Andover, Massachussetts; Philips Medical Systems). Thirty-one consecutive patients were examined with both the HC device and the SE machine.
Results: Median age of the patients was 2.5 years (range 1 month to 17 years), median body weight was 16 kg (range 4 to 62 kg). Among the 31 patients 69 questions (1–4 per patient) had to be answered. The HC machine gave a correct result in 56/69 questions, while 13 were false (19%). We subdivided the questions into six categories: the presence or absence of valvular stenosis or valvular insufficiency, the presence and location of a ventricular septal defect, ventricular function, the presence or absence of pericardial effusion and a rest-group. The rate of missed diagnosis ranged from 0% in diagnosing ventricular septal defect (VSD) or pericardial effusion to 24% in diagnosing valvular insufficiency.
Conclusions: Although there are at the moment restrictions about the use of HC machines in pediatric cardiology our study showed that about 80% of clinical questions could be answered. This is even better for certain categories as pericardial effusion and the presence of a VSD. Furthermore, because of its low weight and compactness the present HC machine was very easy to handle. Therefore, this machine certainly merits a place in pediatric cardiology.

P151
Anti-inflammatory treatment for carditis in acute rheumatic fever: a systematic Cochrane Review
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Objective: To assess the effectiveness of anti-inflammatory agents in preventing or reducing further heart valve damage in patients with acute rheumatic fever.
Method: A meta-analysis was performed using the data extracted from randomised controlled trials comparing anti-inflammatory agents such as aspirin, steroids and immunoglobulins with placebo or controls, or comparing any of the anti-inflammatory agents with one another, in patients with acute rheumatic fever. The presence of cardiac disease one year after treatment was the major outcome criteria selected.
Results: Only eight randomised controlled trials (participants = 996) met the inclusion criteria. Several steroidal agents viz. ACTH, cortisone, hydrocortisone, dexamethasone, prednisone, and intravenous immunoglobulin were compared to aspirin, placebo or no treatment in the various studies. Six of the trials were conducted between 1950 and 1965, whilst the remaining two were done in the last 10 years. Overall, there was no significant difference in the risk of cardiac disease at one year between the corticosteroid-treated and aspirin-treated groups (RR 0.87, 95% CI: 0.66–1.15). Similarly, use of prednisone (RR 1.78, 95% CI: 0.98–3.34) or intravenous immunoglobulins (RR 0.87, 95% CI: 0.55–1.39) when compared to placebo did not reduce the risk of developing heart valve lesions at one year.
Conclusion: There is no benefit in using corticosteroids or intravenous immunoglobulins to reduce the risk of heart valve lesions in patients with acute rheumatic fever.

P152 (see Abstract 123)
Normal values of B type natriuretic peptide in infants, children, and adolescents
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P153 (see Abstract 118)
Prenatal closure of the fetal ductus arteriosus – result of thoughtless pain medication?
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P154 (see Abstract 93)
Enhanced visualisation of the right ventricle by contrast echocardiography in congenital heart disease
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P155
Long-term performance and survival of a steroid-eluting bipolar epicardial pacing lead in children
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Background: Cardiovascular anatomy or small patient size may necessitate the insertion of epicardial pacing leads. Initial studies showed reliable pacing with epicardial bipolar steroid-eluting pacing leads. However, the long-term performance of these leads is uncertain. We analysed long-term survival, impedance and threshold changes of these leads.
Methods: In 56 children, aged 4.9 + 4.8 years 95 epicardial leads (39 atrial, 56 ventricular) were implanted (Medtronic CapSure Epi 10366 or 4968) and connected to various pulse generators. Indications for pacing were postoperative heart block in 27, congenital heart block in 13, sinus node disease in 10 and various in 6 cases. Threshold values and measured data were obtained at 6 months intervals. The mean follow-up was 3.2 + 2.5 years. Statistical regression analyses was done using a GEE regression model. Threshold data were adjusted by the Energy formula $E = 1000 \times \text{voltage}^2 \times \text{pulsewidth/impedance}$ to uniformly compare the data between patients.
Results: Lead survival for atrial leads was 100%, 92%, 87% and for ventricular leads 100%, 94%, 83% at 1, 3 and 5 years respectively. Atrial and ventricular leads demonstrated no significant changes over time in impedance, energy and sensing thresholds (see table).
Conclusion: Long-term follow-up data demonstrate high survival rates for bipolar steroid-eluting epicardial pacing leads with low pacing thresholds and stable sensing thresholds and impedance. Thus, reliable long-term pacing can be assured with these leads.

P156
Adult patients after total repair of tetralogy of fallot: does pulmonary valve insufficiency or QRS prolongation correlate with arrhythmia heart rate variability or QT dispersion
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Introduction: Patients after total correction of Fallot's Tetralogy (TOF) – the group of increased risk of malignant ventricular arrhythmia. Prolongation of QRS duration over 180 ms and/or presence of significant pulmonary regurgitation (IP) are recognized as indicators of risk of this event in that group. Aim of the study was to evaluate cardiac arrhythmia, heart rate variability parameters (HRV) and QT dispersion (QTd) in adult repaired TOF patients with respect to IP and QRS duration.
Material and Methods: Study group : 19 men and 23 women, aged 16–44 years (25.72 ± 6.42), who underwent TOF repair at age of 3–35 years (8.57 ± 6.34), 4 to 29 years ago (16.07 ± 5.02). In all 12-lead ECG for assessing QRS duration and QTd, echocardiography and 24-hours Holter ECG were performed. Control group – 36 volunteers (20 women, 16 men) aged 16–44 years (27.3 ± 8.7).
Results: All patients: sinus rhythm with right bundle branch block. Severe ventricular arrhythmia was observed in 12% of them, more often in patients with significant IP and despite of QRS duration. QTd was 20–180 ms in study group (71.6 + 36.9) and was significantly greater than in controls (p-0.01). The values of SDNN, SDANN-i, SDNN-i were significantly lower in study than in control group (p-0.01, p-0.01, p-0.02), but there were no significant differences for rMSSD or pNN50. There were no significant differences in the values of HRV parameters nor QTd in subgroups of TOF patients distinguished by presence or absence of significant IP nor QRS > 180 ms.
Conclusions: 1/Severe ventricular arrhythmia is observed in 12% of adult repaired TOF patients, mainly with significant pulmonary regurgitation, despite of QRS duration time. 2/24-hours HRV parameters were significantly lower in adult repaired TOF patients and QTd is significantly greater in this group than in controls, which may indicate increased risk of malignant ventricular

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arrhythmia. There were no significant differences in mean values of these parameters according to presence or absence of significant pulmonary regurgitation, nor prolongation of QRS > 180 ms.

P157
Catheter ablation in patients with Wolff-Parkinson-White syndrome, tricuspid atresia, and Fontan circulation
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Aims: Due to scarring and dilatation of the right atrium numerous patients with Fontan circulation suffer from atrial flutter or fibrillation which are very resistant to treatment. We report on electrophysiologic studies in patients with tricuspid atresia after Fontan operation, and WPW syndrome, in whom atrial arrhythmia triggered atrioventricular reentrant tachycardia or caused life-threatening tachycardia.
Methods and Results: Four patients with tricuspid atresia after palliation with a modified Fontan operation (atrio-infundibular connections), and WPW syndrome are presented. Three of them showed paroxysmal orthodromic AV reciprocating tachycardias several times a day and had a history of syncope. A fourth patient presented with a life-threatening tachycardia during atrial fibrillation. In electrophysiologic studies an accessory pathway was localized in the right septal area in three patients. The fourth patient had three accessory pathways at the surgical suture line of the atrio-infundibular connection. All patients could be treated successfully with catheter ablation.
Conclusions: Patients with Fontan circulation and WPW syndrome an accessory pathway should be treated with catheter ablation. In patients with an atrio-infundibular anastomosis an acquired accessory pathway crossing the anastomosis must be considered. In patients planned for total cavopulmonary anastomosis by an extra- or intracardial conduit congenital accessory pathways should be excluded carefully before surgery.

P158
Outcome of neonates with pulmonary atresia and intact ventricular septum (PAIVS) after transcatheter and/or surgical treatment
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From 1994 to 2002, 24 neonates (11 females and 13 males) affected by PAIVS were admitted to our Department. The age ranged from 1 to 18 days and the weight from 2.5 to 4.2 kg. Eighteen patients (pts) underwent interventional procedure with an attempt of radiofrequency perforation (RFP), 5 pts underwent surgery for associated coronary artery anomalies and severe hypolasia of the RV and 1 died spontaneously for extreme dysplasia of the tricuspid valve. In 13 pts the RFP was successfully performed and 7 of them (54%) were discharged in 16–28 days without any other procedure; 2 pts died (15%) for heart failure after RFP and 4 pts (31%) had surgery for persistent severe cyanosis. Two of these 4 pts (mBT shunt) were discharged in 28 days and the other 2 (mBTshunt and outflow patch) died due to post-operative complications. The 9 surviving pts (7 RFP, 2 RFP + mBT shunt) are in follow-up (6–98 m) and 8 of them have had complete recovery of the RV while 1 pt underwent cavopulmonary connection. In 5 pts the attempt to perforate the valve failed and they underwent surgical valvotomy and mBT shunt with immediate good results. During the follow-up it was possible to close the mBT in 4 pts and 1 pt is awaiting a new evaluation. Four of the five pts (80%) who underwent first-choice surgery for unfavourable anatomy died after the mBT shunt and only 1 of them is waiting for bidirectional cavopulmonary connection. In conclusion, RFP is a definitive treatment in favourable cases and a good palliative one in less favourable cases in combination with mBT shunt. The outcome of pts is greatly influenced by the anatomy of the lesion.

P159 (see Abstract 102)
Transcatheter treatment of aortic coarctation. Fifteen years experience
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P160 (see Abstract 124)
Cardiac involvement and myocardial microcirculation disturbance in children with systemic sclerosis
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P161
The echocardiographic evaluation of right ventricular diastolic function in infants and young children with secundum atrial septal defect
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Aim of the Study: The analysis of echocardiographic indexes of diastolic functions of right ventricle (RV) in infants and young children with secundum atrial septal defect (ASD II). The next aim was to establish influence of such clinical indexes as age, weight, height, and body surface of the child, as well as such essential hemodynamic indexes as the size of ASD II, pulmonary to systemic flow ratio (Qp/Qs), pulmonary blood flow index (PBFI), RV end-diastolic volume index (RVEDVI) and RV end-systolic volume index (RVESVI) to disturbances of RV diastolic function in infants and young children with symptomatic ASD II.
Materials and Methods: The study population consisted of 117 patients, 57 infants and 60 children in second year of life, 44 boys and 73 girls. The population age ranged from 4 months and 12 days to 23 months and 8 days (mean 11 months and 10 days ± 5 months and 24 days). This population was divided into three groups: a group with symptomatic ASD II, a group with nonsymptomatic ASD II and a group of healthy infants and young children. Then, the group with symptomatic ASD II was divided into two subgroups depending on the ratio of peak early (E) velocity to peak atrial (A) velocity of tricuspid inflow (E/A vel). All hemodynamic indexes were performed by means of Doppler echocardiography.
Results: Comparison of groups and subgroups of patients revealed abnormal diastolic function of RV in 43% of infants and young children with symptomatic ASD II. There were no significant correlations between both clinical indexes and RV diastolic function indexes as well as such essential hemodynamic indexes and RV diastolic function indexes in this group of patients.
Conclusions: In 43% of infants and young children with symptomatic ASD II there are disturbances of RV diastolic function. These disturbances are independent of the age and the level of

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development of the child. Such indexes as the size of ASD II, Qp/Qs, RVEDVI and RVESVI do not affect disturbances of RV diastolic function in infants and young children with symptomatic ASD II.

P162
Interventional occlusion of right-to-left shunts after univentricular palliation of congenital heart defects by use of Starflex, Amplatzer and Helex-occluders

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Background: Arterial hypoxemia is frequently observed after univentricular palliation of congenital heart defects. Whereas small veno-venous collaterals are easily treated by coil-implantation, occlusion of large collaterals and atypical fenestrations may be technically challenging.
Purpose: To report our experience with device occlusion of large-diameter collaterals and an atypical fontan fenestration connecting the right atrial appendage to an extracardiac conduit.
Patients: 3 patients were treated after univentricular palliation of congenital heart defects for arterial hypoxemia with arterial oxygen saturations ranging from 72% to 82%. Large-sized collateral vessels were identified in 2 patients originating from the superior vena cava or an excluded hepatic vein. In one patient, a fenestration was placed connecting the right atrial appendage to the extracardiac conduit.
Methods: After test-occlusion of the right-to-left shunts and hemodynamic evaluation, the excluded hepatic vein was occluded by use of a 10/8 mm Amplatzer Duct Occluder; the large collateral vessel connecting the superior vena cava to the right upper pulmonary vein was occluded by use of a 33 mm Starflex device and the atypical fenestration was occluded by use of a Helex 15 mm occluder which was implanted by use of a long premounted sheath after removal from its delivery catheter.
Results: Arterial hypoxemia was relieved in all patients by device implantation without peri-procedural complications. Arterial oxygen saturations increased from 72%–82% prior to the intervention to 95%–98% after the intervention. Pulmonary arterial pressure remained low.
Conclusion: In patients after univentricular palliation, transcatheter device occlusion of large-sized collateral vessels is a safe and effective alternative to surgical occlusion. A variety of devices should be available to adjust therapeutic strategies for different morphological features of such connections.

P163
Use of MRI in complicated Kawasaki disease in children

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Introduction: Coronary artery aneurysm and stenosis develop in a significant percentage of patients despite treatment. Therefore in long-term management of patients with coronary aneurysm (CAA)/stenosis, accurate assessment of coronary artery anatomy is important in formulating the plan of management. MRI coronary artery has the advantage of noninvasive assessment of coronary artery anatomy in Kawasaki disease complicated by coronary artery aneurysm. This study aim at defining the usefulness of MRI in assessing coronary artery aneurysm or stenosis compared to coronary angiography in patients with coronary artery aneurysm/stensois complicating Kawasaki disease.
Patients and Methods: Eight patients age ranging from 7 years to 16 years with a history of Kawasaki disease complicated by coronary artery aneurysm were enrolled. The disease activity is quiescent. The follow-up period ranges from 3 months to 12 years. MRI coronary arteries were performed from January to September 2001 in a single center. All patients underwent cardiac catheterization and transthoracic echocardiogram within three years of performing MRI coronary artery.
Results: There were seven males and one female patient. Their age ranges from 7 years to 16 years. All had history of Kawasaki disease with moderate to giant coronary artery aneurysm. Their follow-up period ranged from 3 months to 12 years. MRI detected coronary artery aneurysm (CAA) in 8 cases whereas CCA was documented by coronary angiography in 6 cases only. The sensitivity being 100% but the specificity was 0% in detecting coronary artery aneurysm. Coronary artery stenosis/occlusion was not detected in all the 5 patients, which was detected by coronary angiography. The sensitivity was 0% and the specificity a 100% for detecting coronary stensois/occlusion by MRI.
Conclusion: MRI angiogram can show up coronary aneurysm better than echocardiogram. However MRI cannot detect stensois nor total occlusion nor could it be detected by echocardiogram. Hence coronary angiography is the recommended modality of investigation for delineating coronary abnormality in complicated Kawasaki disease.

P164
A unique occurrence of hypoplastic left heart syndrome in dizygotic twins

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Congenital cardiovascualr malformations in twin and triplet pregnancies are estimated to occur with a frequency of 2% in dizygotic pairs, but up to 10% in monozygotic pairs. Hypoplastic left heart syndrome (HLHS) is well documented to occur within families. Concordant HLHS lesions have also been described in monozygotic twins. This suggests a strong genetic influence in the development of cardiac defects in twins. Some authors propose that the twinning process itself may be implicated in the development of cardiac malformations. This case report documents the unique occurrence of HLHS occurring in a set of dizygotic twins, never before reported in the English language literature. The twins were born at 36 weeks gestation to a 25 year old gravida 3, para 2 mother. She had had no previous history of miscarriages, and no family history of congenital heart disease. The male twin presented on day 1 of life with cyanosis and sock. He was admitted to neonatal intensive care for ventilation and intravenous prostaglandins. Echocardiography showed the typical anatomy of HLHS with mitral and aortic atresia, aortic arch hypoplasia, with a dilated right ventricle and a patent ductus arteriosus to descending aorta. The female twin, initially well, presented with cardiovascular collapse on day 3 of life. Echocardiography showed the identical anatomy to the sibling. Both were managed conservatively once the diagnosis was made. Subsequent autopsy confirmed the echocardiographic findings. Chromososmal analyses on both siblings was normal, as was the FISH analyses for del.22q11 syndrome. Histologic findings on the female twin showed features of acute tubular necrosis, with pulmonary emboli. The male twin who was thrombocytopenic, had features of cytomegalovirus pneumonitis, nephritis and sialo-adenitis.

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P165
Activated renin–aldosterone–angiotensin system in pediatric patients with ventricular septal defect
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Background: Renin–aldosterone–angiotensin (RAA) system was thought to be involved in pathology of congestive cardiac failure. However, there was no human study on change of RAA system in pediatric patients with ventricular septal defect (VSD).
Purpose: To determine the change of RAA system in patients with VSD.
Material and Methods: Twenty-three pediatric patients with VSD were enrolled. After 30 minutes of bed rest, blood samples were obtained from femoral vein. Plasma renin activity (PRA) and serum concentration of aldosterone (ALD) and angiotensin converting enzyme (ACE) were determined. Routine cardiac catheterization was performed to determine hemodynamic variables. Then, we determined the correlation of hemodynamic variables and levels of these hormones and enzymes. Levels of these hormones and enzymes were compared between patients with and without pulmonary to systemic flow ratio (Qp/Qs); 1.5 : 1. In addition, levels of these hormones and enzymes were compared between patients with and without signs of congestive heart failure including hepatomegaly 2 cm below costal margin and respiratory retraction.
Results: Patients' age ranged from 0 to 12 years old (median 1.0) and pulmonary to systemic flow ratio ranged from 0.9 to 4.1 (median 1.7). PRA positively correlated with ALD ($r = 0.84$, $p < 0.001$), but negatively correlated with ACE ($r = 0.50$, $p < 0.01$). PRA and ALD in patients with Qp/Qs 1.5 : 1 were significantly higher than in patients without. PRA positively correlated with systemic vascular resistance ($r = 0.46$, $p < 0.03$) as well as pulmonary to systemic pressure ratio ($r = 0.49$, $p < 0.02$). Patients with hepatomegaly showed significantly higher PRA and ALD than patients without (median 14 vs. 8 ng/ml/hr, $p < 0.03$ for PRA and 410 vs. 175 pg/ml, $p < 0.01$ for ALD) and patients with respiratory retraction showed significantly higher PRA and ALD than patients without (20 vs. 8 ng/ml/hr, $p < 0.03$ for PRA and 340 vs. 180 pg/ml, $p < 0.03$ for ALD).
Conclusions: In patients with moderate VSD, RAA system was activated with increasing systemic vascular resistance and increasing pulmonary blood pressure and involved in the pathology of congestive heart failure of these patients.

P166 (see Abstract 127)
Double orifice mitral valve (DOMV) – clinical presentation, natural history and outcome in children and adolosence
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P167
Value of contemporary myocard bioptate specimens analysis in the diagnostic flow diagram for primary cardiomyopathies in children
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The purpose of this work was to show the importance of myocard bioptate analysis using different methods in diagnosis of primary cardiomyopathies (CMP) in children. According to the guidelines of the Task Force on Cardiomyopathies of the WHO and the ISF of Cardiology, we identified 135 infants children and adolescents with CMP, 55 female (40.7%) and 80 male (59.3%), giving an average occurrence for all CMP of 43.32 on 10,000 patients examined in our outpatient paediatric cardiology clinics. Dilated CMP was found in 52 patients (38.5%), hypertrophic CMP 43 patients (31.9%), and restrictive CMP in 6 patients (4.4%). In nine patients (6.6%) were impossible to classify the cardiomyopathy. The majority of dilated CMP diagnosis were made before the age of 3 years (RR 1.9, 95% CI 1.4–2.47). There were no statistically significant differences in the incidences of dilated and hypertrophic CMP (Z 0.923, p 0.1779), but we encountered significantly lower occurrence of restrictive CMP (Z 6.044, $p < 0.001$). The biopsy of endocard and myocard was done in 22 patients, (12 male and 10 female), age 1 to 17 (average 6.3 yrs). The specimens were examined by light microscope (Dallas criteria) in all patient, in 13 cases direct immunofluorescency and immunohystochemical methods were used, two were examined hystochemically, one by the microscopy in polarised light and 7 by electron microscope. Five bioptates were analysed by PCR method (DNA and RNA of cardiotrophic viruses). Four of ten children with dilated cardiomyopathy had myonecrosis as a consequence of acute myocarditis, and six of them developed signs of late inflammatory processes: immunoglobulin and complement deposits due to chronic immunologic myocarditis. In 4 patients rebiopsy proved complete healing (corticosteroide therapy). In 5 patients the hypertrophic CMP diagnosis was confirmed hystologicaly, one was additionally analysed by electron microscope to rule out the possibility of mytochondriopathy. The finding correlated with the pathology of neuromuscular system (scapulo-peroneal amiotrophy). Pathohystological findings of a bioptate specimens in patients with restrictive cardiomyopathy, showed primary amiloidosis of the heart in one case, fibroelestosis in one another, in 3 children inflammation, and in one case heart tumor (fibroma) was found.

P168 (see Abstract 107)
A new clip device for construction of vascular interrupted anastomosis in congenital cardiac surgery
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P169
Percutaneous closure of moderate-to-large arterial duct (above 2.5 mm). What is the best occluder? Experience with 4 different devices in 116 consecutive patients
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Design of previously employed occluders is not ideal for transcatheter closure of moderate-to-large arterial duct, and their use has been associated with several complications.
Method and Results: This study was performed to compare results of transcatheter closure of arterial duct (>2.5 mm) using the Amplatzer Duct Occluder (ADO) and previous occluders. One hundred and sixteen patients (77 females, 39 males) underwent closure with several devices: Rashkind double umbrella (n = 23, group I), Sideris buttoned device (n = 39, group II), detachable Cook coil (n = 17, group III), and ADO (n = 37, group IV) at a mean age of 74 ± 129 months (weight: 18.9 ± 15.5 kg). Ductal

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diameter was 3.8 ± 1.22 mm. Implantation succeeded in all but 9 (3 in group I, 2 in group II, 3 in group III, and 1 in group IV). Time of fluoroscopy was shorter and full occlusion better on angiography (64%) in group IV despite larger ducts (respectively $p < 0.0001$, $p = 0.0003$ and $p = 0.0015$). Complications included embolisation ($n = 2$, 1 in group I and 1 in group III) and haemolysis ($n = 3$, 2 in group III and 1 in group IV). During follow-up, 12 patients had a second procedure because of residual shunting (5 in group I, 6 in group II, and 1 in group III). Complete occlusion was achieved earlier after implantation (mean 2.6 months, $p = 0.0002$) and rate of complete occlusion was better in group IV (97%, $p = 0.025$).
Conclusions: Transcatheter closure of arterial duct with the ADO is an effective and safe method providing better results than previous occluders. It could be highly recommended for closure of large arterial ducts (>2.5 mm).

P170
Early clinical results of the telemetric adjustable pulmonary artery banding FloWatch™-PAB
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Background: Adjustment of pulmonary artery banding (PAB) may be a challenging procedure in complex congenital heart defects. Whatever the technique used, subsequent re-operations are frequently needed to control the pulmonary blood flow or pressures.
Objective: To determine the safety and performance of a new telemetric adjustable PAB operated with the help of an external control unit that transmits to the implant both energy and commands to further narrow or release the pulmonary artery using radiofrequency waves.
Study Design: Multicenter, prospective, non-randomised single arm clinical investigation.
Patients and Methods: 9 children (median age 3.6 months (range 3 weeks–31 months); median weight 6.9 kg (range 3.5–11 kg)) underwent implantation of the FloWatch™ through median sternotomy in 6 and left thoracotomy in 3. The diagnosis was: multiple ventricular septal (VSD) defects with complex anatomy in 3, single ventricle without pulmonary stenosis in 2, VSD with elevated pulmonary vascular resistance (PVR) in 1, atrio-ventricular canal (AVC) with elevated pulmonary vascular resistance in 1, AVC with diminutive right ventricle in 1, transposition of the great arteries with VSD in 1. All patients had isosystemic systolic pulmonary artery pressure. Additional procedures were performed in 4: atrial septectomy in 2, double aortic arch division in 1, patent ductus arteriosus ligation in 2.
Results: There were no early or late death, nor device-related complications, in a mean follow-up of 16 weeks (range 12–25 weeks). A mean of 5/patient telemetric regulations of the FloWatch™ were required to adjust the tightening of the PAB to the clinical needs (narrowing 79%, releasing 21%). At last follow-up, systolic pulmonary artery pressure was within normal range in all patients (mean 23 ± 5 mmHg). Systemic oxygen saturation demonstrated optimal regulation of the pulmonary blood flow in all according to each specific defect. Two patients were successfully corrected (VSD closure after 4 months, and arterial switch with VSD closure shortly after implantation). The device was easily removed and the pulmonary artery re-expanded spontaneously.
Conclusion: This new device is safe and allows optimal adjustment of PAB in complex heart defects. In children requiring PAB the therapeutic strategies can be expanded by the use of this promising technology.

P171
Particularities in cardiac involvement in children with systemic lupus erythematosus
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Abstract: The SLE disease-typical dyslipidemia, systemic arterial hypertension, increased thrombosis tendency, antiphosholipid antibodies, coronary vasculitis are all mensurable factors prompting an early control but until now they are not a worry in lupic children.
Objectives: Analyse cardiac involvement in a group of asymptomatic children and young adults with SLE, identify the presence of coronary disease risk factors, study their particularities.
Method: Thirty lupic pediatric patients were evaluated under retrospective and transverse study, echocardiographic assessment, scintilography with MIBI under physical stress and investigation of the risk factors for coronary artery disease.
Results: Systemic arterial pressure levels adequate in 53%. Biopsy results renal compromise in 77%. Total cholesterol levels >P90 in 33%; HDL-C <P50 in 67%, triglyceride level >P90 in 53%. The TC/HDL ratio was high in 20%. Several coronary risk factors were found in multiple association in a single patient. Sinus tachycardia 46%, repolarization alterations 60%, hypokinesia and/or cardiac chamber enlargement in 20%. Fifty percent of the group with low to moderate grades of valve insufficiency, anticardiolipin antibodies were detected in 60%, mitral valve involvement (17%). Pericardial involvement was now 3% and in the initial stage in 40%. Myocardial perfusion study revealed 20% with alterations (three patients with persistent uptake area, one with transient ischemic episode).
Conclusions: To permit the individual assessment and selection of the ideal patient management is usefull routine cardiovascular investigation in children and young adults with SLE through regular electrocardiogram, follow-up echocardiographic studies, screening for antiphospholipid antibodies and strict control of the modifiable CAD risk factors.

P172 (see Abstract 128)
Preoperative immunological and serological differences between children with and without post-surgical capillary leak syndrome
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P173
Induction therapy with Daclizumab induction in pediatric heart transplantation
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Daclizumab binds specifically to the Tac subunit (CD 25) of the human high-affinity interleukin-2 (IL-2) receptor that is expressed on the surface of activated lymphocytes. It functions as an IL-2 receptor antagonist inhibiting IL-2-mediated stimulation of lymphocytes, a critical event in the process of allograft rejection. The aim of this study was to evaluate the effect of a Daclizumab induction therapy in heart transplanted children compared to heart transplanted children without Daclizumab induction therapy. Daclizumab was given as induction therapy to 7 heart transplanted children

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(patients (pts) = 7, age 8.7 ± 7.4 yrs, weight 23 ± 12.2 kg, BSA 0.86 ± 0.36 m^2). 1 mg/kg Daclizumab doses were administered on day 0 (0.5 mg/kg before and 0.5 mg/kg after heart transplantation), day 7, day 14 and day 21. Standard immunosuppression consisted in 7 pts of cyclosporine (CsA), mycophenolate mofetil (MMF) and steroids, in 1 pt of tacrolimus, MMF and steroids. In the control group (pts = 24, age 6.9 ± 6.1 yrs, weight 20.6 ± 15.6 kg, BSA 0.82 ± 0.44 m^2) standard immunosuppression in 20 heart transplanted children consisted of CsA, MMF and steroids, 4 pts received tacrolimus instead of CsA. CD 25 ± lymphocytes were totally blocked for a period of several months (44–180 days). During a follow up time of 12.4 months there was no rejection of the grafted heart in the Daclizumab induction group. The 1 year patient and graft survival was 100%. In the control group 12,5% (3/24) pts had a rejection episode, one of them died. Regarding the different individuals of each pt group a niveau of significancy can not be determined. Pt and graft survival of the control group in a follow-up time of 5.5 yrs was 95,8%. There were no clinical side effects in the pts with Daclizumab induction therapy. There was no difference between the two groups regarding infections and malignancies (esp. lymphomas). These results indicate a tendency towards less rejection episodes in establishing an induction therapy with Daclizumab in pediatric heart transplantation. Daclizumab has proven to be safe and effective in preventing organ allograft rejection in pediatric heart transplantation when administered with 4 applications of 1 mg/kg on day 0, 7, 14 and 21.

P174 (see Abstract 126)
Conversion from cyclosporine A to tacrolimus after pediatric heart and heart lung transplantation
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P175 (see Abstract 100)
Stent implantation into the arterial duct in lambs
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P176
First case report of percutaneous closure of a coronary sinus defect with an Amplatzer septum occluder
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Introduction: Coronary sinus (CS) defect is an unusual interatrial communication, it is almost always associated with a persistent left superior Vena Cava (LSVC). The hemodynamic consequence of this association is a certain degree of left-to-right shunting with dilatation of the right atrium and the right ventricle. The management this type of congenital anomaly is usually surgical. We report a successful percutaneous closure of a coronary sinus defect in a young boy with an Amplatzer septum occluder (ASO).
Case Report: A 9.5 years-old caucasean boy with a body weight of 31 Kg was referred to our department for therapy of a CS defect. The child presented with the clinical, chest X-ray and ECG signs of right heart dilatation due to a left-to-right shunt. Echocardiography demonstrated a 5 mm moderate defect with left-to-right shunt, located in the distal part of the CS roof. There was a small LSVC coming from an inominate vein draining through the CS. The cardiac catheterisation was performed under general anaesthesia and under guidance of transoesophageal echocardiography (TOE). The defect's localisation was demonstrated with angiography and sized with a standard Berman balloon catheter. The defect measured in TOE and by balloon sizing 6 mm. To close the defect, an 6 mm ASO was chosen. The device implantation was performed like a standard ASD. Because of the presence of the LSVC the intervention could be easy controlled, with angiographies into the CS with a catheter placed via the LSVC, without the risk of dislocation of the device. The closure could be successfully performed, no residual shunt could be observed during follow up, nor an obstruction of the CS or the mitral valve inflow.
Discussion: Selected CS defects might be also amenable to a percutaneous closure with ASO. The ASO is an ideal device for such atypical located ASDs because of its flexibility and specific safety reserve, allowing to withdraw the device, as long as it is attached to the implantation wire if there rises any complication.

P177
Aortic and pulmonary flow velocity between 12 and 18 weeks gestation measured by transabdominal ultrasound
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Objectives: To measure the flow-velocity waveforms across aortic and pulmonary valves in fetuses between 12 and 18 weeks of gestation, using transabdominal Doppler echocardiography, to define the blood flow in the normal fetus and evaluate changes occurring in the flow-velocity waveforms during this gestational period.
Methods: Cross-sectional data and cardiac Doppler were collected from 86 healthy human singleton fetuses between 12 and 18 weeks of gestation. Flow-velocity waveforms from the fetal ascending aorta were recorded from the five-chamber view and fetal pulmonary artery flow-velocity waveforms were obtained from the echocardiographic short-axis view. The sample volume was placed immediately distal to the semilunar valves. To obtain maximal velocities, the only results used in this study were from fetuses in whom it was possible to have the ultrasound beam parallel to, or at an angle of <20 degrees to the presumed blood flow orientation of the peak flow velocities were measured using the highest modal velocity from three consecutive beats. Fifteen sets of measurements were obtained again by the first investigator (CF) and a second observer (NHS) to define inter- and intraobserver variability. Variability was expressed as difference from the percentagewise mean of the two results. Linear regression was used to establish the relationship between gestational age and aortic flow velocity and pulmonary flow velocity.
Results: Fifty-eight records of flow through the aortic valve and fifty-nine records through the pulmonary valve were obtained. There was a significant relationship between gestational age and flow velocity in the outflow tract of the left ventricle ($r = 0.565$; $p < 0.001$) and in the outflow tract of the right ventricle ($r = 0.422$; $p = 0.01$). Intraobserver variability was 0.6% and interobserver variability was 1.7%.
Conclusions: Cardiac flows through the outflow tract of both ventricles during the early stages of pregnancy may be analyzed by means of transabdominal echocardiography, and may offer an important contribution to the detection and follow up of cardiac abnormalities.

P178 (see Abstract 117)
Doppler evaluation of cardiac function in trisomy 21 and normal fetuses prior to 14 weeks of gestation
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P179

Proximal isovelocity surface area method for assessing rheumatic mitral regurgitation in children

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Objective: The purpose of this study was to determine the feasibility and significance of the proximal isovelocity surface area (PISA) method in children with rheumatic mitral regurgitation (MR).
Methods: Thirty one patients (14 boys and 17 girls, mean aged of 12.3 ± 3.1 years old), with chronic isolated rheumatic MR were evaluated concomitantly by semiquantitative and quantitative Doppler, quantitative two-dimensional echocardiography and PISA methods. Also we compared regurgitant orifice area (ROA), regurgitation volume (RV), systolic and diastolic left ventricular functions and variables reflecting volume overload in group I (grade 1 and 2 MR) and group II (grade 3 and 4 MR).
Results: There were no statistically significant differences in ROA and RV values obtained by PISA method and the quantitative Doppler ($t = 0.421, p = 0.677$ and $t = 0.720, p = 0.477$) but they were different from the same values obtained by two dimensional echocardiography ($t = 5.024$ and $t = 3.992$, $p = 0.000$). There were excellent correlations between ROA, RV and the radii of the PISA hemisphere ($r = 0.882$, $r = 0.925$, $r = 0.880$, $p = 0.000$). We found a very good correlation between ROA(PISA) and left ventricle end-diastolic diameters ($r = 0.763, p = 0.000$), left atrial volume ($r = 0.751$, $p = 0.000$), ratio of the jet area and left atrial area ($r = 0.745, p = 0.000$). There was a good correlation between left ventricle mass index, mitral jet area color flow Doppler mapping at the MR, maximal jet length, cardiac index.
Conclusion: It is concluded that MR can be accurately predicted in children by using the PISA method as like as the Doppler method.

P180

Uselfulness of intracardiac ecocardiography with AcuNav catheter in percutaneous closure of atrial septal defect and patent foramen ovale: preliminary experience

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Background: Percutaneous closure of secundum atrial septal defect (ASD) and patent foramen ovale (PFO) are routinely performed under transoesophageal echocardiography (TEE) guidance and general anaesthesia. The AcuNav (Acuson) is a ultrasound-tipped phased-array catheter, 10 F diameter, frequency from 5.5 to 10 MHz, providing a 90 sector image. Our preliminary experience with AcuNav in percutaneous closure of ASD and PFO is described.
Material and Methods: Intracardiac echocardiography (ICE) was obtained with AcuNav using a standard ultrasound platform (Sequoia, Acuson). TEE was obtained in using HP Sonos 5500 system (Agilent Technologies). The device employed were Amplatzer Septal Occluder (ASO) or PFO Occluder (AGA MedCorp) or STARflex (NMT Medical). 3 patients (1 male, 2 female) had secundum ASD; 2 (1 male, 1 female) had PFO with history of TIA. The mean age was 39 years (13–56), mean weight 63 kg (50–105). The standard protocol of ASD and PFO closure was used. The AcuNav catheter was inserted in the left femoral vein through a 11 F standard sheath, the right femoral vein was the access for the devices delivery. In the first patient both TEE and ICE were performed in anaesthesia, in the remaining 4 only ICE was used, without anaesthesia.
Results: The ASD diameter ranged from 8–28 mm at ICE, the stretched diameter ranged from 8–40 mm; 1 patient had multiple ASD, another patient was not eligible for closure because the stretched diameter resulted of 40 mm, and multiple ASD were evident at ICE during the balloon occlusion. The mean procedure time was 55 minutes (45–155), mean fluoroscopy time was 15 minutes (10–25). In two patients one ASD and one PFO were closed with a 30 mm ASO and 35 mm PFO Occluder respectively. In the other two patients STARflex devices were used: 33 mm for the multiple ASD, and 24 mm for the PFO.
Conclusion: In our preliminary experience the ICE by AcuNav is similar to the TEE, the avoidance of general anaesthesia and a better imaging of the atrial septum with multiple fenestration are the potential advantages. Limitations are the catheter cost and, only for child, the sheath size.

P181 (see Abstract 129)

Slow kinetics of oxygen uptake during exercise in patients with a Fontan-type circulation

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P182

A novel use for improvised Amplatzer device in failing Fontan

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Fenestrating the Fontan circuit between the systemic venous compartment and pulmonary venous atrium is a common practice to augment the systemic output at the expense of desaturation, in patients with relatively high pulmonary vascular resistance. We report the use of fenestrated Amplatzer device for balancing systemic and pulmonary venous circulations. A 5-year-old, child with late presentation of complex cyanotic disease with mitral atresia and hypoplastic left ventricle, initially palliated with pulmonary artery band, atrial septostomy, rebanding, septectomy and cavopulmonary anastomosis was referred to our unit for further management. At cardiac catheter though his mean pulmonary artery pressure was 15 mmhg and was adjudged suitable for completion of Fontan operation. He underwent fenestrated (4 mm) extracardiac total cavopulmonary connection and tricuspid annuloplasty. He had a stormy postoperative period with unstable haemodynamics and large visceral effusions up to 2.5 litters of drainage per day. Subsequently he developed thrombosis in the conduit. Surgical thrombectomy and conduit replacement with a large (10 mm/5 mm) fenestration was made as a lifesaving measure. Following this he recovered slowly with low saturations (55–70%). Six weeks after his re-operation, cardiac catheterisation and graded balloon occlusion was performed to evaluate feasibility of reduction or occlusion of the fenestration. On complete occlusion the PA pressures were unacceptable with fall in systemic output and ischaemic changes on ECG. A custom made 8 mm Amplatzer device with a 5 mm fenestration was subsequently deployed to reduce the right to left shunt and to ensure adequate systemic output. The post procedure saturations were 90% without any haemodynamic deterioration. The child made excellent recovery and is doing well on follow up. This is the first time Amplatzer device has been used for reducing fenestration size in Fontan circulation. Though rarely

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encountered, feasibility of deploying such a device transforms the quality of life in these patients.

P183 (see Abstract 103)
Problems and complications during transcatheter closure of the moderate and large patent ductus arteriosus utilizing Amplatzer duct occluder in 22 patients
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P184
Depression of left ventricular systolic function shortly after closure of large patent ductus arteriosus
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Background: Shortly after transcatheter closure of a large patent ductus arteriosus (PDA), left ventricular (LV) function can be depressed.
Aim of the Study: We attempted to determine if changes in LV function were common after closure of large PDA's.
Methods and Patients: Between January 2000 and April 2002, a total of 43 patients underwent either transcatheter occlusion or surgical ligation of their PDA. Patients were divided into two groups. Group A ($n = 27$) consisted of patients with PDA diameter of >3.1 mm, and group B ($n = 16$) had PDA diameter <3 mm.
Results: The narrowest diameter of the PDA was a mean of 5.31 ± 2.3 mm in group A, while it was a mean of 3.23 ± 0.73 mm in group B; $p < 0.001$. Before the procedure, left ventricular end-diastolic diameter (LVEDd) in group A, was significantly larger than in group B; $p < 0.05$. At base line all patients had normal shortening fraction (SF). At short-term follow up, LVEDd showed a trend towards regression in diameter in group A; p (0.1–0.05), while it remained unchanged in group B. During the same follow up period, left ventricular SF reduced significantly in group A (37.38 ± 4.71% to 27.7 ± 4.6%; $p < 0.001$, while there was no change in the left ventricular SF in the other group (from 39.64 ± 5.8% to SF 35.13 ± 1.718%; n.s. The deterioration of SF in group A was significant and clinically relevant (changes > 20%) in 60% of the studied patients. Over a period of three to six months, there was improvement of LV function in the 16 patients studied in group A.
Conclusions: Deterioration of left ventricular function is observed after closure of the large PDA in children. Echocardiographic studies before discharge are mandatory in these patients.

P185
P wave dispersion in patients with correction of Tetralogy of Fallot
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Background: Atrial fibrillation and flutter frequently occurs in patients with correction of Tetralogy of Fallot (ToF). Recently, P wave dispersion has been used to evaluate the discontinuous propagation of sinus impulse and the prolongation of atrial conduction time. P wave dispersion was shown to be useful for determining patients who had atrial fibrillation history. In this study we evaluated the P wave duration and P wave dispersion in patients with correction of ToF.
Methods: The study group consisted of 25 patients with correction of ToF with the mean age of 16.4 ± 4.2 and 25 age-matched healthy control subjects. The mean duration of follow-up was 11.8 ± 1.77 years after correction surgery. All patients underwent 12-lead surface ECG. P wave duration and P dispersion (Pd = Maximum P wave duration – Minimum P wave duration) were calculated in all the 12 leads of the surface ECG simultaneously recorded. Left atrial diameter was measured using echocardiography in patients and controls.
Results: Maximum P wave duration (94.48 ± 13.25 ms, 86.20 ± 10.16 ms respectively, $p > 0.05$) and P dispersion (26.68 ± 10.82 ms, 17.28 ± 8.71 ms $p < 0.01$) were both found to be significantly higher in patients with correction of ToF than in control subjects. Maximum P wave duration was related to P dispersion ($r = 0.550$, $p < 0.001$), left atrial dimension ($r = 0.353$, $p < 0.05$), and duration after surgery ($r = 0.418$, $p < 0.05$).
Conclusions: Atrial conduction time is prolonged in patients with correction of ToF. Maximum P wave duration and P dispersion are simple ECG markers that might be useful for the prediction of atrial arrhythmias in patients after correction of ToF.

P186
Childhood acute rheumatic fever in Ankara, Turkey
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Background: Patients with acute rheumatic fever, who were admitted to the tertiary hospital in Ankara, during the January 1999 to July 2002 were studied cross-sectionally to verify the clinical profile and followed-up during the acute period.
Methods: All patients were investigated for throat culture, streptococcal serologic study, C-reactive protein, eritrosit sedimentation rate, telecardiograms, electrocardiograms and echocardiographic study.
Results: During the study period, 129 attacks of acute rheumatic fever were observed; 118 were initial attacks and 11 were recurrences. Age on admission was 11.2 ± 2.73 years (mean ± SD, range: 6–21 years). Polyarthritis alone was present in 42 cases (33%), carditis alone in 33 (25%), combined carditis and polyarthritis in 36 (28%), combined carditis and chorea in 15 (12%) and chorea alone was diagnosed in 3 (2%). Nineteen (14%) patients with isolated arthritis and pure chorea had silent mitral and/or aortic regurgitation demonstrated by only echocardiographic investigation but without any significant murmur. Mild to moderate carditis was present in 68 patients (80%). Otherwise severe carditis was diagnosed in 16 (19%); 5 of these patients also had pericardial effusion. Carditis was a dominant presenting manifestation, but appeared to be mild or moderate. The mitral valve was the most commonly affected valve (97%), followed by aortic valve (35%).
Conclusion: The present study indicates that ARF is still a significant problem in Turkey and the observation that 8.5% of the attacks were recurrent reaffirms the need for more effective secondary prevention programs.

P187
DRB1, DQA1, DQB1 genes in Turkish children with rheumatic fever
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Background: Several studies have suggested that genetic susceptibility to rheumatic fever (RF) may be linked to HLA Class II alleles. The objective of this study was to examine the association between

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HLA Class II genes and RF. Although, previous several HLA typing studies based on the serological method was performed, best of our knowledge, this study is the first report which use of the DNA typing technique in the Turkish patients with RF. Additionally, it is the first HLA-DQ typing study in the same population.
Methods and Results: DNA typing HLA Class II genes (DRB1, DQA1, DQB1) was performed in 55 children with RF and 50 healthy unrelated controls by using sequence specific primers (SSP). The frequencies of HLA DQA1*03 (OR: 0.462 $p < 0.05$) allele was significantly decreased in the patient group. A significant deviation in the frequencies of DRB1 and DQB1 alleles was not observed in the patient group.
Conclusions: Our data indicate that, HLA DQA1*03 allele may be a protection factor in RF, in Turkish children group.
Note: This study was supported by the TUBITAK.

P188
Spontaneous closure of secundum atrial septal defects in the era of device and surgical closure

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Aim: Secundum atrial septal defects (ASDs) with significant left to right shunt have been surgically closed to prevent complications. In recent years device closure has gained wide acceptance. The purpose of this study is to address the issues of probability of spontaneous closure and the likelihood of surgical and device closure in the current era.
Methods: All patients with isolated secundum ASD (512) identified from Echocardiography Registry and Cardiac Registry of Singapore Baby & Child Clinic from August 1988 to November 2002 were studied. Patients with patent foramen ovale were excluded. The clinical data, echocardiography, cardiac catheterization and surgical reports were reviewed. Patients with only one visit (198) were excluded from further analysis. The rest (314) were classified into 4 groups: Surgical Closure (51), Device Closure (31), Spontaneous closure (148) and Persistent ASD on follow-up (84). The rates of closure were estimated using the competing risks methodology. Prognostic factors were considered in the modeling of spontaneous closure using cause-specific Cox regression analysis.
Results: Patients with one visit did not differ materially ($p > 0.05$) in terms of demographic and clinical characteristics from those who were analysed. The median duration of follow-up of the 314 patients was 40.6 months. The cumulative incidence of closure (95% confidence interval) was 23% (16% to 30%), 18% (8% to 28%), and 59% (52% to 65%) for surgical closure, device closure and spontaneous closure, respectively. Two most important prognostic factors influencing spontaneous closure were ASD size (HR = 0.76, 95% CI = 0.68–0.86, $p < 0.001$) and age at diagnosis (HR = 0.91, 95% CI = 0.86–0.96, $p < 0.01$).
Conclusion: In the current era of early diagnosis of ASD by easily available echocardiography service, high spontaneous closure rate in patients diagnosed in young age, especially in those with smaller defects, is to be expected. Device closure has occurred almost as frequently as surgical closure, reflecting the current trend towards the acceptance of this relatively new technique.

P189
Diagnosis of atrial septal defect: the value of "Crochetage" sign

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Aim: The ECG "Crochetage" sign (CS) has been found useful in identification of large ASD (Heller et al 1996). The purpose of this study is to determine the prevalence of CS in children and adults with isolated secundum atrial septal defects (ASDs) and to determine its value in the diagnosis of haemodynamically significant and small ASD.
Methods: 12-lead ECGs of 120 subjects (mean + SD age: 7.57 + 12.27 years), comprising 62 haemodynamically significant ASD (34 had surgical closure & 28 had device closure subsequently), 58 small ASD and 120 age- & sex-matched normal controls were studied carefully to identify the presence of CS in the inferior leads (II, III & aVF). Sensitivity, specificity and positive and negative predictive values (PPV & NPV) were computed. Results: The prevalence of CS in at least 1 lead was 64.7%, 39.3%, 34.5% & 10.8% for the 4 subgroups, respectively ($p < 0.01$ for all 3 ASD groups compared to controls). The prevalence of CS was also significantly higher in the Surgical Group compared to the Device Group ($p < 0.05$) and Small ASD Group ($p < 0.01$). The PPV varied from 0.75 to 1.00, while the NPV varied from 0.51 to 0.65. There was no significant difference between the predictive values for haemodynamically significant ASD and those for small ASD ($p > 0.05$).
Conclusion: CS is fairly common in ASD, especially in larger defects. PPV is better than 0.75 if present in 1 lead with PPD approaching 1.00 if present in 3 leads. The NPV is only marginally better then 0.50; dependence solely on this sign will miss about half of the patients.

P190
Natriuretic peptide type B is correlated with clinical outcome, hemodynamic load and systemic ventricular function in children with congenital heart defects and cardiomyopathies

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Introduction: Natriuretic peptide type B (BNP) which is produced mainly in the ventricles of the heart, has been shown to be increased in plasma (P) during systolic dysfunction and different types of hemodynamic overload of the heart. However, the clinical use of analysis of P-BNP in these conditions is still not clarified.
Aim: To study if P-BNP reflect clinical outcome and hemodynamic load of the heart in children with congenital heart defects and cardiomyopathies.
Methods: Consecutive blood samples for analysis of P-BNP during medical check-up, regular pre-operative investigation or intervention (surgery/catheter) in children with congenital heart defects or cardiomyopathy. The children were classified as having clinical symptoms of heart failure (CSHF) or not and hemodynamic load of the heart was evaluated by echo-doppler investigation and/or catheterisation. Possible hemodynamic overload of the heart was classified as: No overload (not sufficient to indicate surgery/catheter intervention according to local praxis), Pressure overload of right and/or left ventricle, Volume overload of right and/or left ventricle and Systolic ventricle dysfunction.
Results: Blood samples were obtained from 137 patients (76 boys, 61 girls), median age 2.9 years (3 months–16.7 years), with a median follow-up time of 244 days (7–1149 days). Twenty-four children had P-BNP > 100 ng/L of which 5 (22%) died or underwent heart transplantation due to end stage heart failure (ESHF) as compared to 2/113 (2%) children with P-BNP < 100 ng/L who died of other reasons than ESHF ($p = 0.01$). Children with CSHF

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had higher P-BNP values, median 141.5 ng/L (18.9–3910) (n = 22) than those without, median 18.2 (0.2–1400) (n = 115) (p = 0.02). Children with No overload had lower P-BNP, median 10.3 ng/L (0.2–28.1) than those with Pressure overload, median 17.9 (0.7–315) who had lower values than those with Volume overload, median 29.8 (5.5–352) (p = 0.002) which in turn had lower values than those with Systolic ventricle dysfunction, median 613 (81.8–3910) (p < 0.0001).
Conclusion: Clinical signs of heart failure, poor clinical outcome and hemodynamic overload of the heart is associated with increased P-BNP levels in children with congenital heart defects and cardiomyopathies.

P191 (see Abstract 99)
Accelerated transcatheter patch occlusion of large patent ductus arteriosus
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P192
Left atrial appendage and patent foramen ovale transcatheter patch occlusion in piglets: role of accelerated fibrin formation
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Fibrin formation has been found responsible for transcatheter patch (TP) adherence on the atrial septum, in experimental atrial septal defects (ASDs). Fully heparinized animals, required 48 hours of balloon support for the ASD occlusion. The timing corresponded well with the clinical experience. Accelerated fibrin formation could shorten polyurethane adherence time and hospitalization in patients implanted with the TP for different applications, including patent foramen ovale (PFO) and left atrial appendage (LAA) occlusions. The occlusion method involves crossing the PFO with a multipurpose catheter, placing an 11 F long sheath either in the left atrium or the LAA, inflating the balloon/patch to 12 mm and obstructing the PFO or the LAA. LAA and PFO TP occlusions were performed in 10 piglets (5 animals for each application). Preformed clot from the same animal was applied on the patch carrying balloon; no heparin was used. Single balloon patch with a proximal nitinol floppy wire was used for the PFO; no proximal wire was required for the LAA occlusion. The patch was released 8 hours later, under fluoroscopy and echocardiography. All patches remained in place after release, occluding either the LAA or the PFO. The animals were followed for periods up to 2 weeks, when they were sacrificed. Autopsy was performed in all animals. The patch remained in place and was endothelialized. We believe that the accelerated patch adherence for these lesions is partly due to their tubular anatomy and their larger attachment surface. In conclusion, accelerated fibrin formation can shorten the required polyurethane patch adherence time, for LAA and PFO occlusions to 8 hours. We speculate that similar timing should be expected for the corresponding clinical applications.

P193
Chronic endothelin-A receptor blockade lowers pulmonary vascular resistance of lambs with increased pulmonary blood flow
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Background: Endothelin-1 has been implicated in the pathophysiology of pulmonary hypertensive disorders. Endothelin receptor antagonists have recently demonstrated efficacy for the treatment of advanced pulmonary hypertension. However, the hemodynamic effect of early treatment on the vasculature of patients with increased pulmonary blood flow secondary to congenital heart disease is unknown. Therefore the objective of this study was to determine the hemodynamic effect of chronic ET-A receptor blockade in a lamb model of increased pulmonary blood flow following in-utero placement of an aortopulmonary shunt.
Methods: Immediately after spontaneous birth, the lambs were treated lifelong with either an ET-A receptor antagonist (PD 156707, 150 mg/kg) or placebo. At 4 weeks of age, the general hemodynamics were obtained and compared.
Results: The pulmonary vascular resistance was significantly lower in the treatment group (n = 6) than in the placebo group (n = 8) (76.5 ± 34.1 vs. 118.1 ± 32.0, p < 0.05). Mean right atrial pressure and systemic pulse pressure were also significantly lower in the treatment group than in the placebo group (4.0 ± 1.2 vs. 5.4 ± 0.8, p < 0.05 and 55.0 ± 17.3 vs. 93.1 ± 21.8, p < 0.05). 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).
Conclusions: Chronic endothelin-A receptor blockade lowers pulmonary vascular resistance in lambs with increased pulmonary blood flow and pressure, and has minimal effects on systemic hemodynamics and heart function. ET receptor blockade may be a therapeutic option for patients with congenital heart disease and increased pulmonary blood flow in which surgical options must be delayed.

P194 (see Abstract 106)
Postoperative course and growth after extracardiac Fontan operation in children under 4 years of age
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P195
Cardiac involvement in patients with Emery-Dreifuss muscle dystrophy
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Introduction: Emery-Dreifuss muscular dystrophy (EMD) is a rare either X-linked or autosomal dominant inherited neuromuscular disorder. Cardiac involvement is commonly seen and may consist of conduction disturbances (atrioventricular block, sinus node dysfunction,atrial paralysis or cardiomyopathy. Case reports: We report on three patients, two adolescents (15 and 16 years) with autosomal dominant EMD and a 20-year-old patient with X-linked EMD all showing cardiac involvement years after neurological onset of the disease. So far, the cardiac manifestations have been rhythm disturbances in all patients. Atypical left atrial flutter and atrial fibrillation occurred in two patients, in the other patient intermittend atrial fibrillation and ventricular tachycardias were recorded. For prophylactic reasons we implanted a cardioverter-defibrillator in all three patients. One female patient developed

severe dilated cardiomyopathy who had to undergo heart transplantation. One year after this procedure she is in good health.
Conclusion: Rhythm disturbances are the predominant feature of cardiac involvement in patients with EMD. In order to prevent sudden cardiac death due to malignant ventricular tachyarrhythmias the implantation of cardioverter-defibrillator is recommended. In patient with severe cardiomyopathy heart transplantation may be a successful therapeutic option.

P196
Dental causes of infective endocarditis in congenital heart diseases
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Hopital Cardiologique Louis Pradel

The aim of this retrospective study was to assess the incidence and outcome of dental infective endocarditis (IE) in congenital heart diseases (CHD). From 1966 to 2000, 153 IE occurred in 140 pts, aged 3.6 d to 78.5 y (median 10 y); 125 (76%) were <18 y of age and 42 were adults. A dental cause of IE was diagnosed in 40 cases (26%). Eight cases (20%) did receive appropriate prophylaxis for dental procedure. Non-compliance with protocols occurred in 8 cases (20%); 7 patients (17.5%) received inappropriate antibiotics given for unexplained prolonged fever, and 11 dental infections were latent (27.5%); in 6 cases, prophylaxis appliance could not be retrieved. Unoperated VSD (10 cases = 25%) and complex cyanotic CHD (10 cases = 25%) were the most frequent underlying CHD; 2 patients had Rastelli tube, 8 had native mitral (2 mitral prolaps) or aortic valvulopathies (20%) and 10 were miscellaneous CHD. Streptococcus was the commonest microbial agent (85% of the cases). Blood cultures were negative in 15%. Most of the cases presented with stable haemodynamical status; 88% had no cardiac failure. Early surgical (<2nd month after IE diagnosis) treatment was needed in 35% of the cases. Four patients died but only one death was related to IE. One-year, three-year and ten-year survival rates were respectively 100%, 96% and 92%. In summary, dental infections account for a significant proportion of IE in CHD, in particular in unoperated VSD and complex CHD; prophylaxis protocols and patients information and compliance should be improved.

P197 (see Abstract 91)
Changing patterns of infective endocarditis in congenital heart diseases over the past decade
D.F. Sylvie, S. Bertrand, S. François, B. André
Hopital Cardiologique Louis Pradel

P198
Prognosis of infective endocarditis in children and adults with congenital heart disease
D.F. Sylvie, S. Bertrand, S. François, B. André
Hopital Cardiologique Louis Pradel

The aim of this retrospective study was to to determine risk factors and long-term prognosis of infective endocarditis (IE) in congenital heart diseases (CHD). From 1966 to 2000, 153 IE occurred in 140 pts, aged 3.6 d to 78.5 y (median 10 y). There was no previous evidence of CHD in 16%; in 84%, underlying CHDs were: ventricular septal defect, isolated (15%: repaired 24%) or not (8%: repaired 60%), complex cardiopathy cyanotic (18%: palliated 77%) or acyanotic (7%: palliated 33%, unoperated 41%), Tetralogy of Fallot (8%: repaired 50%, palliated 36%), aortic (9% including bicuspidia 2.4% and prothesis 4.2%) or mitral (7.2% including MVP 2.4%, unoperated 100%) valvulopathy and persistant ductus arteriosus, atrial septal defect + mitral anomaly (3 = 2 repaired, 1 unoperated), 1 unoperated pulmonary valve stenosis. Route was unknown (31%), dental (25%), post cardiac surgery (19%), cutaneous (9.2%), ENT (6%) or other; 22% of the cases had prophylaxis, 12% did not; in 32.5% prophylaxis was not an indication and 12% received previous «blind» antibiotherapy. Blood cultures were negative in 14%; streptococcus (37%) and staphylococcus (32%, aureus 27%) were the commonest agents. Clinical cardiac complications occurred in 26.3%, embolias in 31%. Vegetations were the most frequent echographic lesion (42.4%). Risk factors for early surgery (31%) were clinical cardiac and embolic complications, echographic vegetations and mitral and/or aortic locations. Thirty-four patients died (20.7%), 1 day to 19.5 y after diagnosis (median 6 mths), from post-IE (48.5%), cardiac non-IE (39.4%) or unknown (12%) causes. Survival was 93.3%, 89%, 87.4%, 76% and 55.6%, at 1 mth, 6 mths, 1 y, 10 y and 25 y. Risk factors for bad prognosis were cardiac clinical complication (global heart failure, pulmonary oedema: $p = 0.004$, cardiogenic shock: $p = 0.0009$), previous palliation ($p = 0.04$), embolias ($p = 0.047$) and period < 1990 ($p = 0.007$); age, underlying CHD, route, bacterial agent, time to diagnosis, prophylaxis, negative culture, site of IE, echographic lesions, early surgery and previous IE did not influence mortality nor survival. Our results show that clinical complications and palliated CHD are risk factors for bad prognosis; mortality is significant but prophylaxis does not influence prognosis and

P199
Predictive value of acidosis on developmental outcome in newborns with congenital heart disease
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Objectives: This study is aimed at the evaluation of parameters for acidosis and their predictive value on developmental outcome in newborns with congenital heart disease.
Background: Attempts have been made to identify indices that may predict outcome or morbidity, as mortality for congenital heart disease has declined. Prenatal diagnosis has been shown to decrease pre-operative acidosis and might prevent the occurrence of disturbed developmental outcome.
Methods: 117 patients, requiring surgery for structural heart disease in the first 31 days of life were included. Diagnosis was established either pre- or postnatally. Pre-operative values of lactate, pH and Base Excess levels were compared to the occurrence of disturbed developmental outcome, i.e. an underperformance of more than 10% on a standardized Dutch developmental scale. Patients were divided into groups according to blood levels of parameters of acidosis, using Receiver Operating Characteristics curves for determining cut-off values for pH, Base excess and lactate.
Results: No significant difference in developmental outcome was found using values for pH or Base Excess as a cutoff level. Pre-operative lactate values exceeding 6.1 mmol/l resulted in a significant increase in impaired development: 40.9% as compared to 15.1% in infants with a pre-operative lactate lower than 6.1 mmol/l ($p = 0.03$).
Conclusions: Pre-operative lactate values have a prognostic value on developmental outcome in newborns with a congenital heart disease. The limited prognostic value of pH can be explained by the fact that pH can be easily corrected, while lactate better reflects the total oxygen debt experienced by these patients.

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P200
Congenital heart disease in pregnancies complicated by maternal diabetes mellitus: clinical insights into the developmental biology of the heart

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Objective: To investigate the incidence and distribution of congenital structural cardiac malformations among the offspring of type 1 diabetic mothers and the influence of preconceptional glycaemic control reflected by HbA1c-levels.
Design: A literature study and multicenter retrospective study.
Methods and Patients: The incidence and pattern of congenital heart disease in our study population and in the literature on the offspring of type-1 diabetic mothers, were compared with the incidence and spectrum of the various cardiovascular defects (CVD) in the offspring of non-diabetic mothers as registered by EUROCAT Northern Netherlands. Medical records were in addition reviewed for glycosylated haemoglobin (HbA1c) during first trimester.
Results: The distribution of congenital heart anomalies in our diabetic study population was in accordance with the distribution encountered in the literature. Approximately half the CVD were conotruncal anomalies as persistent truncus arteriosus (PTA), double outlet right ventricle (DORV)/tetralogy of Fallot (T4F) and transposition of the great arteries (TGA). In our series TGA was most frequently encountered and no DORV were detected. Our series show a significant elevated HbA1c values among mothers with infants with a CVD.
Conclusions: This study shows an increased incidence of transposition of the great arteries, persistent truncus arteriosus and double-outlet right ventricle/tetralogy of Fallot, suggesting a profound teratogenic effect at a very early stage in heart development during conotruncal septation. Our study emphasizes the frequency with which the offspring of IDDM-complicated pregnancies suffer from complex forms of congenital heart disease. Pregnancies with poor first trimester glycemic control are more prone to the presence of fetal heart disease.

P201
Persistent junctional reciprocating tachycardia in the fetus

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Background: Persistent junctional reciprocating tachycardia (PJRT) tends to be a persistent arrhythmia and requires more aggressive therapeutic management. Diagnosis and management of this infrequently occurring tachycardia in the fetus at an early stage is of importance for the prevention of congestive heart failure (CHF).
Methods: A retrospective study of four fetuses with supraventricular tachycardia (SVT) of the PJRT type was performed.
Results: All had sustained SVT (mean of 228 bpm) at a mean gestational age of 34 + 5 weeks, with CHF present in two. All four fetuses had postnatal confirmation of the diagnosis, three fetuses had prenatal characteristics of PJRT on M-mode echocardiography with a ventriculo-atrial (VA)/atrio-ventricular ratio > 1 on M-mode echocardiography suggesting a slow conducting accessory pathway, consistent with PJRT. Transplacental treatment with flecainide was effective in one patient, sotalol and/or digoxin was/were partially effective in the remaining three, two developed sinus rhythm, with short intermittent periods of tachycardia and decreasing signs of CHF, one case showed a minimal decrease in heart rate. Oral propranolol therapy converted two patients postnatally, in the remaining two patients radiofrequency ablation was performed at the age of 5 months and 6 years.
Conclusions: The characteristics of our prenatal PJRT cases include a sustained heart rate not exceeding 240 bpm with a long VA interval, presence of CHF and therapy resistance. Transplacental treatment should be initiated, possibly with a combination of sotalol and digoxin in nonhydropic cases, or flecainide, especially in case of fetal hydrops. Although propranolol is the drug of our choice postnatally, radiofrequency ablation seems indicated in therapy resistant cases with CHF, even in the first months of life.

P202
The sequential safety monitoring procedure as early indicator for deteriorating outcome in congenital heart surgery

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Background: To monitor the outcome of congenital heart surgery, techniques should be implemented to provide continues evaluation. This study assessed the use of the sequential safety monitoring procedure as early indicator for deteriorating outcome.
Methods: Mortality and morbidity rates in 3 periods: 1988–1990, 1991–1995 and 1996–1999 in which 2 different surgical teams operated were evaluated retrospectively. ICU-stay, delayed sternal closure and phrenic nerve paralysis were associated markers of outcome, often considered as near miss situations. Standard descriptive analysis and analysis by the sequential safety monitoring procedure were performed to identify trends. The sequential safety monitoring procedure is an online technique providing case by case information and clear-cut preset alert and alarm boundaries. In this study an overall mortality rate of 4% was considered satisfactory, an increase by a factor 1.5 or more unacceptable. Findings: In period 1 and 3 the same surgical team operated with respectively 5.8% and 4.4% mortality, overall 5.0% (1032/52). Mortality in period 2 was 9.9% (704/70), circumstantial delayed sternal closure and ICU-stay were doubled compared to period 1 and 3. Delayed sternal closure was associated with increased mortality and prolonged IC-stay, phrenic nerve paralysis was associated with prolonged ICU-duration. These retrospective data were evaluated in a prospective fashion, using the sequential safety monitoring procedure. The alert boundary would have been crossed in period 2 after 11 months (64 patients/mortality 7), the alarm boundary after 13 months (87 patients/mortality 11), indicating that the risk of mortality had increased by a factor 1.5 or more. In period 3 the cumulative points stayed well within both the alert and alarm boundary.
Conclusion: The sequential safety monitoring procedure provides accurate and early prospective detection of increasing mortality. It allows, even without requiring the use of near miss situations, timely intervention and prevention of further deterioration of surgical outcome.

P203
Exercise-testing for the detection of hypertension in adult post-coarctectomy patients

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Objective: Successfully operated aortic coarctation patients frequently show a marked systolic hypertensive blood pressure reaction at exercise. In view of this typical hemodynamic pattern, the question is warranted whether blood pressure in daily life is normal when office readings are low. We therefore determined whether exercise-testing in presumed normotensive adult patients after successful repair of aortic coarctation can predict hypertension at 24 hour blood pressure measurement.
Design and Methods: Seventy-three presumed normotensive, successfully repaired, adult post-coarctectomy patients (44 male; mean age 30, range 17–53 years; time after operation 22.9 years, SD 7.4) underwent a maximal, symptom limited treadmill exercise test (Bruce protocol). Blood pressure (BP) was measured every 3 minutes by conventional R.R.K. Daytime BP (7:00–23:00) was assessed by 24 hour ambulatory BP monitoring (Spacelabs 90207). Patients were considered hypertensive when day mean systolic BP = 140 mmHg. A ROC-curve was constructed using maximal systolic BP during exercise as test variable.
Results: Seventeen patients (23%) had daytime hypertension. Analysis of the ROC-curve (area 0.789) revealed that in our patients a cut-off value of maximal systolic BP during exercise =188 mmHg had the highest sensitivity (82%) and specificity (71%). Using day time BP = 135 mmHg as cut-off value, did not result in significant better test characteristics (area 0.806).
Conclusions: Hypertension is a common finding even in presumed normotensive and successfully corrected adult post-coarctectomy patients. Exercise-testing can predict hypertension in the absence of 24 hour ambulatory blood pressure measurement, however sensitivity and specificity are not very high. 24 hour ambulatory blood pressure measurement should be regularly performed for early detection of hypertension in these patients, who already are at high risk for cardiovascular complications.

P204 (see Abstract 98)
Comparison of surgical repair with balloon angioplasty for native coarctation in infants over 3 months of age
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P205 (see Abstract 122)
Prognostic value of balloon occlusion test (BOT) in cadidates for various types of cavo-pulmonary anastomosis
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P206
Increased carotid arterial wall thickness as an indicator of cardiovascular risk in adult patients succesfully operated for aortic coarctation
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Objectives: Despite succesful surgery of aortic coarctation cardiovascular mortality and morbidity, notably premature coronary artery disease and cerebrovascular accidents, are greatly increased in middle aged adult post-coarctectomy patients. This may be due to previous increased blood pressure in the pre-coarctatial arterial conduits. B-mode ultrasound imaging can describe status and changes in intima-media complex thickness (IMCT) of carotid and femoral arterial walls. IMTC is a non-invasive measure and considered a validated endpoint for atherosclerosis.
Methods: Ultrasound investigations were done in 26 normotensive (31.7(SD9.1)years) and 22 hypertensive (34.2 (9.6) years) adult post-coarctectomy patients, and in 26 age and sex matched controls (33.9 (9.9) years). Mean age at operation was 9.5(range 0.8–22.1) years in normotensives and 8.5(0.1–26.9) years in hypertensives. A subject's IMTC was calculated as the averaged measurements of three right and three left carotid arterial wall segments. Hypertension was defined as the daytime mean systolic blood pressure = 140 mmHg registered on 24 hour ambulatory blood pressure monitoring. Student's unpaired t-test was used to compare IMTC's of normotensive and hypertensive post-coarctectomy patients and controls.
Results: IMTC of the entire post-coarctectomy group (0.74 (SD0.10) mm) was increased if compared to controls (IMTC = 0.60 (0.09) mm):DIMT = 0.14 mm, $p < 0.0001$. Differences between normotensive (0.70(0.10) mm) and hypertensive (0.78(0.10) mm) patients and each of these groups with controls, were highly significant (all $p < 0.0001$)(See graph).
Conclusions: Normotensive as well as hypertensive post-coarctectomy patients showed increased carotid IMTC at relatively young age. The findings reflect increased vascular risk in all post-coarctectomy patients regardless of bloodpressure and indicate the need for cardiovascular risk profile evaluation in all patients to improve vascular disease prevention.

P207
Postinfarct and congenital VSD closure with Amplatzer occluders
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Purpose: Results of transcatheter closure of congenital and post-infarct ventricular septal defect (CVSD & PIVSD) with Amplatzer atrial septal occluder (ASO) and Amplatzer muscular VSD occluder (VSO) are presented. The procedure was performed in 11 patients (pts) in one center.
Material and Methods: PIVSD: Procedures were intended in 8 pts, from 2 to 16 (mean 10) weeks after myocardial infarction (MI). Mean age of pts was 62 (ranged from 51 to 76) years (y). One pt had two VSDs (residual post surgery). All pts were in NYHA III/IV class. Transjugular only venous approach was used in 6 pts, transfemoral, after a-v loop creation -in 1 pt. In 1 pt PIVSD could not be entered neither from the venous nor arterial side. CVSD: Two out of 3 pts (9 and 11 y) had perimembranous defects (PVSD) 4 mm in diameter with extension to muscular outflow tract. The third, 2 y old girl after previous surgical debanding and closure of PVSD had a 5 mm midmuscular defect. VSO were implanted through the femoral vein approach in all children.
Results: The procedure was successful in all pts but one, in whom (2 weeks after MI) 24 mm ASO could not achieve a stable position. In one pt with double PIVSD – 2 ASO were used. The size of 7 implanted ASO ranged from 10 to 20 (mean 17) mm and 4 VSO – from 6 to 12 (mean 8) mm. Mean fluoroscopy time was 62 (18–120) min. Two PIVSD pts died during the first week after the procedure because of multiorgan failure. Clinical improvement was observed in all remaining pts. Small residual shunt was diagnosed in 4 PIVSD, while complete closure – in 5 pts. No further complication occurred.

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Conclusions: Despite some technical problems implantation of Amplatzer devices is an attractive option of treatment for patients with congenital and postinfarct VSD's. In the latter – the procedure should be performed – if possible- after postinfarction scar formation.

P208

Histopathological evaluation on children with tachyarrhythmias using endomyocardial biopsy – comparison between radiofrequency catheter ablation and pharmacological therapy

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Purpose: To investigate the clinicopathological relationship between the pediatric patients treated by radiofrequency catheter ablation (RFCA) and those by antiarrhythmic drugs using endomyocardial biopsy (EMB).
Patients and Method: They included 25 patients treated by RFCA: 20 with paroxysmal supraventricular tachycardia (PSVT) and 5 patients with ventricular tachycardia (VT); 27 patients treated by drugs: 13 with PSVT, 14 with VT. Ages ranged from 6 years to 12 years. Following the routine cardiac evaluation and electrophysiological study were performed, EMB was obtained all from the apex of the right ventricle. Histopathology was evaluated with histomorphometric method to calculate average diameter of myocytes, the percent area of fibrous tissue and fatty tissue and small vessel changes using computer assistance.
Results: Myocardial changes of SVT vs. VT on EMB, such as fibrosis, degeneration, disarray and inflammatory cell infiltration, showed 55% vs. 50%, 55% vs. 80%, 75% vs. 65% and 40% vs. 75%, respectively in the patients treated by RFCA, while 62% vs. 50%, 54% vs. 50%, 46% vs 21% and 38% vs. 21%, respectively in the patients treated by drugs. In some cases of the patients with SVT, we found histopathology that suggested postmyocarditic state and hypertrophic cardiomyopathy.
Conclusion: Myocardial histology in the patients both with SVT and VT showed various abnormalities on light microscope as well as on ultrastractural characteristics. It is suggested that some pediatric cases could develop tachyarryhtmias triggered by myocarditis. Persistent myocarditis could be the cause of left ventricular dysfunction without complete suppression of tachycardia, leading to tachycardia-induced cardiomyopathy. On the basis of our findings with EMB, RFCA should be recommended because of rational therapy.

P209

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 investigate autonomic nervous system tone and to estimate the relation between frequency domain HRV parameters and the prevalence of ventricular arrhythmias (VA) in children with mitral valve prolapse (MVP).
Methods: 151 children with MVP and 164 healthy subjects underwent a clinical examination, 24-hour ECG monitoring and echocardiography. 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: VA were recorded in 42% children with MVP. 29 (19%) of the MVP patients manifested infrequent ventricular premature complexes (VPCs), 12 (8%) frequent VPCs, 13 (9%) multiform VPCs, 6 (4%) couplets of VPCs and 3 (2%) had runs of nonsustained ventricular tachycardia. 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: $p < 0.000005$, daytime: $p < 0.009$) values and higher LF/HF ratio (night: $p < 0.000006$; daytime: $p < 0.0002$) than in controls. Children with MVP and VA 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. There was significant relation between the degree of VA in MVP patients and the value of HFNU (daytime, $p < 0.05$) and LF n.u. (night and daytime, $p < 0.05$) parameters.
Conclusions: Our data suggest alterations in sympathovagal balance with reduced vagal and increased sympathetic activity during night and daytime in children with MVP. Alteration in sympathovagal balance with sympathetic predominance is associated with increased prevalence of VA and the higher degree of VA according to the Lown classification in these patients.

P210

Relation of cerebral tissue oxygenation index to central venous oxygen saturation in children

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Objective: Evaluation of the relationship between the cerebral tissue oxygenation index (TOI) measured by near-infrared spectroscopy (NIRS) and central venous oxygen saturation (SvO_2) after corrective surgery of non-cyanotic congenital heart defects in children.
Design: Prospective observational clinical study.
Setting: An intensive care unit for paediatric cardiology.
Patients: Infants and children consecutively admitted to the intensive care unit after corrective surgery of non-cyanotic congenital heart defects.
Methods: NIRS provides a continuous, non-invasive method to measure regional changes in tissue oxygenation. Spatially resolved spectroscopy as an algorithm allows the calculation of the TOI which expresses the ratio between oxygenated haemoglobin (O_2Hb) and total haemoglobin content (tHb) (TOI (%) = (O_2Hb/tHb) × 100) in the observed tissue. Cerebral TOI was compared to SvO_2 taken from a central venous catheter placed in the right atrium and to haemodynamic and respiratory parameters. Pearson`s correlation coefficients and p-values were calculated. Bland-Altman analysis was used to study the agreement between cerebral TOI and SvO_2.
Results: Forty-three children were studied (median age: 2.8 years, range: 7 days to 16.8 years). Simultaneously measured values for SvO_2 (62.2 ± 9.8%, 39.8 – 80.4%) and cerebral TOI (56.7 ± 8.8%, 35.8 – 71.2%) showed a significant correlation ($r = 0.52, p < 0.001$). Bland-Altman analysis showed a mean bias of −5.4% with limits of agreement between 12.8% and −23.7%.

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Conclusion: Cerebral TOI and SvO_2 are not interchangeable parameters, but cerebral TOI may reflect the haemodynamic influence on cerebral oxygenation after cardiovascular surgery. Further work is necessary to confirm the clinical role of continuous non-invasive measurement of cerebral TOI after cardiac surgery in children.

P211 (see Abstract 125)
Myocardial performance, baroreceptor reflexes and coronary flows evaluation using the tilt table in preterm and term neonates

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P212
Evaluation of the diastolic function of the left ventricle before and after ASD closure with an Amplatzer septal occluder (ASO)

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Introduction: It has been often discussed that the implantation of an occlusion device into an atrial septal defect might influence the elasticity of the spetum and therefore the atrial function with an consecutive impact on the ventricular function. The goal of our study was to evaluate the diastolic function of the left ventricle before and after percutaneous closure of an ASD with an Amplatzer Septal Occluder to determine if there is a change of cardiac performance due to device implantation.
Method: In 16 consecutive patients with an ASD with significant left-to-right the diastolic filling patterns of the left ventricle were evaluated with Doppler echocardiography before and after transcatheter closure with an ASO. The following parameters were measured the day before and the day after the intervention: The mitral valve (MV) inflow velocity with the duration of the e wave and the a wave, the pulmonary venous backward flow velocity (PV), the isovolumetric relaxation time (IVRT) and the wall Tissue Doppler (TDI).
Results: The median age at the time of the closure was 7.1 years-old (3.5–15), the median diameter of the ASO was 17 mm (10–22 mm). The statistical analysis was made with paired student's T-test (cf. Table 1: data are expressed in mean ± standard deviation). No significant difference was found in the left ventricular diastolic filling function before and after percutaneous closure of the ASD.

	Before	After	
MV e wave (cm/s)	90 ± 16	97 ± 18	p = NS
MV a wave (cm/s)	53 ± 10	50 ± 16	p = NS
MV e/a	1.75 ± 0.35	2 ± 0.45	p = NS
MV e duration (ms)	185 ± 28	184 ± 42	p = NS
MV a duration (ms)	97 ± 16	103 ± 22	p = NS
PV (cm/s)	26 ± 4	26 ± 4	p = NS
IVRT (ms)	44 ± 7	43 ± 7	p = NS
TDI Ea wave (cm/s)	17 ± 3	16 ± 2	p = NS
TDI Aa wave (cm/s)	9 ± 2	9 ± 1	p = NS
TDI Ea/Aa	1.95 ± 0.45	1.85 ± 0.25	p = NS

NS = not significant

Conclusion: In these 16 consecutive patients the diastolic function of the left ventricle before intervention was normal. The measure of the filling patterns of the left ventricle after percutaneous closure remained unchanged, suggesting that transcatheter closure of an ASD does not influence the diastolic filling of the ventricles, even with devices with marked device mass like in the ASO constructed of a solid metal wire-meshwork.

P213
Chronic Blalock-Taussig shunts in piglets: a model of pulmonary vascular disease by high pulmonary blood flow

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Objective: The purpose of this study was to establish an animal model of high pulmonary blood flow and pulmonary vascular disease.
Methods: Blalock-Taussig shunts were constructed in 5-week-old piglets (n = 6; sham-operated controls, n = 7). Intraoperative measurements of haemodynamics and shunt flow were performed, with a cardiac catheterization after a follow-up period of 3 months. Plasma levels of cGMP and endothelin-1 were measured, and histomorphometry was performed on heart and lung specimens.
Results: Shunt flow was 930 [380–1300] ml/min (median [range]). All animals survived the follow-up period. Shunted animals showed clinical signs of severe congestive heart failure, necessitating a diuretic treatment for about 1 week until recovery. At cardiac catheterization the shunt was patent in all cases. Shunted animals had a significantly higher pulmonary artery pressure (18 [17–28] mmHg, sham group, 15 [11–19] mmHg, $p < 0.05$). Qp/Qs was 1.8 [1.5–2.9]. Circulating levels of cGMP and endothelin-1 were significantly elevated in shunted animals ($p = 0.005$/$p = 0.01$). Histomorphometry revealed dilatation of the intraacinar pulmonary arteries and medial hypertrophy in the shunt group. The ratio of heart weight to body weight was significantly higher in the shunted animals (7.5 [5–7.8] g/kg body weight, sham group, 6.4 [3.7–7.8] g/kg body weight, $p < 0.05$), due to a marked biventricular hypertrophy.
Conclusions: We observed significant haemodynamic, metabolic, and histopathological changes after the creation of chronic Blalock-Taussig shunts in piglets, representing a reliable animal model of pulmonary vascular disease.

P214
Increased stifness of the aorta in tobacco smoking adolescents

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Arterial stiffness may be an indicator of early vascular changes signaling the development of vascular disease, while tobacco is a well-recognized promoter of atherogenesis. The purpose of this study is to measure the aortic stiffness in tobacco smoking adolescents and to investigate the relationship between tobacco smoke and cardiovascular disorders especially atherosclerosis in adolescent ages. Aortic strain (S), pressure strain elastic modulus (Ep) and normalized Ep (Ep*) in tobacco smoking adolescents and the healthy control

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group, were measured by a sphygmomanometer with cuff and transthoracic echocardiography. The study group consisted of 30 healthy cases (M/F: 27/3) as a control group and 30 tobacco-smoking adolescents (M/F: 28/2). The mean ages were 16,1 ± 1.8 and 16,2 ± 1.4 years respectively. The number of cigarettes smoked per day was 31 ± 7.1 and the duration of smoking were 3,1 ± 1.1 years. S, Ep and Ep* measurements were 0.198 ± 0.042, 215 ± 16.7 and 2.84 ± 0.42 in tobacco smoking adolescents and 0.262 ± 0.056, 152 ± 17.8 and 2.2 ± 0.66 in normal control respectively. S, Ep and Ep* measurements were statistically significant in tobacco user adolescents compared to the control group. HDL levels were lower in this group than the control group but statistically insignificant. These parameters suggest that this group has cardiovascular risk factors in adolescent ages. These findings may be important to determine the relationships among cardiovascular risk factors especially tobacco at the adolescent-adult transition.

P215

Prenatal evaluation of the pulmonary veins in the fetuses with heterotaxy syndrome

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For the fetuses with heterotaxy syndrome, the anatomy of the pulmonary veins is important information for estimating the risk for postnatal outcome. Although recent development of echocardiographic equipment allows us to assess detailed anatomy from prenatal period, the prenatal assessment of the complex pulmonary venous anatomy is still difficult. Furthermore, prenatal detection of the pulmonary venous obstruction has not been established. Hence, we examined the accuracy of the prenatal diagnosis of the pulmonary venous anatomy. We also compared the Doppler flow patterns at the pulmonary veins in prenatal period and those in the postnatal one. From 1999 to 2002, 6 fetuses with heterotaxy (5 with right atrial isomerism and 1 with left atrial isomerism) were detected prenatally, and had postnatal confirmation by echocardiography and/or cardiac catheterization. Detailed visualization of the pulmonary veins was possible in 5 cases. However, in one case with complex pulmonary arteries' anatomy and multiple collateral arteries, partial anomalous pulmonary venous return (the left upper pulmonary vein to the innominate vein) had not been diagnosed prenatally. By the Doppler study in prenatal echocardiography, pulmonary venous obstruction had been suspected in 2 fetuses. In the first case with common chamber of the pulmonary veins returned to the superior vena-cava, prenatal Doppler study detected increased blood velocity (1.4 m/sec) with continuous flow pattern at the junction between the common pulmonary vein to the superior vena-cave. However, postnatal study showed that the obstructive site was between the superior vena-cava and the right atrium, and the pressure gradient of only 10 mmHg. In the other case, Doppler study detected abnormal continuous flow pattern but normal blood velocity (45 cm/sec) at the site between the common pulmonary vein to the atrium. Postnatal echocardiography showed no significant pulmonary venous obstruction, even after the postnatal increase of pulmonary blood flow. In conclusion, prenatal echocardiography can accurately detect the pulmonary venous anatomy. However, the detection of the partial anomalous pulmonary venous return seemed to be difficult in some fetuses with complex anatomy. Abnormal pulmonary venous blood flow pattern may not directly relate to the postnatal obstruction, so that prediction of postnatal pulmonary venous obstruction is difficult.

P216

Identification of potential arrhythmogenic conditions by neonatal EKG screening

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Background: In 1998, Schwartz et al demonstrated that "a prolongation of QT interval in the neonatal period was strongly associated with SIDS". We submitted to EKG "screening" a population of neonates, to verify: 1) prevalence of QTc interval prolongation; 2) prevalence of the long QT syndrome; and 3) prevalence of other arrhythmogenic conditions.
Methods: ECGs were recorded on the 3rd or 4th day in 8808 neonates born between January 1996 and November 2002. All infants were healthy and born at full term. Neonates hospitalised at NICU were enrolled, excluding very premature and sick newborns. EKG: QT intervals were usually measured in D2, and corrected for heart rate using Bazett's formula. Neonates with QTc > 440 msec underwent additional ECGs on the 3rd week of life. A Holter monitoring was performed on the 4th week of life in babies with QTc prolongation in order to: (a) evaluate 24-hours QTc variability; and (b) institute preventive measures. Results. Of the 8808 infants, a QTc interval > 440 msec was found in 156 (= 1.8%). 9 of these, showing a QTc interval prolongation at birth (>460 msec) confirmed in the following weeks, received therapy with Propranolol. During a 2.5 years follow-up (4.y to 10 m.) QTc returned within normal values in 6 infants who discontinued beta-blockers therapy at the end of 1st year. In 3 babies the QTc interval persisted prolonged (490,500 and 590 msec respectively) and therapy was not stopped. Molecular analysis was negative in two. In one of these infants, family history of LQTS was present. The results are still awaited for the third. A Wolf-Parkinson-White (WPW) syndrome (= δ wave) was present in 5 babies (symptomatic in one). Epidemiological remarks. We have registered the following data: (1) prevalence of a persistent QTc interval prolongation in the first month of life = 9/8808; (2) prevalence of LQTS = 3/8808; (3) prevalence of WPW syndrome = 5/8808. Due to the small sample size caution shall be use in the interpretation of these results. A larger ongoing multicentric study will contribute to assess the prevalence of these ECG abnormalities.

P217

Long-term cardiac follow up of severe twin-to-twin transfusion syndrome

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Introduction: In twin-to-twin transfusion syndrome (TTTS) vascular placental anastomoses lead to unbalanced shunting of blood from one twin to the monozygotic co-twin. The hypervolemic recipient develops myocardial hypertrophy and congestive heart failure. In addition, congenital heart disease of the right ventricle and pulmonary artery are reported. In contrast, the hypovolemic donor rarely develops heart disease.

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Objective: To evaluate long term effects on cardiac structure and function in genetically identical twins with a different prenatal hemodynamic and environmental situation.
Patients and Methods: Prospective follow up study of 82/89 survivors of 73 consecutive twin pregnancies with severe TTTS treated by laser coagulation of placental anastomoses. Pre- and postinterventional fetal echocardiography and study of feto-placental hemodynamics were performed at a median gestation age of 21.7 weeks and postnatal cardiac follow up including echocardiographic study of systolic and diastolic myocardial function at a median age of 21.1 months.
Results: In spite of a high rate of prenatal cardiac dysfunction (27/47 recipients) or pathological Doppler findings (8/35 donors) the majority of twins showed no signs of even minor cardiac involvement at follow up. Pulmonary stenosis was only recorded in recipients (3/47 = 3.6%; CI 0.76–10.3%, bimodal distribution) and more common than in the general population (prevalence 0.378‰). Dilatation of severe pulmonary stenosis had to be performed in 2/3 cases. Other forms of congenital heart disease were equally distributed between the donor and recipient cohort.
Conclusion: This study illustrates the remarkable capacity of the heart for ventricular remodeling once causal factors are removed. On the other hand it supports the hypothesis, that alterations in fetal hemodynamics may result in some form of structural heart disease. Given the progression of pulmonary stenosis either prenatally or after birth, intrauterine and postnatal follow up of these children is warranted for early intervention.

P218

Transcatheter closure of atrial septal defect in young children: results and follow up

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Objectives: This study sought to analyze safety, efficacy and follow-up results of percutaneous closure of secundum atrial septal defect (ASD) in young children.
Background: Results of ASD transcatheter closure in adults are widely reported but there are no large published series concerning young children.
Methods: Between December 1996 and September 2002, out of 590 patients treated at our institution 56 were children aged less than 5 years old. Indications for closure were: elective closure in 38 patients, frequent respiratory infections in 9, failure to thrive in 2, liver transplantation in 6 and fenestrated Fontan in 1. The procedure was carried out under general anesthesia with fluoroscopy and transesophageal control. Two different devices were used: the CardioSEAL/StarFLEX (CS/SF) and the Amplatzer Septal Occluder (ASO). Basal physical examinations and echocardiograms were performed prior to the procedure and at follow-ups (1,6, and 12 months and yearly thereafter).
Results: The mean age at closure was 3.6 ± 1.3 years. CS/SF was used in 10 subjects while ASO was used in 46 patients. No deaths or any major complications occurred. the total occlusion rate was 87% at procedure rising to 94% at discharge. The mean follow-up was 18 ± 14 months. No major or minor complications occurred. The occlusion rate rose to 100% at 12 months of follow-up. Symptomatic patients improved significantly.
Conclusions: In the current era and in experienced hands, ASD closure can be performed safely and successfully even in very young children.

P219

Transcatheter closure of the patent ductus arteriosus with the Amplatzer duct occluder in very young symptomatic infants

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Background: Transcatheter occlusion of Patent ductus arteriosus (PDA) using various occluders and coils is a well established alternative to surgical duct ligation. The use of Amplatzer duct occluder (ADO) is reported in children and adults while there are only occasional reports in symptomatic infants and very young children.
Aim: To report the immediate and short term results using the ADO to close moderate to large sized PDAs in a pediatric population of very young symptomatic children.
Patients and Methods: From March 2000 to September 2002, 13 out of 85 subjects undergoing PDA percutaneous closure at our institution were very young symptomatic children. Age at intervention ranged from 6 months to 3 years. Patients with clinical and echocardiographic findings of moderate to large PDA and weighing > 4500 gs were included in the study. In all subjects ADO was used. Prophylactic antibiotics with cephalosporin were administered during the procedure. All patients were discharged 24 h after the procedure on no medication.
Results: Mean age and mean weight at procedure were respectively 19 ± 11 months and 9.3 ± 3.5 kgs. Indications for closure were: failure to thrive in 7 patients, frequent respiratory infections in 6 pts. Median PDA diameter at its narrowest point determined by angiography was 4 mm. Delivery of the device was successful in all patients. In 5 patients there was a trivial residual shunt immediately after the procedure. Mean procedure and fluoroscopy time were 76 ± 8 minutes and 9 ± 2 minutes, respectively. The following ADO devices were used: 6 × 4 mm: 8 patients; 8 × 6 mm: 4 patients; 10 × 8 mm: 1 patient. No complication occurred. All patients were followed up for an average of 16 ± 7 months. Patients with recurrent respiratory infections had no significant recurrences and subjects with failure to thrive increased significantly their growth. The absence of obstruction of the left pulmonary artery and of the descending aorta was confirmed during the follow-up. No other complication occurred.
Conclusion: Even in very young children percutaneous closure of moderate to large PDA can be performed safely and successfully.

P220

Electrophysiological study in patients after repair of tetralogy of Fallot: correlation with late potentials

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We compared signal-averaged electrocardiography with electrophysiological study in patients after correction of tetralogy of Fallot to determine if potentially correlations exist between the two methods for assessment of risk for ventricular tachycardia (VT). The study group consisted of 30 patients with correction of tetralogy of Fallot with the mean age of 16.5 ± 3.9 years (mean age at surgical correction was 4.6 ± 3.2 years). The mean duration of follow-up was 11.9 ± 1.8 years (ranged, 9 to 15 years) after corrective surgery. Holter recordings, signal averaged ECG, and electrophysiological study was performed. Signal averaged electrocardiograms were examined for the duration of low amplitude QRS signal (LAS), duration of total QRS, and root-mean-square voltage of the

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terminal 40 msec of the QRS (RMS-40). During electrophysiological study; single, double, and triple programmed ventricular premature stimuli were introduced into ventricular paced rhythm in two areas (apex and outflow tract) of right ventricle during basal state and isoproterenol infusion. Patients were classified by electrophysiological study results as having no inducible VT, non-sustained VT or sustained VT. All patients had right bundle branch block, with a mean QRS duration of 148 ± 30 msec (ranged, 85 to 224 msec). The mean LAS was 34.3 ± 37 msec (ranged, 4 to 135 msec) and the mean RMS-40 was 63.1 ± 47 μV (ranged, 4 to 169 μV). There were no patients with sustained VT. Non-sustained VT was detected in 5 patients (16.6%). Duration of low amplitude QRS signal = 35 msec yielded a very high predictive accuracy (sensitivity = 80% and specificity = 92%) for non-sustained VT inducible by electrophysiological study. Similar sensitivity but less specificity was seen using the criterion of total QRS duration = 135 msec and RMS-40 = 60 μV. In conclusion, probably due to the relatively short mean follow-up time after surgery, we could not found sustained VT in any patient. However, the presence of non-sustained VT indicates a specific patient group in whom close follow-up may be needed. Duration of low amplitude QRS signal = 35 msec is easily measured variables with a good sensitivity and specificity for identifying patients with non-sustained ventricular arrhythmias.

P221

Reactive oxygen species regulate the hypoxia-inducible transcription factor HIF and its target genes VEGF and PAI-1 in pulmonary artery smooth muscle cells

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Pulmonary hypertension is associated with increased levels of reactive oxygen species (ROS) and with hypoxia. Both stimuli lead to proliferation and thrombosis frequently observed with this disease. Hypoxia-inducible factors (HIF) regulate the transcription of vascular endothelial growth factor (VEGF) and plasminogen activator inhibitor-1 (PAI-1) under hypoxia due to enhanced stabilization of the HIF-alpha subunits. Activation of the HIF pathway is also observed under normoxia in response to thrombin but the mechanisms involved are not completely clear. Since ROS act as signaling molecules we investigated the role of ROS in the HIF pathway in pulmonary artery smooth muscle cells (PASMC). Stimulation with hypoxia, thrombin, and H_2O_2 enhanced HIF-alpha mRNA and protein levels and induced VEGF and PAI-1 expression. Treatment with the antioxidants vitamin C and N-acetylcysteine decreased these responses. Transfection of a p22phox antisense vector or a dominant negative Rac1 to inhibit the activity of the ROS-generating NADPH oxidase diminished HIF-alpha, VEGF and PAI-1 levels. These results show that activation of the HIF subunits is sensitive to ROS production in PASMC and suggest a role of this pathway in promoting thrombosis and proliferation in pulmonary hypertension.

P222 (see Abstract 114)

Tissue factor expression is promoted by thrombin via Rac and the p21-activated kinase PAK in pulmonary artery smooth muscle cells

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P223

The benefit of using triple-lumen catheter to monitor left atrial pressure

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Left atrial pressure (LAP) monitoring provides a useful option for management of hemodynamic status in pediatric open-heart surgical patients during the postoperative period. We used a triple-lumen catheter placed into the left atrium transseptally to measure left atrial pressure. Twenty children that were operated in our clinic were included for this study. There were 11 male and 9 female. A 5-Fr. triple-lumen radio-opaque polyethylene catheter was used for the procedure. After the repair of the primary cardiac defect, the distal end of catheter was repositioned transseptally and advanced into the left atrium. The proximal and middle line's distal orifices were left in the right atrium. Distal line was used as a left atrial pressure line while middle line as a central venous pressure line and proximal line as a route for fluid infusion or drugs administration. No mortality and no catheter related complication was observed. No failure or complication occurred during withdrawal of the catheter.

P224 (see Abstract 95)

Value of the novel isovolumetric acceleration index in the evaluation of ventricular systolic function in patients following corrective surgery of TOF

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P225

The effect of high dose melatonin on cardiac ischemia-reperfusion injury: an experimental study

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Melatonin, is the most potent scavenger of toxic free radicals, has been found to be effective in protecting against pathological states due to reactive oxygen species release. This study was performed in order to establish the effects of high dose of melatonin for protection against ischemia-reperfusion (I/R) injury in rats' heart. Forty male Sprague-Dawley rats were used in this study. They were separated in four groups of ten. Left coronary artery occlusion was induced in rats by ligating for 10 minutes followed thereafter by release of the ligation (reperfusion). Group A was control group. Group B was subjected to myocardial ischemia-reperfusion without any treatment. Group C was subjected to myocardial ischemia-reperfusion with melatonin treatment before ischemia. Group D was subjected to myocardial ischemia-reperfusion with melatonin treatment before reperfusion. After 20 minutes of reperfusion, blood samples were obtained for biochemical studies and animals were sacrificed for histological, immunohistochemical examination of myocardial tissue. We found that cTn-T levels were significantly increased in group B as compared with all groups. We showed that in Group C where rats were treated with melatonin before ischemia, cTn-T values were significantly lower than group B and D. In addition MDA and antioxidant enzymes, such as SOD and MPO in melatonin treated groups, were lower compared with group B. The differences were statistically significant ($p < 0.05$).

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Histopathologic and immunohistopathologic studies were also supporting the effectiveness of melatonin. Our study suggests that high dose melatonin, appears to confer protection against cardiac ischemia-reperfusion injury in rats by scavenging the free readicals and could have a potential clinical use in the management of myocardial ischemia.

P226
Cardiac structural and functional abnormalities in growth hormone deficient children before and six months after growth hormone replacement therapy

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This study included 45 patients with growth hormone (GH) deficiency, 30 males and 15 females with mean ages of 11.5 ± 4.6 years. Clinical evaluation included: resting heart rate, blood pressure, auxological data: height standard deviation (SD), height velocity SD, body mass index (BMI) and body surface area (BSA). Blood count, serum electrolytes, kidney function tests were also done. Echocardiography assessed left ventricular diastolic and systolic dimensions (LVEDD, LVESD), left ventricular fractional shortening (FS), ejection fraction (EF), left ventricular mass index (LVMI). Thirty patients received recombinant human GH (rhGH) therapy for six months (rhGH gp); other fifteen patients formed a control group. All patients were fully re-evaluated 6 months from the initial assessment. Initially, there were no significant differences between rhGH group and control one in auxological, laboratory and echocardiography data (all were within normal range for age and sex). GH therapy significantly improved the height SD, but not BMI, nor resting heart rate. Systolic blood pressure showed significant elevation by 3.6%with GH therapy. Although echocardiography data for both groups were within normal both initially and 6 months later, GH therapy resulted in significant increase in LVEDD (4.33 ± 0.41 vs. 3.46 ± 0.48 cm, $p < 0.05$), decrease in LVESD (2.48 ± 0.44 vs. 2.72 ± 0.28 cm, $p < 0.05$) and improvement in FS (42.36 ± 8.58% vs. 25.73 ± 7.2%, $p < 0.05$), and increase in LVMI (116.16 ± 40.47 vs. 86.54 ± 28.93 gm/m^2, $p < 0.05$).
Conclusion: Although children with GH deficiency have cardiac structure and function within normal ranges for their ages and sex, GH replacement therapy improves their cardiac function. However, long term studies are needed to guard against possible ventricular hypertrophy.

P227
Cleft mitral valve: characteristics and outcome

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Background: Few studies have addressed the morphologic features of cleft mitral valve and the outcome of the patients. The pathogenetic features including the developmental relation with atrioventricular septal defect remain unclear.
Methods and Results: Patients with atrioventricular canal, ventriculo-arterial discordance and hypoplastic ventricles were excluded. Twenty-two patients had a cleft on the anterior leaflet of the mitral valve identified by echocardiography at a median age of 0.5 years (0 to 10.6 years), with attachments of its components to the ventricular septum in 19 cases. Ten patients had a more than mild mitral regurgitation and left ventricular outflow tract obstruction by the cleft occurred in 3 cases. Associated cardiac lesions and extracardiac features were present in 13 and 10 patients, respectively. During the median follow-up period of 1.5 years (0 to 11.8 years), 2 patients died from extracardiac causes and one neonate from severe subaortic obstruction. Conservative mitral valve repair was performed in 10 patients at a median age of 5.2 years (1.3 to 10.6 years). One patient was reoperated on for mitral stenosis associated with aortic valve stenosis. Follow-up echocardiography demonstrated a moderate mitral regurgitation in 2 unoperated patients and a moderate mitral stenosis in 2 operated patients.
Conclusions: Cleft of the anterior mitral valve leaflet comprises a wide clinical spectrum with associated cardiac and extracardiac features. Important morphologic differences exist with atrioventricular septal defect although the 2 lesions may be related pathogenetically. Conservative surgical repair always seems possible. Long-term echocardiographic follow-up of the operated and unoperated patients is warranted.

P228
Consumption of blood products during long-term mechanical circulatory support in children: a comparison between two different perfusion systems

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Background: Different mechanical circulatory support (MCS) systems are used in children and even small infants with intractable heart failure. However, the need for anticoagulation leads to hemorrhage with subsequent use of blood products. We compared the coagulation disorders and the need for blood products in children treated either with ECMO or with the pulsatile pneumatic ventricular assist device, Berlin Heart.
Methods: We retrospectively reviewed the course of 64 children who were on MCS for more than 2 days during the course of 8 days between 1994 and July 2002 in our center. Thirty children with a median age of 7.4 [0.4–16.8] years, and median body weight of 25.5 kg were on Berlin Heart support, whereas 34 children with a median age of 1.8 [0.01–16.8] years, and median body weight of 9.2 kg were supported with ECMO. Anticoagulation was accomplished by continuous infusion of heparin in all cases. Target activated clotting time was 140–160 sec in the Berlin Heart and 160–180 sec in the ECMO group. Red blood cell count, platelet count, aPTT, AT III and Fibrinogen were measured twice daily. Depending on blood loss and the coagulation disorder red blood cells (when Hb < 9 g/dl), fresh frozen plasma (when Fibrinogen < 180 mg/dl) and platelets (when <70 000/ml) were substituted.
Results: There were no preoperative differences in haematologic parameters between the two groups. The mean daily substitution was significantly higher in the ECMO group. In the Berlin Heart group the platelet transfusion was 4.3 ml/kg/d vs. 24.6 ml/kg/d in the ECMO group. Red blood cell substitution in the Berlin Heart group was 17.2 vs. 60.3 ml/kg/d in the ECMO group. The substitution of fresh frozen plasma was 8.5 ml/kg/d in the Berlin Heart group and 46.9 ml/kg/d, in the ECMO patients. Nevertheless the mean daily values for hemoglobin, platelets and fibrinogen were lower in the ECMO group. There were fewer bleeding complications and lower overall mortality in the Berlin Heart group.
Conclusion: Compared to ECMO the use of the Berlin Heart in children results in less blood loss and lower consumption of blood products.

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P229

Simultaneous relief of right pulmonary artery stenosis and closure of residual Waterston shunt using a covered stent

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Introduction: Anastomosis of the ascending aorta to the right pulmonary artery (RPA), the so-called Waterston shunt, was undertaken as a palliative procedure for children with cyanotic congenital heart disease due to obstruction of the pulmonary outflow tract with reduced pulmonary blood flow. Many complications of this shunt have been documented, among others RPA stenosis, side different pulmonary perfusion, and endocarditis. Stent mounted endovascular grafts, so-called covered stents, have recently found wide spread use in aortic and periphereal angiologic interventional procedures. Their use in coronary diseases is under investigation. So far, there are only few reports about the use of covered stents in congenital heart disease and its sequelae.
Case Report: We present the case of a 35-years-old male adult, who had had a Waterston shunt for palliation of tetralogy of Fallot at the age of two years. Subsequently he underwent corrective surgery at the age of 12 years. Regulary, the Waterston shunt would have been closed at that time. Due to pulmonary regurgitiation and right ventricular failure a valved conduit was implanted at the age of 34 years. Unfortunately, the patent shunt and a concomitant RPA stenosis persisted. Both were eliminated simultaneously by implantation of a covered stent into the RPA. A Jomed Stentgraft 38 mm mounted on a 12 × 40 mm Cordis Powerflex balloon catheter was used. There were neither residual shunt nor residual RPA stenosis.
Conclusion: Implantation of a covered stent can simultaneously eliminate residual shunt and relief pulmonary artery stenosis in selected patients after repeated surgery for congenital heart disease.

P230

Doppler echocardiographic predictors of outcome in newborns with persistent pulmonary hypertension

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Background: The echocardiographic investigation of newborns with persistent pulmonary hypertension has been limited to the assessment of pulmonary blood flow and pressure. Features of intracardiac shunting and echocardiographic predictors of outcome are not well described.
Objectives: To review in newborns with persistent pulmonary hypertension echocardiographic characteristics of shunting and ventricular performance, and to identify the predictors of ECMO therapy, death and response to inhaled nitric oxide.
Patients and Methods: We retrospectively studied the echocardiographic features of the newborns referred to our neonatal intensive care unit for persistent pulmonary hypertension of the newborn who were randomized to receive inhaled nitric oxide or conventional therapy.
Results: Among 85 neonates studied with persistent pulmonary hypertension, an extrapulmonary right-to-left shunt through either a foramen ovale and/or a ductus arteriosus was present in 80 (94%) cases, with a right-to-left or bidirectional atrial shunt in 62 (73%) patients and right-to-left or bidirectional ductal shunt in 62 (73%) cases also. Left and right ventricular function were normal or only mildly depressed in the majority of the patients, whereas left ventricular index was decreased ($<2\,L/m^2/min$) in 61% of cases. In multivariate analysis, a right-to-left ductal shunt correlated with death (odds ratio, 7.8; 95% confidence interval, 1.2 to 52.8; $p = 0.04$), and there was a tendency toward greater use of ECMO therapy in patients with a predominant left-to-right ductal shunt (odd ratio, 0.13; 95% confidence interval, 0.01 to 1.22; $p = 0.07$). In the 40 patients randomized to receive inhaled nitric oxide, 28 had a positive response, defined by a 20% reduction in the oxygenation index on the post-ductal arterial blood gas. A left-to-right atrial shunt increased the risk to be a nonresponder to inhaled NO in multivariate analysis (odd ratio, 7.46; 95% confidence interval, 1.23 to 45.1; $p = 0.028$).
Conclusion: Patients with predominant left-to-right shunting at ductal level commonly required ECMO. Patients with predominant left-to-right shunting at atrial level were poor responders to inhaled NO. Patients at highest risk for mortality were those with exclusive right-to-left shunting at ductal level. Careful Doppler echocardiographic screening of patients with presumed persistent pulmonary hypertension may refine treatment groups and identify risk factors.

P231

Early survival and late outcome of direct aortic implantation of anomalous left coronary artery origen from the pulmonary artery

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Objective: To determine the early and late outcome of patients presenting with anomalous left coronary artery origen from the pulmonary artery (ALCAPA) who had repair by direct aortic implantation.
Methods: From 9/99 until 08/02, 9 patients with ALCAPA were repaired by aortic re-implantation. The mean age was 17.7 months. Prior to repair, 7 (77%) patients were in heart failure including 4 (44%) infants presenting in extremis requiring ventilatory and inotropic support. Pericardial hood was used in two patients.
Results: Hospital survival was 100%. Two children required left ventricular assist device (LVAD) for 2 and 6 days, resp. Follow up is complete, median follow up time is 24 months (5 to 40 months). So far, there were no late deaths and no need for re-intervention. At follow-up, echocardiography demonstrated significant improvements in left ventricular ejection fraction (61 + 9% vs. 28 + 19% preoperative, $p < 0.0001$), degree of mitral regurgitation (moderate or higher 15% vs. 44% pre-repair, $p < 0.02$), and wall motion abnormalities (22% vs. 89% pre-repair, $p < 0.002$). Both children who had had LVAD showed normal ejection fractions and ventricular dimensions at follow-up. Normalization of ejection fraction occurred within 1 year of repair.
Conclusion: Anatomic repair of ALCAPA by aortic re-implantation yields excellent early survival and late functional outcome even in critically ill infants. The institution of temporary LVAD improved the postoperative outcome in severe left ventricular failure.

P232

High serum levels of atrial and brain natriuretic peptide after transcatheter ASD closure

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Background: Many studies have reported that natriuretic peptides are effective markers for evaluating cardiac situations in patients with congenital heart disease. Atrial septal defect (ASD) is one of the most common congenital heart diseases. In patients with ASD plasma A-type natriuretic peptide (ANP) and B-type natriuretic peptide (BNP) levels may be elevated. We evaluated the ANP-BNP plasma level modifications after ASD transcatheter closure during a mid-term follow-up.
Methods and Results: We studied 44 consecutive patients with an uncomplicated ASD and 10 healthy control subjects matched for age and sex. All patients underwent ASD transcatheter closure. Blood samples were obtained from patients and controls. Plasma ANP and BNP levels were measured directly with specific immunoradiometric assay kits (Shiono RIA ANP assay kit and Shiono RIA BNP assay kit, Shionogi Co., Ltd., Osaka, Japan). The ANP and BNP concentrations compared to controls are significantly higher before transcatheter closure (53.5 ± 57.8 pg/ml vs. 14.5 ± 8.3 pg/ml, $p < 0.0001$). At 1- and 6- months post closure, the ANP concentrations decreased but are still more elevated that control subjects (Fig. 1). Different from ANP, BNP persists elevated at 1- and 6-month after closure (Fig. 2). BNP serum levels correlate positively with the age of the patients at the transcatheter procedure.
Conclusion: These results testify that atria and ventricles do not normalize at medium term after ASD transcatheter closure.

P233 (see Abstract 115)
Differential proteomic profiling of right ventricle after 3 weeks of RV pressure overload in young rats
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P234
Long-term follow-up of left atrial and left ventricular epicardial pacing leads in children
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Background: Left atrial and left ventricular epicardial pacing with pacing lead insertion through a lateral thoracotomy is an alternative approach for epicardial pacing. However, long-term follow-up data of left atrial and left ventricular pacing leads are scarce. We analysed survival and performance of left atrial and left ventricular pacing leads.
Methods: In 34 children, aged 6.9 ± 4.8 years, 24 DDD(R) and 10 VVI(R) pacing systems were implanted using bipolar steroid eluting pacing leads (Medtronic CapSure Epi 10366 or 4968) and various pulse generators. Pacing leads were inserted through a left lateral thoracotomy in 33 and sternotomy in 1 child and fixated on the left atrium or left ventricle respectively. Congenital heart disease with previous cardiac surgery was present in 24 children. Indications for pacing were postoperative heart block in 13, sinus node disease in 10, congenital heart block in 9, and various in 2 cases. Threshold values and measured data were obtained at 6 months intervals. The mean follow-up was 3.7 ± 2.4 years.
Results: Lead survival for atrial leads was 95% and 86% and for ventricular leads 96% and 88% at 1 and 5 years respectively. Atrial and ventricular lead follow-up data demonstrate only significant changes in impedance and ventricular sensing (see table).
Conclusion: Left atrial and left ventricular epicardial pacing leads inserted through a left lateral thoracotomy demonstrate a high probability of survival with persisting low pacing thresholds and adequate sensing thresholds at long-term follow-up. Thus, reliable permanent pacing can be achieved with epicardial access through a lateral thoracotomy.

	1 Year	5 Year	
A Imp Ohm	671 ± 61	732 ± 105	p = 0.01
A V @0.5 ms	1.4 ± 0.5	1.3 ± 0.5	ns
A S mV	3.9 ± 1.8	4.0 ± 1.7	ns
V Imp Ohm	721 ± 119	784 ± 148	p = 0.02
V V @0.5 ms	1.5 ± 1.0	1.2 ± 0.5	ns
V S mV	10.2 ± 3.7	11.6 ± 3.8	p = 0.04

P235
Persistence of left superior vena cava in the pathogenesis of left ventricular outflow tract obstruction
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Introduction: Left superior vena cava (LSVC) is clinically relevant during administration of cardioplegia in cardiopulmonary bypass, cavopulmonary anastomosis, pacemaker lead implants and cardiac transplantation. It is present in 0.1–0.4% of the general population. The association of LSVC and coarctation has not been reported before.
Objective: To evaluate the association of LSVC in the pathogenesis of left heart outflow obstructions by non-invasive methods and hypothesize the possible cause.
Methods: This retrospective study was conducted in children who had surgery for left ventricular outflow tract obstruction at a tertiary referral centre from 1st June 2000 to 30th June 2002. Case notes and echocardiogram reports were studied with the corresponding operation notes. Patients with hypoplastic left heart syndrome and complex heart disease with isomerism and those with unobstructed morphologic left ventricle were excluded from the study.
Results: Fifty-three patients were identified to have had surgery for left ventricular outflow tract obstruction. Amongst them thirty-eight had associated coarctation of the aorta (71%). Twenty patients with coarctation needed repair in the neonatal period (20/38, 52.6%), and eighteen were repaired after 4 weeks of age. Persistent LSVC was identified in 7 patients (11.5%). However, in babies needing coarctation repair in the neonatal period, the proportion of persistent LSVC was much higher (5/20) at 25% compared to the rest (2/33, 6%).
Conclusions: Persistent LSVC is more common in patients with aortic coarctation than in the general population. This association is stronger in severe coarctations, presenting in the neonatal period. We hypothesise that the direction of flow from the left SVC to the coronary sinus being counter-current to that of the flow from the umbilical vein to the foramen ovale, the presence of a dominant LSVC, reduces the effective blood flow into the left heart. This results in the hypoplasia of the left heart and its ouflow tract. Preferential flow through the arterial duct maintains flow related post-ductal arch development. This may contribute to justa ductal coarctation, arch and left heart hypoplasia presenting in the early neonatal period.

P236 (see Abstract 109)
One stage correction for aortic arch obstruction and ventricular septal defect
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P237 (see Abstract 120)
Amiodarone: efficacious treatment of refractory fetal tachycardia
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P238
Heartrate-variability a sign for an effective therapy in infants with cardiac failure due to left-to-right-shunting
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Background: Infants with signs of cardiac failure due to a left-to-right shunt have a stimulated neurohumoral system. Betablocker (BB) lead to a damping of the neurohumoral system. Does betablocker lead to an improvement of symptoms and is the heartrate-variability helpful in steering the treatment?
Method: 25 infants (age 72 ± 44 days) were followed with signs of cardiac failure due to left-right shunting. We measured in all infants daily weight gain (wg), respiratory rate (rr), heartrate (hr) and the heartrate-variability (sNN50gess, SDANN, SDNN, RMSSD) before and after we started either propanolol or metoprolol (target dose 1,5 to 2 mg/kg/d). The heartrate-variability parameter were analyzed in a multivariant analysis ($p > 0,05$ as significant). The medical therapy was always meant to stabilize the infant untill surgery was possible.
Results: All infants improved clinically and gained more weight then before starting betablocker. RMSSD and sNN50ges reflected well the clinical improvement of these infants, whereas the other parameters such as SDNN or SDANN showed no correlation to clinical signs.
Discussion: The heartrate-variability parameter RMSSD and sNN50ges reflected well the clinical improvement in infants. We used these parameter to help to decide if the betablocker dose is sufficient. The other heartrate parameters did not reflect the clinical situation of the infants at all. All infants had no problems during or after the following operation.

	rr	wg	hr	before BB	after BB
SDNN	ns	ns	$p < 0,01$	40,8 ± 27,1	42,5 ± 15,3
SDANN	ns	ns	ns	35,5 ± 14,3	37,0 ± 13,6
SNN50ges	$p < 0,05$	$p < 0,01$	ns	915,7 ± 1460	4121 ± 7157
RMSSD	$p < 0,05$	$p < 0,01$	ns	12,5 ± 5	18 ± 10,2
Wg				0,7 ± 16,1	35,6 ± 21 gr/d
Rr				60,9 ± 13,7	51,6 ± 10,5/min
Hr				140,3 ± 14,8	120,8 ± 11,2/min

P239 (see Abstract 110)
Long-term mortality and complication rate in children with mechanical prosthesis: a 30 years single center experience
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P240
Non connection of the left pulmonary artery in two neonates with DiGeorge syndrome and cono-truncal or great arteries anomalies: importance of early surgery for the recovery of the involved lung
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First Case: A 2-day-old neonate was transferred to our Unit for uncertain echocardiographic arrangement of the aortic arch (AA) and transient cyanotic episodes. Physical examination, chest X-ray, ECG were unremarkable. The echocardiogram (ECHO) showed: right AA, as first branch the left carotid artery, as second branch the right carotid, as third branch the right subclavian artery. Left subclavian artery was unclearly identified and postulated as retroesophageal vessel determining a vascular ring. A ductus was detected draining into the right pulmonary artery (RPA), whereas the left pulmonary artery (LPA) was small, non connected to the main pulmonary trunk (MPT) and receiving flow from an AA connection. MRI confirmed the hypothesis. An angiography was performed for the surgical plan. DiGeorge syndrome was also clinically assumpted and recognized by genetic study. Surgery was performed within the first month of life reconnecting LPA to MPT and disconnecting LSA to eliminate the vascular ring. After a 4-month follow-up no signs of stenosis were detected by ECHO at the level of LPA reconnection with good flow.
Second Case: A 1-day-old neonate was admitted to our Unit for truncus arteriosus (TA). ECHO showed TA, right AA, mirror-image brachiocephalic branching, pulmonary trunk emerging from the common trunk and continuing as a single RPA. LPA was not connected to the pulmonary common trunk but was detected as a small extrapericardial vessel receiving continuous flow from an arterial connection originating from AA. DiGeorge syndrome was later confirmed. Complete surgery, performed on the 27th day of life, restored direct connection between the pulmonary trunk and the LPA.
Conclusions: An early and accurate neonatal diagnosis of discontinuity of one pulmonary artery is important in order to restore a bilateral pulmonary flow and normal function of the involved lung, reducing operative risks in more complex cardiac diseases. Our two cases of non connection of a pulmonary artery, in DiGeorge syndrome, suggest that so-called "absent" pulmonary branch artery could be present in the first weeks of life becoming obliterated with time.

P241
Clinical course and management strategies for hemolysis following catheter closure of patent arterial ducts
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Residual flows following transcatheter coil or device closure of the patent ductus arteriosus (PDA) can result in hemolysis. Management of hemolysis after coil closure often requires emergent elimination of residual flows. We reviewed the data of 611 patients who underwent transcatheter PDA closure at our institution from July 1998–December 2002. Five (0.8%) patients (age: 6–63 years) developed overt hemolysis (after coil occlusion in 4 and after Amplatzer device closure in one). All patients had significant residual flows after the procedure and, except one patient with a 3 mm duct, all had large ducts (4.6–12.5 mm). In one patient, hemolysis occurred 3 months after coil occlusion following a period of uncontrolled hypertension. Hemolysis was associated with a fall in hemoglobin of 3–6 g/100 ml ($n = 3$), jaundice ($n = 2$) and renal failure ($n = 1$). Hemolysis subsided spontaneously in one patient and 4 patients required flow elimination. Deploying additional coils in 3 patients eliminated residual flows. In one patient (after Amplatzer device closure for 12.5 mm duct with aneurysm) flow

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persisted after 25 additional coils, transient balloon occlusion and gel foam instillation. Flow elimination was eventually achieved through thrombin instillation after balloon occlusion of the ampulla. All patients recovered completely and were well on follow up. In our series, hemolysis was relatively rare and essentially restricted to the large ducts with residual flows. Residual flow at the end of the procedure merits careful monitoring because of the potentially serious effects of hemolysis and aggressive elimination of residual flows is often necessary to control hemolysis.

P242
Left ventricular echographic changes with recombinant enzyme therapy in infantile Pompe disease

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Pompe disease is a lethal cardioskeletal myopathy in infants and results from genetic deficiency of the lysosomal enzyme acid alpha-glucosidase (GAA). The purpose of this study was to investigate the echocardiographic cardiac changes with the intravenous recombinant human GAA (rhGAA) enzyme therapy.
Methods: ECG, echocardiographic and Doppler data from the first infant enrolled in a protocole rhGAA enzyme therapy, was serially collected: before treatment, at 1, 2, 3, 4, 5, 8, 11 and 15 month after initiation of therapy. Measured parameters included: ECG Sokoloff index, left ventricular diastolic diameter (LVDD), posterior wall thickness (PWT), interventricular septum thickness (IVST). LV mass and LV mass index were calculated. Doppler records of mitral PHT, subaortic velocity and intraventricular gradient were assessed at each time points. The patient was 2.5 months old at onset of rhGAA enzyme therapy.
Results: Initial clinical symptoms of cardiac failure and muscular hypotonia resolved within 1 month after onset of therapy. Sokoloff ECG index dramatically decreased from 200 mm to 35 mm within one-month therapy, and maintained within normal range (28 to 40 mm). Table shows the LV echographic changes: LVDD ranged within normal value; increased initial PWT and IVST turned to upper limit of normal value within 3 months. LV mass decreased from 75.8 g (before therapy) to 35 g (3rd month), 31 g (5th month) and 29 g (15th month). Initial LV mass index was 316 g/m^2 and decreased to 142 g/m^2 (3rd month), 91 g/m^2 (5th month) and 65 g/m^2 (15th month). Doppler mitral PHT did not change significanly, but inversed A and E mitral waves normalized. Intraventricular obstruction resolved at 1 month-therapy evaluation.

Time (mth)	LVDD (mm)	PWT (mm)	IVST (mm)
Before	20	11.5	16
1	24	8.5	14
2	22	7.5	7.5
3	23	6	8
5	23.5	5.9	7.5
8	23	5.5	5
15	24	4.8	6

Conclusion: ECG Sokoloff index is the earliest parameter of LV changes. LVPWT, IVST, LV mass and LV mass index turn to normal range within 3 months enzyme therapy, and remain stable up to one year therapy.

P243 (see Abstract 116)
N-Terminal Pro-B-Type natriuretic peptide. Normal plasma levels from birth to adolescence. Elevated levels at birth, and in patients with heart disease

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P244
Survey on prophylaxis and management of infective endocarditis in patients with congenital heart disease. Japanese nation wide survey

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Guidelines for prevention and management of infective endocarditis (IE) in children with congenital heart disease (CHD) have not been established. The aim of this study is to clarify the incidence, practical prevention and management of IE in patients with CHD in a Japanese nation-wide survey. A written questionnaire was sent to members of the Japanese Society of Pediatric Cardiology and Cardiac Surgery. Information was obtained from 236 cardiologists in 228 institutions. Four hundred and eight patients with IE hospitalized during 1997 to 2001 (1/173 admissions with CHD). Prevention of IE for CHD was undertaken by 92% of cardiologists. Penicillins (73%) and less frequently cephems (18%) were prescribed for oral IE prevention. The Duke criteria were used as clinical criteria in 38%. Blood culture was performed only once in 40%. Penicillins and aminoglycosides (38%) were frequently administered for management of culture-negative IE. Dose and duration of antibiotics for prevention and management of IE revealed large variations among cardiologists. It appears that prevalence of IE in CHD is rising. Practical prevention and management of IE in patients with CHD revealed large variations than expected. Our study may be helpful in making a future guideline of IE in CHD.

P245
Balloon versus surgical therapy as the initial palliation for pulmonary atresia with intact ventricular septum – multicenter study in Korea

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There is a controversy with regard to the role of balloon valvuloplasty (BV) as the initial palliative therapy for pulmonary atresia (PA) with intact ventricular septum. From 1993 to 2001, 146 patients with PA were analyzed to evaluate this issue. 78 underwent BV while 68 had surgery, it depended on the policy of each institution. The two groups are nearly identical with respect to age, weight and Z-value of pulmonic valve, but Z-value of tricuspid valve were -0.8 ± 1.6 in BV group, and -2.2 ± 2.1 in surgery group ($p < 0.001$). BV was successful in 54 of 78 (69%). In conclusion, BV is superior to surgery as the initial palliative therapy for PA

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with regard to the biventricular repair and survival rate. RV size and Z-value of TV are crucial for biventricular repair of PA.

		Bi-repair	mortality
Palliation	BV	58/78(74%)	13/78(17%)
	surgery	20/68(29%)	18/68(26%)
RV size	mild hypo	40/46(87%)	3/46(7%)
	severe	15/55(27%)	13/55(24%)
TV	$Z > -3$	55/87(63%)	17/87(20%)
	$Z < -3$	2/23(9%)	4/23(17%)

P246
Aortic root dilatation in Marfan's Syndrome, when it develops?

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Aims: To determine when Marfan Syndrome (MFS) patients develop aortic root dilatation and if it increases with age. When we can say the aorta is normal and how it grows in people who do not dissect.
Methods: 160 patients <40 years of age with MFS were reviewed. We looked at those with an abnormal cardiac examination and dilated aortic root. 2D-Echocardiography on first visit and on subsequent follow up was measured at sinus of valsalva and comparing it to normal value in relation to body surface area. Group I (n = 31): who have normal aortic root when first time seen but they became abnormal on subsequent follow up. Group II (n = 84): Those who have abnormal aortic root when first time seen. Group III (n = 45): Includes all patients who have normal aortic root and stayed normal throughout the follow up period. Skeletal manifestations, eye involvement and those who are on B-blockers were all recorded.
Results: MFS is more common in males than females (1.4:1). The age range was 11 months to 40 years. Family history is positive in 75%. Skeletal abnormalities are present in 95%. Eye involvement was found in 18%. 37 patients (23%) have significant cardiac murmur. Echocardiography shows that 115/160 (72%) have aortic root dilatation, 77/115 (72%) patients with dilated aortic root are <19 years. 32 patients (20%) have mitral regurge and 29 (18%) have mitral valve prolapse, 10 (6.25%) with aortic regurge and 6 patients (3.75%) have congenital heart disease, of these bicuspid aortic valve and ASD are the commonest. 14 patients (9%) had surgery for aortic root replacement.
Conclusion: Aortic root dilatation develops early in MFS 69% in our population by 19 years of age. It does increase with age but definitely less after the age of 19 years. There is no age limit to say the aorta will stay normal, but it does grow variably in those who did not dissect.

P247
Contegra-Venpro conduits in the right ventricular outflow tract: results after intermediate-term follow-up

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Aim of the Study: Intermediate-term assessment of the Contegra-Venpro graft in the right ventricular outflow tract.
Patients and Methods: Between 5/2000 and 9/2002, 56 Contegra-Venpro conduits (12 to 22 mm), originating from bovine jugular veins, were implanted in the right ventricular outflow tract during primary repair (n = 26) or redo surgery (n = 30).
Primary Repair: Ross operation (n = 10, age 11.5 ± 2.4yrs), TOF (n = 7, age 0.2–48 yrs), truncus arteriosus (n = 8, age 0.3–1.7 mths) or Rastelli repair (n = 1, 1.8 yr).
Redo surgery: Pulmonary regurgitation after TOF repair (n = 19), after critical pulmonary stenosis (n = 1) or complex VSD closure (n = 1) and pulmonary stenosis after truncus repair (n = 6) or Rastelli repair (n = 3) (age 12.6 ± 3.6 yrs). Follow-up was performed serially at 1, 6, 12 and 24 months; mean 11,4 mths (0.2–27.3 mths).
Results: Hospital mortality was 3.5% (2/56): 1 died from Staphylococcus aureus septicemia after 12 days, 1 from enterococcal endocarditis after 12 weeks. During follow-up 15/56 patients (26.7%) developed significant supravalvular stenosis at the distal anastomosis with systemic or suprasystemic RV pressure, requiring intervention. This was more frequent in the patients who underwent primary repair at young age (0.3–36 mths) for truncus arteriosus or TOF (9/15 = 60%), whereas the incidence was 6.6% (2/30) in the redo group and 10% (1/10) after Ross operation.
Secondary Procedures: balloon valvuloplasty in 7 pts 10,4 ± 1,4 mths after surgery, stent implantation at bifurcation in 8 pts (bilateral n = 6, unilateral n = 2) 8.3 ± 3.1 mths after surgery. In 4 patients the graft was subsequently explanted because of severe restenosis after stent implantation (n = 3, time interval 12–19 mths) or because of Streptococcal endocarditis (n = 1, time interval 11 mths). In the remaining 41 patients, there was some valve thickening and shrinkage of the graft, with a mild to moderate valvular and supravalvular stenosis (0–60 mmHg Doppler gradient). In 49 patients pulmonary regurgitation was mild, in 6 patients moderate and in 1 patient (who died early from endocarditis) severe.
Conclusion: Intermediate-term results of a Contegra-Venpro graft in the right ventricular outflow show a high incidence of significant stenosis at the distal anastomosis, especially in infants and young patients. Moreover, the graft seems to be less resistant to bacterial endocarditis.

P248
New technique of right heart bypass in congenital heart surgeries with autologus lung as oxygenator

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Background: Modifications have been made in the cardiopulmonary circuit to reduce the inflammatory deleterious effects and the cost. We present our experience of one such right heart bypass (RHB) circuit utilizing autologus lung as the oxygenator.
Methods: From September 2001–August 2002, 15 patients underwent congenital heart surgeries with this technique. Thirteen patients underwent Bi-directional Glenn (12–24 months, 6–10 kgs). One patient (26 yrs) underwent central shunt with enlargement of the confluence and left pulmonary artery. Another patient (18 months) underwent one and half ventricle repair. The bypass circuit consisted of a reservoir with an in-built heat exchanger and a roller pump along with a cardiotomy sucker. The left pulmonary artery/main pulmonary artery were used for arterial return and the venous circuit was connected to innominate vein and the inferior vena cava (IVC) if required.
Results: There were no hospital deaths. The mean flow achieved on RHB was 0.57 ± 0.3 l/min/m^2, central venous pressure (CVP) was 3.3 ± 1.8 (0–7) mmHg, and mean arterial pressure could be maintained satisfactorily in all the patients (54 ± 14 mmHg). The Mean RHB time was 54 ± 14 mins. Mean CVP was 10.1 ± 2.4 mmHg after the procedure and the saturation was similar to

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that of RHB (88 ± 8%). The mean amount of drainage was 9.1 ± 4.2 cc/kg/24 hours. Avoiding the oxygenator and reducing the number of tubes achieved a combined cost savings of 40% for all the procedures.
Conclusion: Right heart bypass is a simpler, easily reproducible, and less expensive alternative to conventional cardiopulmonary bypass. This technique allows effective decompression of superior vena cava and adequate oxygenation. It in addition, predicts saturation after the Glenn shunt. It can also be applied to construct central shunts and pulmonary artery reconstructions with cost containment.

P249

Low s-albumin in children as a risk factor for coronary heart disease

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Background: Low serum albumin has been reported to be a risk factor for Coronary Heart Disease (CHD). Albumin has antioxidant properties, it inhibits the production of free hydroxyl radicals and may inhibit endothelial apoptosis. Infections may modify the risk factors for coronary heart disease (CHD). Vascular pathology and altered lipoprotein metabolism seem to be associated with inflammation and infectious diseases. They are associated with increase of acute phase proteins, which reflect the strength of an infectious stimulus. Endothelial cell damage during an acute insult like infection may induce subendothelial oedema, which during repeated insults may permanently thicken the arteries.
Patients and Methods: 2458 individuals, aged 9, 12, 15, 18 and 21 years, were investigated in the first visit in 1983 and again 3 years later. During the 2 weeks before their second follow-up visit in 1986, 114 subjects out of 2458 (4.6%) had a history of infection before the follow-up visit. The fasting serum lipid values in 1983 served as "normal" values of patients (n = 95), which were compared to values in 1986. Paired student's t-test was used for statistical method.
Results: Acute phase proteins were elevated after infection. Serum CRP, haptoglobin, α1antitrypsin, orosomucoid and ferritin correlated significantly to a change in serum HDL cholesterol, HDL/total cholesterol and total cholesterol between 1983 (no infection) and 1986 (infection), ($p < 0.0001$–0.0003). These lipoproteins and lipid fractions were lower during the infection than during the control (healthy) period ($p < 0.04$–0.0001). The mean value for albumin in control group (1983) was 45 g/l and for infection group (1986) = 38 g/l ($p < 0.001$).
Conclusion: The "good" cholesterol, serum HDL cholesterol and serum albumin decrease during an infection. Many physiological mechanisms suggest that albumin is not only an acute phase reactant but low s-albumin has an independent role as a risk factor for CHD. Especially repeated albumin lowering insults might be etiological factors for CHD.

P250

Hypertrophic cardiomyopathy – whom does it affect and how serious in childhood?

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Introduction: Hypertrophic cardiomyopathy (HCM) in infancy and childhood is a rare but serious disease. Studying results of patients with HCM is important to get better insight in clinical outcome related to the underlying cause.
Aim: Giving an overview of children with HCM in our institution, describing clinical course of HCM related to the cause. Furthermore, reporting the output of screening for familiar HCM.
Methods: Data charts from all patients with HCM according to our database in the period 1974–2000 have been analysed regarding cause, mode/onset of presentation, outflow tract obstruction, therapy and clinical outcome. In addition, data from patients screened for familial HCM in the past 10 years have been analysed.
Results: HCM was found in 42 patients. Fifteen patients (36%, aged 2.6 ± 4.8 yrs) had syndrome-related HCM, 10 had Noonan's syndrome. Symptoms at presentation were found in 7 cases (all Noonan's), 8 had outflow tract obstruction (all Noonan's), of whom 4 underwent surgical reduction of muscle tissue, 5 were on medication (β-blocker/Ca antagonist), and 2 children died. Five patients (12%, aged 1.4 ± 2.8 yrs) suffered underlying metabolic disease, 3 of them died at young age. Thirteen patients (31%, aged 5.6 ± 4.8 yrs) had familial HCM, only 1 presented with symptoms, 2 showed outflow tract obstruction, 1 underwent surgery to reduce muscle tissue, 1 was put on a β-blocker, and 1 suddenly died. Seven patients (17%, aged 4.6 ± 5.6 yrs) showed familial negative HCM without other cause, of whom 4 presented with symptoms, 2 showed outflow tract obstruction, 1 was put on a Ca-antagonist, and 2 children died. Two infants (5%) had other miscellaneous underlying disease, 1 infant died. Screening for familial HCM revealed 10/111 positive cases, of whom 80% had a first degree affected family member.
Conclusions: HCM in infancy is mainly due to syndrome-related (especially Noonan's syndrome) and metabolic disease with a poor prognosis. At school age both family related and non-related HCM is predominant with the worse prognosis in childhood in the latter. Screening for familial HCM in childhood is mainly negative.

P251

Detection of allo- and autoreactive antibodies in patients with protein losing enteropathy (PLE) in Fontan patients by flow and laser scanning cytometry (LSC)

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PLE is a feared late complication occurring 5–10 yrs after Glenn/Fontan type of cardiac surgery with 5–15% of the patients exhibiting a substantial decrease of serum protein by increased secretion of protein. The mortality among patients with a manifest PLE is >60% but its aetiology is yet completely unknown. In a follow up study we analysed patients after Fontan surgery over a period of five years by flow cytometry (FCM) and serology. One patient developed PLE nine months after surgery. Its immune sequel was compared to that of seven patients with a manifest PLE, PLE-free Fontan patients and healthy controls.
The immune alterations after PLE is comparable for all affected children. It includes the dramatic selective loss of >80% of the circulating alpha, beta T-cell receptor positive CD3+ 4+ cells. In our essays we tried to find the rational for the selective cell loss. We developed various assays for the FCM and LSC to quantify binding of autoantibodies to cells and tissues. With an FCM based assay we found in the serum of 25% of the PLE patients antibodies binding to leukocytes, especially to T-helper cells. Serum of PLE free Fontan patients and of healthy controls was negative. In 25% of the patients with manifest PLE we found antibodies against myocardial structures by LSC. None of the PLE patients but one Fontan patient without PLE had antinuclear antibodies. Because of these

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results we hypothise that autoimmunity is at least associated if not participates in the aetiology of PLE.
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P252
Realtime blood flow quantification by phase-contrast MRI using single-shot EPI combined with SENSE
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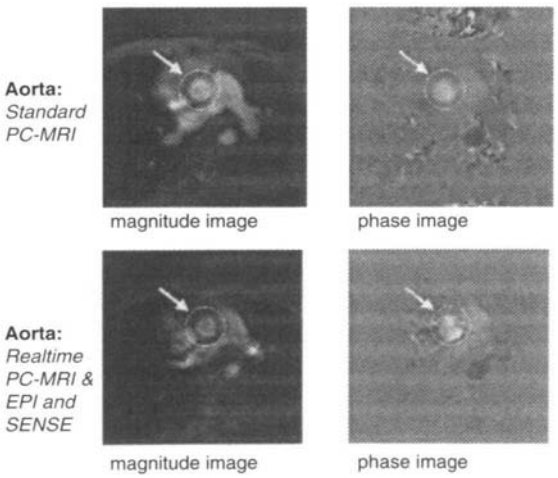
Purpose: To evaluate free-breathing realtime phase-contrast MR techniques using non-triggered single-shot EPI (Ssh-EPI-PC) and sensitivity-encoding (SENSE) for beat-to-beat cardiac stroke volume quantification.
Method: 3 non-triggered Ssh-EPI-PC sequences were tested in-vitro, and applied in a total of 44 subjects to quantify stroke volumes in the ascending aorta/pulmonary artery. We used Ssh-EPI-PC with (a) SENSE-reduction factor (SF) of 3 (n = 40/31); (b) SF 3 with fat suppression (n = 26/21), both protocols with in-plane resolution 2.3 × 3.1 mm and phase-intervals 40 and 46 ms; (c) SF 4, increased flip-angle of 40, symmetrical in-plane resolution 2.7 × 2.7 mm, phase-interval 39 ms (n = 11/10). Results were compared with conventional retrospectively-ECG-gated phase-contrast MRI (PC-MRI).
Results: Aortic (pulmonary) stroke volumes were underestimated by a mean of 10–13% (13%) using Ssh-EPI-PC with SF 3, both with/without fat suppression, with a wide scatter, compared with conventional PC-MRI using Bland/Altman analysis. In contrast, Ssh-EPI-PC with SF 4 differed only by 2–3% (both vessels), with a range of −19% to +17% for aorta and −18% to +17% for pulmonary artery (=mean ± 2SD). High repeatability but some flow overestimation was observed in-vitro with any Ssh-EPI-PC method, suggesting EPI-related in-vitro phase-errors, whereas conventional PC-MRI was accurate. Beat-to-beat stroke volume variation in-vitro was 7.4 ± 0.6%, and 16.8 ± 5.4% in-vivo (45 measurements in a subpopulation of 10 participants).
Conclusions: Ssh-EPI-PC with SF 4, flip-angle 40, symmetrical in-plane resolution of 2.7 × 2.7 mm and phase-interval of 39 ms, allows reliable realtime beat-to-beat quantification of aortic and pulmonary stroke volumes. Use of smaller SFs and fat suppression was unfavourable.



P253
Differences in the acute immune response between homograft and xenograft implantation
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Little is known about the host reaction to a xenograft implant. The more and more frequent usage of xenograft implants requires a comparison of the host response to homograft and xenograft implants. Differences in the immune response of the host between homo- and xenograft implantation may be useful for the understanding of the long time reaction and perhaps rejection. Therefore, we compared immunological reactions before (−1d), one day (1d), 2 days after surgery (2d) and at discharge (>3d). The following groups were analyzed: homograft (group 1, n = 6), xenograft recipients (group 2, n = 9) and reoperations changing from homograft to xenograft implants (group 3, n = 8). Complement factors (C1, C3, C3d, C5a, C5), serological data (TNF, sICAM, PECAM, sE-, sL-, sP-selectin, neopterin) and haemogram parameters (white and red blood cells, hemoglobin, hematocrit) were examined. Group1 had lower levels of the complement factors C1 at 1d (p = 0.002 vs. group2, p = 0.02 vs. group3) and 2d (p = 0.003 vs. group2, p = 0.01 vs. group3). Furthermore, group1 had lower levels of C3 1d (p < 0.0005 vs. group2) and 2d (p = 0.0008 vs. group2, p = 0.02 vs. group3). In contrast, group1 recipients had higher C5a levels at >3d (p = 0.03 vs. group2, p = 0.02 vs. group3). Serological data: Group2 had higher sICAM levels −1d (p = 0.01 group 1 and 3 respectively). Group1 had lower PECAM levels -1d (p = 0.03 vs. group2, p = 0.04 vs. group3). Furthermore, the difference remained significant compared to group3 at 1d (p = 0.03) and >3d (p = 0.003). Group2 had lower concentrations of IL-2r than group3 at 1d (p = 0.02), 2d (p = 0.03) and >3d (p = 0.02). Blood picture: group1 had a higher lymphocyte count than group3 (p = 0.003). Furthermore, group1 has a higher monocyte count at 1d (p = 0.01 vs. group2, p = 0.04 vs. group3). There were no significant differences in all other parameters. We conclude that there were no specific differences in the acute immune response between homograft and xenograft implantation during the first contact with the implant. The study has to be continued to test for differences which may appear later after surgery. If the rejection of xenografts is not much more, equal or even less harmful than that of homografts the implantation of xenografts shows hardly disadvantages.
Support: Deutsche Gesellschaft für Konderkardiologie, Herzkind e.V.

P254 (see Abstract 97)
Early cardiopulmonary functional improvement following transcatheter atrial septal defect closure
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P255
Catheter closure of large adult patent ductus arteriosus using simultaneous double or triple implantation of 0.052 inch gianturco coil
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Background: Technical difficulties still remain for catheter closure of adult patent ductus arteriosus (PDA) because of its morphological feature and calcified ductal wall. Such difficulties have been

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brought the high incidence of residual leakage and hemolytic complications.
Purpose: To evaluate the technical advantages of simultaneous double or triple implantation of 0.052 inch Gianturco coils for adult PDA patients.
Patients and Methods: Twenty-two adult PDA patients were treated by coil occlusion. Their median age was 49 (range 18 to 77) years, and median size of ductus was 4.1 (2.5 to 7.8) mm. Clinical symptoms such as palpitation or significant cardiomegaly were found in all patients. Thirteen patients were treated by 0.052 inch coil (0.052 group) and nine patients were treated by 0.038 inch detachable coil (0.038 group). 0.052 inch coil was implanted by 3 French bioptome hold control system though a 4 French long sheath. To implant 2 or 3 coils simultaneously, delivery catheters, which were introduced from femoral vein or artery, were passed through the ductus. Angiography or color Doppler echocardiography was used to confirm residual shunt. Systemic hypertension required medication was complicated in 14 patients.
Results: Complete occlusion at immediately after the procedure was confirmed in 11 patients in 0.052 group and in 8 patients in 0.038 group. Complete occlusion rate at 1 month after the procedure was obtained in all in 0.052 group and in 8 in 0.038 group. Compare to 0.038 group, 0.052 group has significantly short fluoroscopic time (12 vs. 24 minutes, $p < 0.05$) and less number of coil used (2.2 vs. 3.1, $p < 0.01$). Complications such as coil migration, hemolysis, LPA stenosis were not found in our patient population. Significant elevation of systemic arterial pressure, especially diastolic pressure, was detected in all, however such elevation declined to pre-operative level within 12 hours.
Conclusion: Simultaneous double or triple implantation of 0.052 inch Gianturco coil has the high acute complete occlusion rate and shorter procedure time compare to 0.038 inch detachable coil. Because this procedure is simple and matched any type of ductus arteriosus, it may have clinical implication for catheter closure of relatively large adult PDA.

P256
Value of tissue Doppler strain rate in the evaluation of right ventricular function in patients following corrective surgery of tetralogy of Fallot (TOF)

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Objective: Strain rate imaging is a promising method of displaying myocardial deformation properties and is unlike to myocardial Doppler velocity independent of the global heart motion. This study was carried out to evaluate the right ventricular function in postoperative TOF using tissue Doppler and applying this novel parameter.
Material and Method: In 10 asymptotic postoperative TOF patients and in 10 aged matched normal subjects, basal and middle RV free wall myocardial velocity and strain rate were examined in the apical four chamber view.
Result: Systolic and early diastolic myocardial velocity in the right ventricular basal and middle segments were significanly lower in patients with TOF patients compared to normal group($p < 0.01$). On the other hand, the strain rate was significantly lower only in the middle segment during systole ($p < 0.01$) and early diastole ($p < 0.01$). Significant correlation was found between the measured systolic ($r = 0.98$, $p < 0.01$) and early diastolic strain rate ($r = 0.97$, $p < 0.01$) in the basal and middle RV free wall and their corresponding locations in the intraventricular septum. No correlation was found between the LV free wall strain rate and intraventricular septum.
Conclusion: In tetralogy of Fallot, the middle segment of the right ventricular free wall appears to be the more affected region in the studied segments both during systole and diastole. Abnormal wall motions in the intraventricular septum seems to be related to same underlying pathology affecting the RV. To determine the culprit for global RV systolic dysfunction further regional analysis of the right ventricular wall motions is needed.

P257
Trial balloon occlusion for large patent ductus arteriosus with elevated pulmonary vascular resistance

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The feasibility of temporary balloon occlusion of PDA in the catheterization laboratory allows assessment of hemodynamic effects of duct closure. However, the utility of this maneuver has not been adequately tested. Eleven patients with large PDA and severe PAH and elevated pulmonary vascular resistance (PVR) suspected after clinical and non-invasive evaluation underwent cardiac catheterization for suitability for closure, from July 1998 to December 2002. Patients with lower limb saturation <85% and those with clinically obvious large left to right shunts were excluded. Trial balloon occlusion was done using compliant balloons (24–33 mm). The PA systolic pressures increased, remained unchanged or fell by <10 mmHg after balloon occlusion in 4 patients who were felt to be clearly inoperable. Seven patients were thought to have significant fall in PA pressures after balloon occlusion and were advised surgery. Ducts were closed with the Amplatzer PDA occluder in 1, coils in 3 and, surgically in 3. We compared the hemodynamic data between patients with a sustained reduction in PA pressures (RV systolic pressures <2/3 systemic) after duct closure ($n = 5$) with those who did not have a sustained reduction/considered inoperable ($n = 6$). The parameters that were significantly different between the two groups were the following (a) baseline PVR/SVR ratio <0.5 ($p < 0.01$). (b) On balloon occlusion: PA mean/AO mean (0.49 ± 0.08 vs. 0.74 ± 0.2; $p < 0.01$), absolute PA diastolic pressures (31 ± 7 mmHg vs. 45 ± 17 mmHg; $p = 0.04$) and, fall in PA diastolic pressure (33 ± 13 mmHg vs. 13 ± 9 mmHg, $p = 0.006$). The parameters with borderline significance were absolute values of PA mean and decline in PA mean pressure after balloon occlusion and baseline lower limb PO_2 levels.
Conclusion: A decline in PA mean pressures to approximately half systemic levels during balloon occlusion and a fall in PA diastolic of >30 mmHg could predict a favourable outcome after closure of PDA with severe pulmonary hypertension. These parameters need to be assessed in larger prospective studies.

P258
Patient dose in paediatric interventional cardiology: effect of additional beam filtration

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In interventional paediatric cardiology, the extensive use of X-rays can result in high radiation doses. Therefore, one has to take all

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measures to keep the dose as low as possible in view of the high radiosensitivity of paediatric patients. The aim of the study was to determine the effective dose allowing a risk estimate for stochastic effects such as cancer and leukaemia. Furthermore, the effect of additional beam filtration on the patient dose was investigated. 22 patients (12 male, 10 female) with a mean age of 3 years (range: 0.1–10 years), were referred to the interventional cardiology department for diagnostic cardiac catheterisation (n = 10) or for other interventional cardiological procedures (n = 12). All patients were examined on a biplane X-ray tube configuration (Philips Integris BH5000). In half of the patients, normal beam filtration settings were used. For the other half of the patients, the additional filtration of 0.2 mm Cu available on the equipment was inserted. The effective dose (ICRP60) was determined by Monte Carlo simulation on a patient specific anthropomorphic phantom and based on DAP measurements and recording of the beam characteristics. For the calculation of the life-time risk for stochastic effects, we used the gender-dependent risk factor for the age of 0–10 years from the multiplicative model recommended by ICRP 60. For the diagnostic cardiac catheterisation, the mean effective dose was 6.1 mSv (range: 1.0–20.1 mSv). The cine contributed for a mean of 58% in the dose. The other interventional cardiological procedures (asd, dilatation, ...) resulted in a mean effective dose of 10.2 mSv (range: 1.6–40.0 mSv). The cine had a mean contribution of 50%. The use of additional filtration, resulted in a mean dose reduction of 19% without degrading the image quality. Averaging over all procedures, a mean life-time risk estimate of 1 per 1000 is obtained. The calculated effective doses are as high as for adult interventional cardiology. Therefore, it is important to monitor the patient dose by DAP instrumentation and to use additional beam filtration to keep the effective dose as low as possible in view of the sensitivity of the paediatric patients.

P259

Transcatheter closure of coronary fistulas at the drainage point using coils or other devices

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Transcatheter closure of coronary fistulas by closing the feeding vessel may put normal side branches at risk and may result in residual shunt if multiple feeders are present. In order to determine the feasibility, safety and efficacy of selective drainage point closure of coronary fistulas we reviewed our single center experience from June 1996 until November 2002. Seven out of 8 patients underwent selective closure at the drainage point. Patient age ranged from 10 days to 18 years (median 6.9 years), patient weight ranged from 3.6 to 64.8 kg (median 21.0 kg). Origin was from the right coronary artery (2), from left coronary artery branches (3) and from both in 2. Drainage was into the right ventricle (4), the right atrium (2) and the main pulmonary artery (1). A single feeder was identified in 4 patients and 3 or more feeders were identified in 3 patients. Smallest diameter was from 1.6 to 3.2 mm (mean 2.3 mm) with an ampulla of 3.5 to 11.0 mm (mean 6.8 mm). QP/QS was 1.0 to 1.6 (mean 1.2). An arterio-venous wire rail was used in 4 patients and implantation was retrograde in 3 and antegrade in 4 patients. One to 10 coils (mean 4.6) and 1 Amplatzer PDA Occluder were implanted. Fluoroscopy time was 10.3 to 83.7 minutes (median 46.8 minutes). Complete occlusion was achieved in all patients at 24 hours. There were no ischemic EKG changes and no major complications. Supra-ventricular tachycardia occurred in 1 patient and required cardioversion. There were no late complications. Selective occlusion at the drainage point of coronary fistulas appears feasible, safe and effective.

P260

Five year single center experience with the Amplatzer atrial septal occluder

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Device occlusion of atrial septal defects (ASD) has become an alternative to surgical closure. Ongoing review of short- and medium-term results is needed to confirm its utility. 272 patients were considered for ASD device occlusion from 8/97–12/02. In 43 patients, device placement was not attempted because of unsuitable anatomy. Two hundred and twenty nine patients, aged 1.4–88.1 years (median 12.6 years) and weighing 8.7–142.7 kg (median 44.7 kg) underwent device implantation. Thirty-five patients had 2 or 3 defects. Qp/Qs was 0.4–6.5 (mean 1.9). Echocardiographic defect size was 1–27 mm. Stretched defect diameter was 4–40 mm. Fifteen additional interventions were performed: 2 pulmonary artery stent placements, 2 coil occlusions of surgical shunts, 1 pulmonary balloon valvuloplasty, 6 patent ductus arteriosus coil occlusions, 1 coil occlusion of a systemic to pulmonary vein collateral, 2 radiofrequency ablations, and 1 device closure of a ventricular septal defect. All implantation attempts were successful. Device size was 4–40 mm (median 17 mm). Two occluders were implanted in 8 patients. Implantation was from the right internal jugular vein in 6 patients. Procedure time was 34–212 min (median 98.0 min). Fluoroscopy time was 8.2–42.6 min (median 16.8 min). There were no device embolizations, no strokes and no deaths. Total early and late complication rate was 12.2% (28/229). There was 1 major complication (0.4%): hemopericard requiring surgery after a disk eroded through right atrial wall and into the aortic root within 12 hrs. There were 27 minor complications (11.8%): transient atrial ectopy in 11, migraine headaches in 7, atrial fibrillation or flutter in 3, transient AV block in 3, marker band detachment from delivery sheath in 1, transient ST elevation in 1, blood transfusion in 1. Most patients were discharged within 24 hrs. Complete occlusion of the defect was achieved in 199/227 (87.7%) at 24 hrs, in 131/143 (91.6%) after 6 months, and in 115/125 (92.0%) after 1 year. Residual shunt was more prevalent in multiple and larger defects ($p < 0.05$). All residual defects were 2 mm or less in diameter. No late complications were observed after 6 months. Closure of ASD using the Amplatzer septal occluder is safe and effective during early and medium follow-up.

P261

Ministernotomy in infants and children for correction of tetralogy of Fallot, atrioventricular septal defects and, ventricular septal defects

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Ministernotomy (MS) is gaining acceptance corrections of simple lesions like atrial septal defects (ASD) because of its cosmetic appeal. We have extended the application of MS for repair of tetralogy of Fallot (TOF), ventricular septal defects (VSD) and atrioventricular septal defects. From January 2001–December 2002, MS was employed for 139 patients (including 94 secundum ASDs) undergoing congenital heart surgery. This report describes the results of the 45 patients (16 infants, 21 females, median age 36 months,

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range: 2–120 months; median weight 10 kgs, range: 4–38 Kgs) who underwent VSD closure (n = 23) and repair of TOF (n = 17), partial AV canal (n = 4) and, cortriatriatum (n = 1). The skin incision was 2–3 inches long and sternum was split in its lower half with the manubrium intact. The thymus was excised in all. Only conventional instruments were used. Aorta, superior vena cava and, inferior vena cava were cannulated in all. Cardioplegic arrest was achieved antegradely. Retraction stitches facilitated access thereby eliminating the need for retraction by the assistant. Five underwent ligation of a patent ductus arteriosus. All VSDs were closed with Gore-Tex patch/pretreated pericardium. 21 patients required leaflet detachment of the tricuspid valve for closure of VSDs. Ten patients undergoing TOF repair required transannular patch. The left pleura was opened widely in all so as to give space to the heart. One infant (VSD) had myocardial edema and the split sternum was closed 12 hrs later. Perioperative details included: bypass time of 143 ± 53 minutes, cross clamp time of 84 ± 42 mins, median ventilation of 14 hrs (range 8–50), median ICU stay of 28 hrs (range 12–72 hrs). There were no hospital deaths. All the patients were discharged on day 7 with no wound infections or fractures. At 3–12 months follow-up, all are asymptomatic on no medications, with no hypertrophic scars. MS is safe and cosmetically appealing and free from wound infections. Our experience shows that MS can be safely applied for major lesions like VSD, TOF and AVSD and for very small infants (>2 months, >4 Kg) as well.

P262
QT dispersion in children with breath-holding spells

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Breath holding spells are common in infancy and early childhood and are frequently referred to pediatric cardiology clinics for exclusion of heart disease. Although these spells were previously considered as benign, recent data indicates late development of syncopal attacks, increased incidence of epilepsia and mental retardation during follow-up. Autonomic dysregulation, increased vagal stimulation leading to cardiac arrest and cerebral ischemia is considered as the cause of the clinical situation. Iron deficiency anemia is commonly associated with these spells. The aim of our study was to investigate QT dispersion for assessment of the risk of lethal dysrythmia and sudden death in this patient group. The study group consisted of 43 children (19 girls and 25 boys) between 3–108 months of age (mean ± SD = 22.7 ± 17.7 months); and the control group consisted of 25 children (12 girls and 13 boys) between 3–57 months of age (mean ± SD = 22.9 ± 15.1 months). Cardiac or neurologic disease were excluded in all patients. QT interval was measured; corrected QT interval (QTc), QT dispersion (QTd) and QTc dispersion (QTcd) were calculated using 12 lead surface electrocardiograms of the patients and the control group. There was no statistically significant difference in terms of QT and QTc intervals between the patient and control groups while QTd and QTcd values were significantly increased in patients with breath holding spells comparing the healthy children. QT dispersion was 59.5 ± 35.9 msn and 44.8 ± 11.9 msn in patients and control group respectively ($p < 0.05$). QTc dispersion was 102.1 ± 41.9 msn and 79.6 ± 24.6 in patients and control group respectively ($p < 0.01$). In conclusion, increased QT dispersion may be due to non-homogeneous repolarization of the ventricles in patients with breath holding spells. Further investigation for dysrhythmia and sudden death is needed in this patient group in order to clarify this relation.

P263
Peripheral pulmonary stenoses in twin-twin-transfusion recipients

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Development of right ventricular outflow tract obstruction at the infundibular and valvular level, as well as tricuspid regurgitation and cardiomegaly in the recipient twin in twin-twin-transfusion syndrome (TTTS) have been described. We describe 3 patients with significant peripheral pulmonary stenosis (PPS) in addition to the changes mentioned above:
Pat 1: Second twin, known TTTS recipient; laser coagulation at 17 wks GA. Fetal echo showed cardiomegaly and congestive heart failure. Elective C-section was performed at 30 wks. Echocardiography showed infundibular and pulmonary valve stenosis. During catheterization at 6 wks of age for balloon valvuloplasty hypoplastic left and right pulmonary arteries were found. The left pulmonary artery diameter (LPAD) and the RPAD were both 2.8 mm (calculated normal values 4.3 and 4.6 mm resp.). A second BVP was performed at age 5 mo which showed normal diameters. The other twin also had to undergo BVP for pulmonary valve stenosis without PPS. Both twins are being followed by our Cardiology Clinic and are doing well.
Case 2: Recipient second twin; fetal echo showed pericardial effusion; polyhydramnios required amniocentesis 8 times. Postnatally the patient developed resp. distress syndrome. Echo showed biventricular hypertrophy, pericardial effusion, and PPS. Cardiac catheterization at age 18 mo showed bilateral high grade PPS with suprasystemic pressures in the right ventricle and main pulmonary artery. The LPAD and RPAD were both 2.9 mm (calculated normal values 8.3 and 8.9 mm resp.). The A multislice-CT also showed high grade stenosis of the RPA and hypoplastic intrapulmonary vessels on the right as well as peripheral stenosis of the LPA with dilated intrapulmonary vessels and increased perfusion of the left lung. Because of inoperability the patient was started on Diltiazem and Lasix and is stable on that regimen.
Case 3: Second twin, C-section at 34 + 4 weeks, postpartal development of RDS. Echo showed a dysplastic and stenotic pulmonary valve with a gradient of 68 mmHg. BVP was performed at 2 weeks of age. The LPAD was 3.1 mm (calculated normal value 3.9 mm), the RPAD was 2.7 mm (calculated normal value 4.2 mm). Studies have shown an increased level of endothelin 1 in the recipient fetus in TTTS which may lead to vascular changes in the peripheral pulmonary vessels.

P264
Influence of pacemaker lead localisation on mean and long-term outcome in pediatric patients – a single center study on 111 patients

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Objective: To assess the influence of pacemaker lead localisation on mean and longterm outcome in pediatric patients we reevaluated all patients younger than 18 years who had a primary pacemaker (pm) implantation in our center between 1997 and 2001 with special emphasis on lead localisation.
Results: 111 patients were included in the study, 9 of whom died within the study period. Mean age at the first implantation was 6.8 ± 6.2 years, mean weight 23.4 ± 19.8 kg and mean

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length 108 ± 40 cm. Indication for pm implantation was complete heart block in 91%, sinus node dysfunction in 7% and long QT-syndrome in 2%. 64% of the patients had epicardial leads initially, 36% endocardial. The mean follow-up was 5.5 ± 5.1 yrs per patient. The total follow-up time was 611 patient-years. In the group with epicardial leads the pm had to be replaced mean 2.3 ± 1.4 times compared to 1.2 ± 1.4 in that with endocardial leads ($p = 0.004$). The interval up to the first pm replacement was 3.8 ± 2.8 yrs in epicardial and 5.4 ± 2.7 yrs in endocardial leads ($p = 0.004$). Mean 1.3 ± 0.7 lead revisions per patient were necessary in the epicardial, mean 1.2 ± 0.4 in the endocardial group ($p = 0.8$). 23 out of 71 (32%) epicardial and 6 out of 40 (15%) endocardial leads had to be replaced. Freedom from reintervention – pacemaker and/or lead revision – after 5 years was 38% in the epicardial and 70% in the endocardial group ($p = 0.001$).
Conclusion: In the mean and long term follow-up endocardial pm leads are superior to epicardial leads concerning the need of pm changes, the duration of the implanted leads and the number of lead revisions. The freedom from reintervention after 5 years is nearly twice as high in the endocardial compared to the epicardial group. Therefore whenever possible endocardial leads should be implanted even in the pediatric patient group.

P265
Effect of angiotensin converting enzyme inhibitors on exercise response of children with mitral insufficiency
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Background: Mitral insufficiency (MI) causes heart failure by volume overload, and an increase in atrial natriuretic peptide (ANP) levels due to stretch of atrium. Angiotensin converting enzyme inhibitors (ACEI) and ANP have similar effects like vasodilatation, natriuresis and diuresis. The aim of this study is to determine the exercise capacity in MI and response to ACEI treatment.
Subject-Methods: Patients with MI were divided into two groups: Digoxin group (DG)(10 F, 2 M; aged 10–18 years, mean 14 ± 0.72 years, under digoxin therapy for at least one year) and control group (CG) (8 F, 4 M; aged 8–17, mean 13.5 ± 0.81 years). None of the patients in each group had symptom of heart failure. ACEI treatment was given to DG and ANP levels, left ventricular systolic functions and exercise capacities were evaluated before and after enalapril therapy.
Results: ANP was significantly higher before enalapril both at rest and peak exercise in DG group.There were no significant differences between the groups after enalapril treatment. Left ventricular end-diastolic volumes of DG were significantly higher than the control group's values on admission. There were no significant differences between the DG and CG after enalapril. The ratio of reaching target heart rate in DG before and after enalapril were found 92.66 ± 1.00% and 97.33 ± 0.72% respectively ($p < 0.05$). There were no significant differences between the groups both on admission and after enalapril. Exercise duration time (EDT) of DG and CG were 8.95 ± 0.67 minutes and 11.02 ± 0.47 minutes on admission, respectively ($p < 0.05$), and it increased up to 10.12 ± 0.56 minutes in DG with enalapril ($p > 0.05$).
Conclusion: ANP is a good parameter to determine the silent heart failure, and it is recommended to monitor the ANP levels during the treatment of heart failure. Enalapril should take place at an early stage of heart failure in MI, because even if patients are on digoxin therapy, their heart failure may progress silently. Enalapril treatment increases exercise capacity in MI patients even they are on digoxin therapy.

P266
Left ventricular mechanics and tissue characterization in hypertensive children: what the earlier markers of heart damage?
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Aim of our study was to non-invasively assess LV mechanics and tissue characterization in hypertensive children in order to define the earlier markers of heart damage and the risk of cardiovascular morbility and mortality. We studied 21 untreated pts (aged 12.1 ± 6 yrs) with a recent diagnosis (<6 months) of high blood pressure at ambulatory blood pressure monitoring. All pts underwent an echocardiographic evaluation. Sex- and age-specific cut-off levels for LV mass/height 2.7 (LVMI) and relative wall thickness (RWT) were defined to assess LV geometry, as normal, concentric remodeling (CR), concentric hypertrophy(CH), eccentric hypertrophy (EH). As load-independent index of myocardial contractility, the relation between the midwall rate-corrected velocity of circumferential fiber shortening (mwVCFc) and meridional end-systolic stress(es) was defined. LV diastolic function was evaluated by the mitral flow indexes [peak E, peak A, E/A ratio, deceleration time (DT)] and isovolumic relaxation time (IVRT). Ultrasonic tissue characterization of the LV myocardium was performed by integrated backscatter (IBS) calculating the magnitude of cyclic variation (CV), which reflects the contractile function, and the averaged myocardial intensity normalized to pericardium, which is directly related to the myocardial collagen content. In addition 35 age-BSA-matched normal subjects were used as control group.
Results: LV geometry was abnormal in 5/21 pts (2 CR, 1 CH, 2 EH) (23%). LVMI, significantly correlated with mean 24-hours systolic pressure ($r = 0.46$; $p < 0.05$), was greater than 99th percentile in 4/21 (19%) and >51 g/m 2.7 in 2/21 (9.5%) pts. The midwall VCFc-es relation was normal in all pts. Based on 95% CI generated from normal controls our study population showed prolonged IVRT in 12/21(57%) pts and DT in 11/21 (52%) pts, incresad peak A in 14/21 (66%), decreased E/A ratio in 12/21 (57%) pts. Finally, both at IVS and PW, comparable values of CV and averaged myocardial intensity were detected.
Conclusions: An early echocardiogrphic evaluation of LV diastolic function and geometry allows us to identify a subset of hypertensive children with heart damage and increased risk of cardiovascular morbility.

P267
Left ventricular rotation and torsion in Situs Inversus Totalis are not the inverse of the ones in Situs Solitus
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Introduction: Due to the specific helical pattern of the myofibers in the left ventricular (LV) wall, it is found, both in model studies and experimental studies on humans and dogs, that myocardial fiber shortening is about evenly distributed over the LV wall and that LV torsion (axial gradient in rotation) is nearly uniform along the LV long axis. Hypothesis: We expected that in Situs Inversus Totalis (SIT) the same, though inverse, deformation pattern as in Situs Solitus (SS) could be found due to mirror image organ position, mirror image cardiac looping, and mirror image cardiac fiber orientation.

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Methods: healthy children and adults with SS or SIT were studied using Magnetic Resonance (MR) Imaging Tagging. LV rotation was estimated from MR tag displacements in five short-axis slices. Torsion (T) was calculated using data on rotation (rot) and midwall radius (r) from 2 adjacent slices according to: T = (rot_u − rot_l)/d * (r_u + r_l)/2 in which u and l refer to the upper and lower slice, respectively, and d denotes the distance between the 2 slices.
Results: In SIT LV systolic rotation for all five slices had an inverse pattern compared to SS. However, instead of showing a gradual increase in rotation from base to apex, rotation showed an increase from base to equator, and a decrease from equator to apex. As a result, apical and basal torsion were opposite, with apical torsion as in SS and basal torsion inverse as compared to SS.
Discussion: The deformation pattern in SIT can be explained using data from a histological study (Kaibogaku Zasshi 1989; 64(1): 36–45) showing that apical and superficial basal fiber orientation follow the pattern of the heart in SS, whereas in the deeper basal layers fiber orientation is inversed.
Conclusions: Cardiac myofiber structure likely develops from two sites, i.e. apex and base. In the normal heart, the resulting helical structures are synergistic, resulting in the common myofiber pattern. In SIT, normal fiber structure near the apex meets inverse symmetry near the base at the equator. Therefore, in SIT torsion near the apex is as in Situs Solitus, whereas torsion near the base is inversed.

P268

Systemic ventricular rotation and torsion are independent of the morphological type of the ventricle

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Introduction: The left ventricular (LV) and the right ventricular (RV) wall have about the similar helical pattern of myofibers. Transmural helix angle courses typically range from +60° at the subendocardium to −60° at the subepicardium. In healthy humans with systemic ventricles of left ventricular morphology (normal situation) it is found, due to the specific myofiber pattern, that myocardial fiber shortening is about evenly distributed over the LV wall and that LV torsion (axial gradient in rotation) is nearly uniform along the LV long axis.
Hypothesis: We expected that systemic ventricular rotation and torsion in persons with both atrio-ventricular and ventriculo-arterial discordance (cc-TGA) would be the same as in normal persons, since the myofiber structure is not really altered by the ventricular morphology.
Methods: 2 adults with cc-TGA and no other cardiac abnormality were studied using Magnetic Resonance (MR) Imaging Tagging. Systemic ventricular rotation was estimated from MR tag displacements in five short-axis slices. Torsion (T) was calculated using data on rotation (rot) and midwall radius (r) from 2 adjacent slices according to: T = (rot_u − rot_l)/d * (r_u + r_l)/2 in which u and l refer to the upper and lower slice, respectively, and d denotes the distance between the 2 slices.
Results: Systemic ventricular rotation for all five slices and torsion patterns between slices 1–2, 2–3, 3–4, and 4–5 were similar to the ones in normal persons. All slices showed counterclockwise rotation (as viewed from the apex) during the first 150 ms of the systole. Thereafter, they started to rotate in clockwise direction. The amount of rotation increased from the base to the apex. As a result, torsion, being the axial gradient in rotation, was about uniform for all 4 sections during systole.
Conclusions: Systemic ventricular rotation and torsion are independent of the morphological type of the ventricle. The axiom in pediatric cardiology that the right ventricle is not apt to act as a systemic ventricle might be true, but not because of differences in microscopic wall characteristics. More likely macroscopic differences are responsible, e.g. chordal attachments of the systemic atrio-ventricular valve to the inlet septum giving easily rise to valve incompetence.

P269

A new echo technique in management of paediatric patients

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Background: An innovative tool a portable echocardiographic system is one of the latest achievements in cardiac evaluation. This ultrasound stethoscope has not been widely used in paediatric population. The aim of this study was to investigate the utility of a new portable echocardiography machine (ACUSON-Cypress/Siemens) as a bedside screening system.
Methods: We studied 30 pts (17 female, mean age 5 years) referred for cardiac evaluation at Cardiorheumatology out-pts department. All pts underwent clinical evaluation, ECG and X ray, miniaturised echocardiographic examination (US) with ACUSON and subsequent complete 2D examination (2DE) performed by a blinded and independent observer as a reference. The portable instrument used was ACUSON Cypress/Siemens, weighting 2.4 kg, measuring 21 cm × 40 cm × 35 cm with Color Flow Doppler.
Results: Good images were obtained in 28/30 pts (92%). When a comparison of 2DE was made, no difference were found in terms of pericardial effusion (5/5), intracardiac mass (1/1), valves regurgitation (8/8) and complex congenital heart anomalies (6/6), as well as in evaluation of LV function. Inter-observer variability on ACUSON was excellent: correlation coefficient of 0.92, mean difference 2%, 95% limits of agreement −12 to + 8%, kappa value of agreement 0.7.
Conclusion: This portable ultrasound machine is a feasible device, allows accurate assessment of both LV systolic function and Color flow with excellent interobserver agreement. It can be effectively applied for detailed evaluation of cardiac morphogenesis and haemodinamical data in paediatric population.

P270 (see Abstract 105)

Pediatric cardiac surgery risk stratification: The pediatric cardiac care consortium (PCCC) categories reflect mortality and length of stay in a large German unit

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P271

Surgical repair of ALCAPA: Long-term follow-up with emphasis on the mitral valve

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Background: The appropriate approach for the anamalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is immediate surgical correction with restoration of a 2-coronary circulation. However there is still some concern in regard to the

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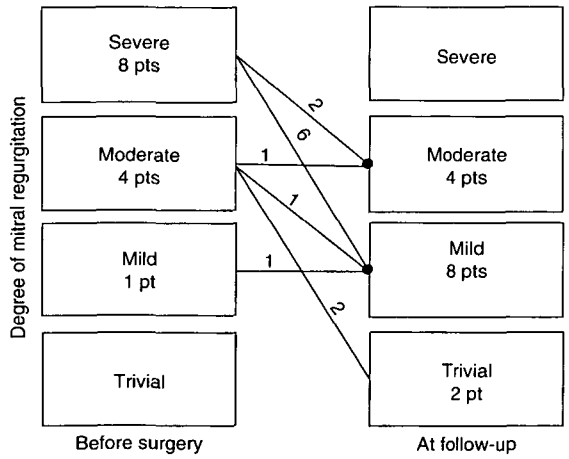
potential for coronary insufficiency and for the evolution of mitral regurgitation (MR) in the long-term follow-up.
Methods: 13 consecutive patients (7M/6F) underwent a 2-coronary repair of ALCAPA at a mean age of 4.1 years (range 1.3 months–14 years). Nine pts underwent aortic coronary reimplantation with punch tecnique under hypotermic cardiopulmonary bypass while 4 pts underwent terminal anastomosis between left subclavian artery and left main coronary artery. Additional procedures were: pulmonary transcatheter valvuloplasty at 23 months of age in one pt and mitral chardae shortening for severe non-ischemic anterior leaflet prolapse in another one. In 12 patients reparative surgery on the mitral valve was not undertaken, even when a severe preoperative MR was present. After repair no pt needed mechanical circulatory support.
Results: There have been no early or late deaths after a mean follow-up of 7.9 ± 6.7 years. Left ventricular ejection fraction markedly improved after 1 week from repair (37.7 ± 12.7% vs. 49.6 ± 10.1%, $p = 0.0006$) with a further improvement in the long-term (49.6 ± 10.1% vs 64.7 ± 7.7%, $p < 0.0001$). MR degree showed a significant decrease in the follow-up, with no patient presenting severe MR (Figure). Coronary angiography was performed in 12/13 pts after 2 to 4 years from surgery and showed no coronary stenosis or occlusion. Myocardial Tc99-perfusion scans performed in all patients in the follow-up showed small anteroseptal or anterolateral perfusion defects in 4 patients with recent normal coronary angiograms. Additional procedures in the follow-up were: an aortic valve replacement for severe aortic regurgitation at 14 years of age and a pulmonary baloon valvuloplasty for severe supravalvular stenosis at 18 months from surgical repair.



Conclusion: Although prompt surgical intervention yield excellent results and gradual myocardial recovery, the possibility of recurrent subclinical ischemia remains, thus justifying regular postoperative follow-up. As mitral regurgitation showed a significant decrease in the follow-up, in our experience, mitral valve repair should not be performed in all patients at the time of coronary repair.

P272
Outcome in fetuses with abnormal viscero-atrial situs

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Background: Fetal cases with abnormal viscero-atrial situs present a serious problem for a correct prenatal counselling; therefore we wanted to analyse retrospectively the features and outcome of our cases.
Material and Methods: Out of 670 cases (c.) diagnosed in our Centre to have congenital heart disease (CHD) by fetal echocardiography between January 1985 and August 2002, 41 (6.1%) were found to have an abnormal viscero-atrial situs (AVAS). Their data – type of CHD and outcome in utero and after birth were retrospectively analysed. Fetal diagnosis was always compared with postnatal or postmortem diagnosis.
Results: 10 c. had situs viscerum solitus (SVS) with destrocardia, 12 had situs viscerum inversus (SVI) and dextrocardia, 4 had SVI and levocardia, 8 had left isomerism (Lis) and 7 right isomerism (Ris). 8 cases had isolated AVAS and normal cardiac anatomy (1 with extracardiac anomaly), 16 had complex atrioventricular defect (mostly associated to Lis and Ris), 6 had univentricular heart, 3 – double outlet right ventricle, 1-double outlet left ventricle, 1 truncus, 2 siblings had pulmonary atresia and hypoplastic right ventricle, 3 had single or multiple VSD and one had corrected transposition. Four fetuses with Lis had associated total atrioventricular block (AVB). There were 2 cases with chromosomal and 3 with extracardiac anomalies.
Outcome: 13 opted for termination of pregnancy, of 28 cases that continued pregnancy, 2 died in utero, 11 (39.3%) died after birth – 3 postoperatively and 15 survived (53.6%). Mortality was higher in Lis – in 5/7 (71%) of cases that continued pregnancy, mainly due to associated AVB.
Conclusions: AVAS was mostly associated to complex CHD and the outcome in such cases was poor, (mainly in those with Lis and AVB). No impact on the outcome was found in cases with structurally normal heart.

P273
Thirty-day mortality in pediatric cardiac surgery after centralization in Sweden 1994–2001

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Background: Concentration of pediatric cardiac surgery from four to two centres was achieved in Sweden almost 10 years ago. The process of centralization started in 1993 with 95% of the surgery having been performed in two centres until 1998 and thereafter all surgery being concentrated to these two centres. We report the results of all pediatric cardiac surgical procedures in Sweden during the time period 1994–2001.
Materials and Methods: All open and closed surgical procedures were registered and classified regarding age at surgery and 30-day mortality. Open heart surgery was categorised according to three complexity levels, grade III representing the most complex surgery such as for example the Norwood operation, total cavopulmonary anastomosis and open heart surgery in premature babies (for details see Lundström et al., Pediatric Cardiology 2000; 21: 353–357).
Results: The total number of procedures was 4737 with a decrease per year from 709 procedures in 1994 to 476 in 2001. The overall 30-day mortality was 1.8% (86 patients). Twelve patients out of 1094 (1.1%) died after having had a closed surgical procedure and 74 patients of 3643 (2%) died after open heart surgery. In open heart surgery mortality decreased from 3.6% in 1994 to 0.8% in 2001 having been less than 2% during the last three years of the study period. The proportion of complex surgery (grade III) increased from 38% to 45% during the study period, as did the proportion of open heart surgery in infants <1 year which increased from 45% to 60% in later years.

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Conclusion: The decrease of the number of surgical procedures is probably explained by a significant decrease of the birth rate in Sweden during the study period, and also by an increase of catheter interventions in congenital heart disease (approximately 150 per year in later years). The low mortality in later years is remarkable bearing in mind that the proportion of complex surgery and open heart surgery in infants less than one year of age increased significantly. The centralization of pediatric cardiac surgery has been a prerequisite for the development and performance of the most complex surgical procedures in Sweden.

P274 (see Abstract 92)
Percutaneous Pulmonary Valve Implantation for right ventricular outflow tract lesions after congenital heart surgery

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P275
Five-year follow up after pedatric cardiac surgery in 1996 in Sweden

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Background: The much improved short term results in pediatric cardiac surgery in later years emphasise the need to focus also on long term results. Complete and nation-wide long-term follow up studies after surgical treatment of congenital heart defects reduce the risks of confounding selection bias. The concentration of pediatric cardiac surgery in Sweden from four to two centres since 1993 greatly facilitates long term follow-up studies to be performed.
Materials and Methods: Survival of 575 children under the age of 18 years who in 1996 underwent cardiac surgery in Sweden was studied using surgery and cardiology registries in the two centres. Survival of all patients were also cross-checked against the death registry of Swedish National Board of Health. Causes of deaths were analysed through hospital notes and by autopsy reports. An additional 41 surgical procedures were performed in a third institution but were not included.
Results: The 575 patients had surgery at a median age of 0.88 years (0 days–17.1 years). 541 patients were alive at five year after surgery (five-year survival 94%). Ten patients out of the 34 who died did so within 30-days after surgery, 19 between one and 12 months after surgery and five later than a year after surgery. Subsequent surgery was performed in 18/34 patients who died and 11 of them died within 30 days after the subsequent surgical procedure. Eighteen of the deaths occurred in children who had surgery for univentricular hearts (nine after Norwood surgery). Five infants with modified Blalock-Taussig shunts died. Eight of the children who died had syndromes and/or chromosomal aberrations (Down syndrome in four). Two non-cardiac deaths occurred, one due to gastroenteritis with necrotizing enterocolitis and one due to an intracerebral hemorrhage (patient on coumadine treatment). One death due to chronic rejection of a cardiac transplant occurred.
Conclusion: The five year survival of 94% establishes a fairly high standard, although comparison with results from other countries is difficult due to lack of information. Most deaths occurred later than 30 days after the surgical procedure, which emphasises the need for continuous long-term recording and analysis.

P276
Gadolinium filter for pediatric cardiac catheterisation: a new safe tecnique

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Despite echocardiography, still many pediatric patients (pt) with congenital heart disease have to be submitted to haemodynamic cardiac catheterisation in first years of life. At this ages the radiation risk is particularly relevant owing to high tissue radiosensitivity, large percentage of body tissue involved by radiation and possible repeated procedures. We investigated the in vivo effectiveness of using a gadolinium filter for cardiac angiography of paediatric patients (0–6 years) to reduce the patients' exposure while maintaining image quality. During 10 interventional procedures data were acquired using an Exposure Area Product (E.A.P.) meter and by filtering the incident X-ray beam with a foil of 0.1 mm of gadolinium. We compared the images obtained on the same pt with and without filter, while effective dose per frame estimates were calculated from EAP measurements. When the filter was employed, the mean radiological doses decreased from 10% to 20.3% in the individual pt, mean of 14.3%, without any observable difference in image quality. In conclusion, gadolinium filter can improve the iodated contrast medium visualisation, maintaining the same diagnostic image quality but significantly reducing radiological exposure to paediatric pt.

P277
Fate of homograft conduits after surgery for congenital heart defects in children

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Background: Cryopreserved homografts are routinely used at corrective surgery for many congenital heart defects, most often in the subpulmonary position. Homograft failure due to stenosis or insufficiency (or both) sooner or later necessitates replacement. We studied the fate of homografts inserted in patients less than 18 years of age in our institution.
Materials and Methods: Retrospective analysis of patients <18 years, who had an homograft inserted in our institution during 1992–1996 (study endpoint was May 1st 2002). Parameters studied were types of congenital heart defects, age at primary insertion of the homograft as well at replacement operations, types of surgical procedures, mortality, type of homograft etc.
Results: Sixty patients had their first insertion of an homograft in 1992–1996. Fifty-seven patients had the homograft inserted in the subpulmonary position, of whom 20 underwent Ross surgery. Three patients had the homograft implanted in the aortic position. Death occurred in 10 patients (16,7%) of whom five died within 30 days of the operation and five thereafter. Homograft function was satisfactory in 72% of the patients five years after homograft insertion. In total 11 homografts were replaced during the study period at a median time after the homograft insertion of 3.2 years (1.7 months–8.9 years). Patients with truncus arteriosus

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were most common among those who needed replacement surgery (six out of seven patients). Homograft function at five year after the surgery was satisfactory in 42% of patients who had the homograft insertion before the age of one year but in all who were older than one year at the time of homograft insertion ($p < 0.001$). No child died at homograft replacement.
Conclusion: Age at homograft insertion is a risk factor for early homograft failure, with infants younger than one year at the insertion being clearly at risk for needing early replacement. Patients with truncus arteriosus were common among those who needed replacement surgery. Deaths were not related to homograft replacement.

P278
Left ventricular torsion is nearly uniform along the left ventricular long axis in healthy children
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Introduction: Within the left ventricular (LV) cardiac wall muscle fibers are oriented in a characteristic helixlike pattern. Measured transmural helix angle courses typically range from $+60°$ at the subendocardium to $-60°$ at the subepicardium. Due to the specific helical pattern of the myofibers, it is to be expected that the LV will exhibit torsion (axial gradient in rotation) during the ejection phase. We wanted to estimate LV short-axis rotation and torsion patterns at different levels between the base and apex.
Methods: 12 healthy children were studied using Magnetic Resonance (MR) Imaging Tagging. LV rotation was estimated from MR tag displacements in five short-axis slices. Torsion (T) was calculated using data on rotation (rot) and midwall radius (r) from 2 adjacent slices according to: $T = (rot\_u - rot\_l)/d \star (r\_u + r\_l)/2$ in which u and l refer to the upper and lower slice, respectively, and d denotes the distance between the 2 slices.
Results: All 5 LV short axis slices showed counterclockwise rotation (as viewed from the apex) during the first 100–150 ms of the systole. Thereafter, they started to rotate in clockwise direction. The amount of systolic rotation increased from the base to the apex. As a result, torsion, being the axial gradient in rotation, was about uniform for all 4 sections during systole.
Conclusions: LV rotation is gradually increasing from base to apex, and, hence, LV torsion (axial gradient in rotation) is nearly uniform along the LV long axis in healthy children.

P279
Interrelationship of left ventricular wall motion, pulmonary venous flow, and transmitral flow in healthy children
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Aim: To assess the myocardial velocities of the mitral annulus, left ventricular posterior wall and midseptum in healthy children, to determine the reference values of myocardial wall motion and to compare these parameters with transmitral and pulmonary venous velocities.
Methods and Results: Seventy-two children (36 females/36 males) who had no systemic or cardiac pathologies were examined. The mean age of the study group was 6.73 ∓ 61617; 5.10 years (0.1–17.75 years, median: 6.71 years). Each parameter was measured twice in end inspirium and in end expirium. The tissue velocities were similar in males and females ($p > 0.05$). The longitudinal velocity of the heart in early diastole (E) had positive correlation with age ($p < 0.05$; midseptum velocities $r = 0.57$, posterior wall velocities $r = 0.56$, mitral annulus $r = 0.56$). The tissue velocities were not influenced by respiration ($p > 0.05$). The myocardial velocities of different segments of the left ventricle are not correlated with the transmitral or pulmonary venous flows in children ($p > 0.05$).
Conclusion: This study, while determining the reference values of tissue velocities in a large healthy pediatric group, will pioneer new studies concerning pediatric patients.

P280
A prospective single center study on percutaneous ASD closure with the Helex Septal Occluder
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Background: Over the past decades catheter closure of secundum ASD has become a standard procedure in interventional cardiology. The Helex Septal Occluder is device which has been used frequently in children but rarely in adults. We report our experience in ASD closure with the Helex device.
Methods: Since 1992 transcatheter closure of interatrial communications has been performed in our center in 1184 patients. In 36 selected patients with a secundum ASD we implanted the Helex occluder (mean age 49 ± 18 years, range 15–74). The device was used only in smaller defects (stretched diameter ranged from 7 to 20 mm, mean 15 ± 3). 3 patients had no anterior septal rim.
Results: The implantation was successful in all 36 patients. All procedures could be performed without general anesthesia. The mean procedure time was 45 ± 16 minutes (mean X-ray time 9 ± 5 minutes). Patients could be discharged after 0.9 ± 0.4 days. The Qp:Qs ratio significantly decreased from 1.5 ± 0.3 before closure to 1.0 ± 0.1 after 6 months ($p < 0.01$). The mean pulmonary artery pressure was normal before closure (18 ± 6 mmHg) and remained unchanged after 6 months (17 ± 5 mmHg). The RVED diameter decreased from 36 ± 7 mm to 35 ± 5 mm. Immediately after implantation 27/36 patients had a complete closure (2 patients with residual shunt had a multiperforated septum). After 6 months 32/36 patients had complete closure, 4 had a small residual shunt (RS) measured by TEE doppler. 1 RS occluded spontaneously 1 year after implantation, 3 persisted (no hemodynamic relevance, $Qp/Qs < 1.1$) No complications occurred during hospital stay. During follow up (0.5–29 months, mean 11 ± 8) a small thrombus on the device was diagnosed in one patient (asymptomatic, resolved under heparin). One patient showed an asymptomatic device wire fracture (no residual shunt) and one patient suffered from atrial fibrillation (cardioversion with sotalol successful).
Conclusions: Catheter ASD closure with the Helex Occluder is a very safe procedure, if the patients are selected carefully. Residual shunts occur occasionally but they are usually small and not hemodynamically relevant. This device is a good alternative to other devices in smaller ASD's.

P281 (see Abstract 121)
Cardiac disturbances in insulin-dependent diabetes mellitus in child
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P282

Electrocardiographic and echocardiographic modifications occurred during the hemodialysis session in children with chronic renal failure

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The Aim of the Study: was to find out the main electrocardiographic and echocardiographic modifications occurred after the hemodialysis session in children with chronic renal failure (CRF).
Methods:
Patients: 17 children with CRF included in a hemodialysis program, aged between 5 and 18 years. In all patients we performed immediately before and 4 hours after the hemodialysis: Electrocardiography (ECG); Echocardiography (Echo), in order to asses the systolic function of the left ventricle(LV).
Results: The ECG modifications observed in our patients were especially the modifications of the QT interval segments: insignificant shortening of the Q-aTc, significant shortening of the Q-oTc and significant increase of the Q-eTc interval (from 0.37 ± 0.036 to 0.42 ± 0.039 ms, $p < 0.001$), well correlated with the decrease of the potassium level. The Echo modifications observed in our patients were : a significant decrease of the right ventricle (RV) diameter ($p < 0.05$) and of the end-systolic ($p < 0,001$) and end-diastolic ($p < 0.05$) LV diameter; a mild increase of the ejection (from $56.8 \pm 14.51\%$ to $65.11 \pm 14.09\%$) and shortening (from $25.22 \pm 9.60\%$ to $30.73 \pm 9.58\%$) fractions, without significant modifications of the LV wall thickness.
Conclusions: The hemodialysis session induces : the improvement of the main parameters of the systolic function of LV and the decrease of the RV dimensions; the improvement of the varied segments of the QT interval, whose increase occurred in CRF represents an enhancing factor for the risk of cardiac arrhythmias. The ECG and Echo follow up is very useful in order to prove the efficiency of the hemodialysis session in improving the systolic function of the LV and in reducing the risk of cardiac arrhythmias induced by the electrolytic disequilibrium and showed by the QT interval modifications.

P283

Left ventricular myocardial function in congenital valvar aortic stenosis assessed by ultrasound tissue velocity and strain rate techniques

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A pilot study was performed to reveal the potentials of new echo/Doppler techniques for the detection of myocardial changes due to congenital valvar aortic stenosis. Twenty-four patients (age range 0.1–17 years), with various degrees of aortic stenosis and 24 age- and gender-matched, healthy children were enrolled in this study. Conventional echo-Doppler, tissue velocity- and strain rate measurements were carried out using the apical four-chamber view and transthoracic long axis view. All patients had normal fractional shortening of the left ventricle (>28%). Although the sum of septal and ventricular wall thicknesses was significantly increased in the patients' group ($p < 0.001$), only six of the 24 patients showed left ventricular hypertrophy. In tissue velocity mode, systolic and early diastolic wall velocity acceleration was significantly reduced in both views. Peak systolic and early diastolic wall velocities as well as strain rate values in the four chamber view were significantly reduced in the patient group. The decrease was highest for the strain rate values in all cases. In conclusion, strain rate values at different moments within the heart cycle might become important parameters in the assessment of myocardial impairment. Further studies are indicated to assess the correlation of these parameters with the severity of stenosis, left ventricular hypertrophy and irreversible myocardial function changes.

P284

Long-term follow up of balloon angioplasty for native aortic coarctation in neonates

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Background: The role of percutaneous transluminal balloon angioplasty (PTBA) for treatment of native aortic coarctation (CoAo) in neonates is controversial as it produces a rapid decrease of isthmic gradient and improves symptoms of congestive heart failure but on the other hand carries an high early recoarctation rate and vascular complications.
Methods: We reviewed clinical and haemodinamic data of 81 consecutive neonates 1 to 90 days old (mean 23 median 14 days) undergoing PTBA in our hospital between 1984 and 2001. Late follow up (range 12 to 213 months mean 107) data were avaliable in 68 patients (pts). CoAo was isolated in 44 neonates (54%) while in 37 it was associated to others cardiac defects (46%). Median weight of pts at procedure was 3.2 Kg (range 1.2 to 5.4 Kg). The vascular approach used was femoral vein in 6 pts, femoral artery in 36 pts and more recently right axillary artery in 39 pts.
Results: Short term success of PTBA was achieved in 71 pts (88%), 8 neonates were surgically treated and 1 underwent second PTBA during first stay for inefficacy of PTBA or early restenosis. 1 neonate died during the procedure for iliac artery damage. At late follow up 38 pts (56%) are clinically stable with only transcatheter treatment (1 to 3 PTBA mean 1.7). 30 pts required surgery 2 at 148 months after PTBA. MRI or angiography was performed in 50 pts at least 2 years after PTBA and only 2 small aneurysms were detected. Obstruction of the femoral artery was detected in 23 pts with transfemoral approach (65%) while vascular complications were noted in only 2 pts with axillary approach (5%).
Conclusions: PTBA of native coarctation is a safe and effective therapeutical option in selected neonates. High incidence of restenosis is present but the result of a second PTBA is more stable. Vascular complications are reduced in our experience with right axillary approach and use of low profile catheters. PTBA may delay the timing of surgical correction when the neonate reaches a greater weight and a more stable clinical condition.

P285

Pulmonary arteries underdevelopment – diagnostics and treatment in children with HLHS after Norwood procedure

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Background: Operative treatment in children with HLHS still remains a multistage and high risk procedure. Therefore the possibility of percutaneous treatment of its complications is very important as it allows avoiding additional surgical procedure and leads to increase of time intervals between consecutive stages of surgical treatment.

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Material and Methods: Between 1996–2002 34 children (age 1.5–10.5, mean 6 months; BSA 0.22–0.38, mean 0.32) with HLHS after first stage Norwood procedure underwent invasive cardiac diagnostics. 17 patients underwent cardiac catheterization with selective angiography, 17 – retrograde interarterial angiography.
Results: LPA diameter ranged from 2.9 to 14, mean 6.14 mm. In 8 children we observed severe LPA hypoplasia. 1 patient in this group was operated using classic Norwood procedure (method I), 5 patients were operated using modified Norwood procedure (method II) and in 2 patients pulmonary artery was anastomosed with free wall of right ventricle (method III). RPA diameter ranged from 4.8 to 11 mm, mean 6.96. In 6 patients we found RPA hypoplasia, one of them was operated using method II, 5 – method III. In 4 children we found hypoplasia of both pulmonary arteries (3 operated using method II, 1 – method III). Only one patient required balloon angioplasty of coarctation of the aorta. In 9 children after second stage of surgical treatment (bidirectional Glenn procedure) we performed next cardiac catheterization. LPA hypoplasia was found in 8 of them. In 5 children in this group LPA balonoplasty was performed. In 1 patient we succeeded to obtain permanent increase of LPA diameter, 2 required stent implantation 4 and 9 months after balonoplasty. In 7 patients stent implantation in LPA was performed with good effect. 3 patients from interventionally treated group are nowadays after third stage of surgical treatment – Fontan's procedure.
Conclusions: In majority of patients with HLHS after stage one Norwood procedure we observed stenosis or hypoplasia of one or both pulmonary arteries. Frequency of pulmonary arteries underdevelopment was independent on Norwood procedure technique. Percutaneous treatment such as balonoplasty and stent implantation is a method of choice in pulmonary arteries stenosis or hypoplasia in children with HLHS between stages of operative treatment.

P286
A novel device to prevent stroke in patients with atrial fibrillation

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Background: In elderly patients up to 1/3 of all strokes are attributable to atrial fibrillation. Since the left atrial appendage is the source of thrombus formation in these patients, its occlusion may become an effective treatment in patients who are suboptimal candidates for life-long anticoagulation. Basic principals of this new technique are similar to other interventional procedures in paediatric cardiology. Therefore, interventional paediatric cardiologists will become involved.
Methods: The device consists of a self-expanding Nitinol-cage (ranging from 15 mm to 32 mm in diameter) which is covered with ePTFE. Small anchors along the struts help stabilizing the occluder in the LAA. It is delivered by a specially-designed 12 Fr transseptal sheath. LAA occlusion was attempted in 68 patients (54–85 years, 71 ± 8 years) who had atrial fibrillation and were suboptimal candidates for warfarin therapy. These patients were followed by flouroscopy, echocardiography and chest-x-ray.
Results: Device implantation (mean procedure time = 74 ± 30 min) was successful in 67/68 patients. Due to a hematoma during groin access the procedure was interrupted in one patient. Five patients developed pericardial effusion. In three of them pericardiocentesis was performed without further sequelae. Device exchanges to optimal implant size were performed in 10 patients. Follow-up ranges from 2 to 16 months (9 ± 4 months, 50 patient years). Since the implantation no further stroke has occurred.
Conclusion: Percutaneous closure of the LAA with this new device for prevention of embolism in patients with atrial fibrillation is feasible and safe. This new technology may become an alternative in patients who are not suitable for anticoagulation therapy.

P287
Impaired development of the right mamma after right anterolateral thoracotomy in pre-pubescent female patients

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Background: It is presumed that a right anterolateral thoracotomy (ALA) reaches more favorable cosmetic results than a median sternotomy (MS). We investigated a subgroup of pre-pubescent female children under 12 years of age at time of operation, with regard to breast development and scoliosis in adolescence. To our knowledge, this study represents the largest reported cohort at long-term.
Methods: Between 1974–1984, a total of 599 patients (pts.) had atrial septal defect (ASD) closure at our institution. 184 pts. were pre-pubescent females. 95 pts. (72 ALA, 23 MS) were contacted to our questionnaire analysis. Of these, 61 pts. (46 ALA, 15 with MS) were examined at our institution, including ECG and echocardiography. Degree of scoliosis and breast volume difference were measured by a three dimensional surface and spine scanning apparatus ("Formetric II", Diers Ò). Using photographs of the upper chest, breast symmetry was described by an index. Mean follow-up time was 23 years.
Results: Due to questionnaire analysis, impairment of self-esteem in regards to the cosmetic outcome was significantly lower in the ALA group ($p = 0.036$). Accordingly, 76% (ALA) vs. 39% (MS) considered the cosmetic result excellent ($p = 0.008$). Still, upon our calculated index, severe asymmetry, with a smaller right breast, occured in 61% of the ALA group. Breast volume difference >20% occured in 55% of the ALA group. In the MS group, no apparent asymmetry was detected ($p < 0.001$). Orthopedic examination showed no difference in incidence of scoliosis compared between ALA and MS group and compared to normal population. None of the pts. had residual ASD.
Conclusions: Although ALA in pre-pubescent female patients affects breast development significantly, most women do prefer it to a scar following MS. To our mind, a standard median sternotomy can not be justified for correction of ASD related complexes. As an alternative to ALA other less invasive approaches should be considered, such as right posterolateral thoracotomy, subxyphoid approach, partial sternotomy.

P288
CardioPat, a computer programme, uses a medical matrix as a new approach to the input of data in a clinical environment

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Purpose: In a clinical environment storing medical data is not only a legal obligation, it is also essential to daily clinical work and

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scientific work. A well organized computer network with a central database represents a good infrastructure for retrieving data, but entering data into a computer remains a problem. CardioPat represents a new approach to the input of data. Its main instrument is a medical matrix developed by the author. Given a key diagnosis, it opens a network of related words. This facilitates the input of further data.
Methods: In 6 years of research and programming, the author developed a computer application, CardioPat, that allows the input of almost all medical data: patient's history, findings, diagnoses, examination results (ecg, echocardiography, ultrasound, x-ray etc.), daily notes and other items. In contrast to the conventional approach, CardioPat tries to collect different data of the patient simultaneously, thus achieving a more efficient input of data. For example, if the diagnosis for a patient were aortic stenosis, CardioPat would display items that relate to this diagnosis. As soon as the user opens the entry form for echocardiography he would get, along with standard items, a list of items that relate to aortic stenosis: diameter of aortic valvar ring, number and shape of aortic valvar cusps etc. The advantage of a medical matrix is the connection and cross-connection of medical data. The computer keeps a data dictionary, in this case for paediatric cardiology, and can present the entry form according to the given data. The user may of course decide to add other items to the entry form. CardioPat offers a dynamic system for data entry. The programme is written in Borland Delphi and uses an Oracle database system. The data dictionary holds more then 600 "medical items". CardioPat can be used in German or English. Operating system: Microsoft Windows '95, '98, NT4, 2000 or XP.
Conclusion: The introduction of a data entry system (CardioPat) improves and accelerates the acquisition of daily clinical data. It also permits retrieving data quickly and simply, for example for scientific or statistical evaluation.

P289
Paediatric cardiology and telemedicine: two year's experience of cooperation with remote hospitals
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A program of telemedicine was initiated in November 2000 between the Paediatric Cardiology Department of a tertiary care hospital in Lisbon and paediatricians and obstetricians from three remote Portuguese hospitals: two in continental Portugal and one in the Azores Islands. The images were transmitted by a telemedicine link across three integrated service digital network (ISDN) lines to a regional paediatric cardiology unit for interpretation by a consultant paediatric cardiologist. Over 25 months, a total of 192 real-time teleconsultations in 175 patients were carried out, concerning 92 fetal exams (48%), 68 evaluations in newborns (35%) and 32 children (17%). 30 of the total (35%) were urgent transmissions. There were 76 positive diagnosis (43%) in 50 newborns, 16 children and 10 foetuses. Structural congenital heart disease was the most frequent diagnosis, observed in 58 patients, including 7 complex anomalies (12%). In all of these complex anomalies, the segmental arrangement and the main diagnosis were correctly assessed by telemedicine. In only one case, with a telemedicine diagnosis of atrial septal defect, this was not subsequently confirmed by the paediatric cardiology. Thirteen patients required urgent transfer to Lisbon, while a medical team from the tertiary hospital travelled to the local hospitals and performed a surgical treatment of large patent ductus arteriosus in three premature newborns and a percutaneous atrioseptostomy in one newborn with transposition of the great arteries and severe dessaturation unresponsive to prostin, avoiding the transport of unstable patients. The other patients were referred for follow-up in peripheral clinics. In our experience, real-time telemedicine with online echocardiography, conducted by a paediatric cardiologist, is an important tool in the diagnosis or exclusion of paediatric cardiovascular diseases. It has an important role in continuing medical education of staff from local hospitals, specially in the field of pre and post-natal echocardiography. Telemedicine has significant medical, economical and social benefits for patients and families in remote areas and this is particularly true for paediatric cardiology.

P290 (see Abstract 94)
Left Ventricular Structure and Function in Adolescent Swimmers
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P291 (see Abstract 119)
Ectopia cordis in prenatal life
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P292
Effect of Carvedilol on left ventricular systolic and diastolic function in children with dilated cardiomyopathy
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Introduction: Carvedilol has been widely used in adult cardiomyopathy, but very few studies have demonstrated its effects in children.
Objectives: To evaluate systolic and diastolic left ventricular function in children and adolescents with dilated cardiomyopathy (DC) and correlate with outcome.
Material and Methods: Prospective study utilizing Doppler echocardiography to evaluate carvedilol effect on systolic and diastolic left ventricular function in 13 children aged 10 months to 15 years (6.8 ± 1.5 years) with dilated cardiomyopathy. Ejection fraction (EF) was calculated from M-mode measurements and patterns of diastolic dysfunction were classified as normal (N), abnormal relaxation (AR), pseudonormal (PN) and restrictive (R). All patients were on standard therapy when carvedilol was added.
Results: An initial echocardiogram was performed before carvedilol and at monthly intervals. All patients improved their EF from 29.6 ± 2.7% to 44 ± 4.4% after a mean period of 1.2 ± 0.3 years. Diastolic pattern improved from restrictive to normal in 5 and to a pseudonormal in 3. Modified NYHA functional class improved in 8 patients, 3 underwent heart transplantation, 1 in in the waiting list and 1 died.
Conclusions: Carvedilol improves left ventricular systolic and diastolic left function in children with DC as well as their functional class, allowing a better life quality while awaiting for a heart transplantation or even delisting in some cases.

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P293

Glucose-insulin infusion improves cardiac function and energetics during fetal tachycardia

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Introduction: While sustained fetal tachycardia may lead to cardiac failure, hydrops, and intrauterine death, neonatal tachycardia is generally better tolerated. There are many metabolic substrates in neonatal myocardium, but fetal myocardial energy production is almost solely glycolytic. We hypothesized therefore that increased substrate availability by glucose-insulin (GI) infusion would improve fetal myocardial responses to tachycardia.
Methods: We used three porcine models: (1) An isolated fetal heart model (n = 14); (2) An in-vivo fetal model (n = 13); and (3) An in vivo closed chest neonatal model (n = 13). Each animal was randomized to control or GI treatment during tachycardia. In model 1 the controls received normal Krebs-Henseleit perfusion; the GI hearts received double glucose concentration (11 mM) and insulin (1 mU/ml). In models 2 and 3 the GI animals received insulin (100 mU/kg/h) in a 20% glucose solution. The fetuses were 7–14 days preterm, neonates 7–12 days old. Cardiac function was monitored with high fidelity pressure catheters in LV. All animals/hearts were exposed to 90 minutes of pacing at 250–330 bpm after which the hearts were frozen and chemically analyzed.
Results: See table. The isolated hearts in the GI group showed no decline in dP/dtmax during pacing while the controls declined 16% ($p < 0.04$). The in-vivo fetal hearts in the GI group stayed stable or increased in dP/dtmax during pacing, the controls fell ($p < 0.03$). Tau was stable or declined in the GI group and rose in the control group ($p < 0.02$). Myocardial glycogen content was higher and lactate lower in the GI group compared to controls ($p < 0.01$). There were no functional differences in the neonates (study 3) despite higher glycogen content in the GI group ($p < 0.03$).
Conclusion: GI infusion during fetal tachycardia has a beneficial effect on myocardial energetics and cardiac function. These observations may have direct clinical relevance to the management of fetal arrhythmia.

P294

Gene expression profiles in the closing ductus and the great vessels

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The pharmacological approach to ductal patency is currently focused on prostaglandin metabolism. The role of other vascular factors for ductal remodelling is under discussion. Aim of the present study was to define patterns of gene expression in the arterial duct and the surrounding elastic arteries around the time of physiological ductal closure.
Animals and Methods: Ductus, aortae and pulmonary arteries were obtained from 60 Sprague-Dawley rat foetuses resp. pups from 10 litters. The vessels were studied on prenatal day 19, immediately after delivery and 3 days post partum. RNA was isolated from pools of vascular tissue using the SV total RNA Isolation System (Promega, Mannheim, Germany). After reverse transcription the resulting c-DNA was used for thermocycler RT-PCR. 3′ and 5′ specific primers (Gibco, Karlsruhe, Germany) were used for endothelin, PGE synthase, prostanoid-receptors EP1, EP2, EP4, TGF-beta-receptor2 and 3, cyclooxygenase1 and 2, EGF and PDGF-A and PDGF-B, eNOS and iNOS. The PCR-products were separated on 1.5% agarose gel, stained with ethidium bromide, photographed on a transilluminator and compared to GAPDH expression as a reference.
Results: On foetal day 19 all but one (EGF) of the selected genes were expressed at the same or higher levels in the ductus compared to the adjacent arteries. Relative expression of PDGF-A, TGF-beta-receptors2 and 3, prostaglandin-receptor EP4 exceeded the expression of the same genes in both arteries by factor two. At birth expression of PGE synthase, PDGF-B and cyclooxygenase1 was twice as high in the ductus compared to both elastic arteries. Cyclooxygenase2 and eNOS expression in the ductus was half that of the two elastic arteries. On day 3 post partum prostanoid receptor EP4 expression in the arterial duct exceeded that in both elastic arteries by factor two. The rest of the selected genes were expressed at comparable levels in the vessels studied.
Conclusions: Perinatally, the gene expression profiles in arterial duct and surrounding elastic arteries vary by vessel type and time of study. The up-regulation of growth-factor-receptor-genes and growth-factor genes in ductal tissue before closure suggests an important role for the reorganization of the ductus wall.

P295 (see Abstract 112)

Magnetic Resonance Imaging in children with ventricular arrhythmias

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P296

Supraventricular tachycardia (SVT) in children with Ebstein's malformation

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Ebstein's anomaly is rare malformation and affects about 0,3% of children with congenital heart disease. In Ebstein's anomaly supraventricular tachyarrhythmias (SVT) are common and Wolf-Parkinson-White syndrome (WPW) is known to be a congenital substrate for these arrhythmias. The aim of the study was to analyze type and clinical characteristics of paroxysmal SVT in 30 children with Ebstein's malformation (20 girls, 10 boys) aged from 1 day to 12 years (yrs) (mean 16 months) with follow-up from 5 days to 18 yrs (mean 8 yrs). Associated cardiac defects had 23 pts (77%): atrial septal defect – 12 pts, pulmonary stenosis or atresia – 7 pts, ventricular septal defect – 2 pts, corrected transposition- 1 pt, pulmonary regurgitation – 1 pt. In 11 pts. Balloon angioplasty of pulmonary stenosis had 2 pts, in 1 pt angioplasty with stent implantation was performed. Other pts underwent cardiosurgical interventions: Blalock- Taussig shunt, cavopulmonary Glenn shunt, bi-directional cavopulmonary Glenn shunt, valve reconstruction with aortic homograft implantation. In 10 pts intraventricular conduction was found. Eleven pts (27%) had WPW syndrome with right-sided pathways in 10 and multiple pathways in 1 pt. Episodes of SVT had 10 children. Type of SVT was ortodromic in 8 pts, orto- and antidromic in 2 pts. During SVT hemodynamic deterioration and dessaturation were observed. For control of tachyarrhythmias

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sotalol, amiodarone, propafenone were used with variable effect. In 2 pts ablation procedure was performed with excellent result.
Conclusion: Episodes of SVT in pts with Ebstein's anomaly and WPW syndrome are often drug refractory and in some cases ablation procedure may be the only successful therapy.

P297 (see Abstract 101)
Use of covered Cheatham-Platinum stents in aortic coarctation and recoarctation
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P298
Changes of the cardiac surgical treatment strategies on premature babies 1975–2002
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Nowadays, due to the development of cardiac surgery, pediatric cardiology and anesthesia, almost every congenital heart disease (CHD) can be corrected totally or partially. The increasing number of surgical corrections will lead to better life quality.The surgical mortality has decreased significantly, even in the most complex cases. Only few cases can not be treated surgically. The aim of this study was to examine these positive changes, what they mean in the treatment of premature babies with CHD. These patients are endangered because of their age as well, they require special treatment and the surgical treatment has always meant high risk, urgent interventions.
Patients: Between 01 Jan.1975 and 31. Dec. 2002. 427 premature babies were operated on. Overall mortality: 81 patients (18.9%). The patients were divided into 3 subgroups by their weight: I: 470–1500 gr. – 14 patients, II: 1500–2000 gr. – 88 pts., III: > 2000 gr. – 325 pts. Detailed mortality: group I.: 0%, group II: 23 (18.9%), group III.: 58 (17.8%).There were 64 corrective surgical procedures performed mainly after 1998. The successful operations in the smallest weight groups were follows: On ECC: TAPVR (1600gr), TGA – Art. Switch (1800 gr), VSD (1800 gr), Aortico-pulmonary fenestration (2000 gr), HLHS – Norwood procedure (2200 gr), A-V defect (2300 gr), Interrupted aortic arch + VSD (2300 gr), Truncus arteriosus (2500 gr). Without ECC: Coarctation of the aorta (930 gr), PDA (470 gr).
Conclusion: Nowadays the possibilities and the chances of the corrective procedures of CHD in those patients with bodyweight of over 2000 gr and in those that are mature babies are the same. In those patients with bodyweight 1500–2000 gr procedures without ECC had good results, on-pump procedures had higher mortality rate, but the long-term results were preferable. At the moment only procedures without ECC are performed on babies with weight under 1500 gr – but with higher risk. Our effort is to perform ECC operations in patients with bodyweight less than 1500 gr.

P299
Immunization status of children with congenital heart disease in new millennium
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The immunization schedule by the child council of ministry of health in mandatory for every child in our country. However, children with congenital heart disease seem to have incomplete immunization because of various reason. This study included to determine the immunization status of children with congenital heart disease. A questionnaire was proposed to every parent and immunization records inspected for 119 children with congenital heart disease and 102 healthy age matched subjects. The regular schedule consists of BCG, Dyphteria-Pertussis-Tetanus-Polio, maesles, hepatitis B vaccines. Incomplete immunization status was significantly higher in children with congenital heart disease than control's ($p < 0.01$). Incomplete immunization status was significantly higher in acyanotic group (24.6%) than the the cyanotic group (8.3%) for measles. Hepatitis B immunization had been found the most frequent among others for the first two months in both groups (98.6% in acyanotic group and 998.8% in cyanotic group). Low body weight, frequent infections, frequent hospitalisation are the most frequent reasons of incomplete immunization in the study group. Primary care physicians, pediatricians must be informed and trained about the primary care of chronically ill children like congenital heart disease children. Besides regular schedule pneumpococcal and haemophyllus influenza B, varicella and MMR vaccines must also be recommended. Live virus vaccines must be avoided in immunologically deprived children.

P300
Closure of coronary artery fistula with the Amplatzer duct occluder: preliminary results from the USA Registry Group
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Objective: Although rare, coronary artery fistula (CAF) can cause myocardial ischemia, aneursymal dilation of the involved coronary artery, endocarditis, and congestive heart failure. Closure of hemodynamically significant CAF is recommended. Transcatheter coil closure is safe, however, multiple coils are needed for large fistulas and, occasionally, residual shunt may persist. We report use of the Amplatzer Duct Occluder (ADO) device for closure of large and complex CAF, from the uSA registry group.
Methods: Seven patients (pts) had attempt to close CAF with the ADO. Four pts had symptoms of shortness of breath (n = 2) and chest pain (n = 2), and 3 were asymptomatic. The median age was 3.8 years and the median weight was 19 kg. The size of the fistula ranged from 3 to 14 mm. The CAF originated form left coronary artery in 3, and right coronary artey in 4 pts. The CAF drained into the right atrium (n = 2), right ventricle (n = 2), right pulmonary artery (n = 1) and other locations. Six to 9F sheaths size were used for delivery. The median size of the device was 8/6 mm (range 5/4 to 12/10 mm). The CAF was closed by arteriovenous loping (n = 3) or directly from the venous side.
Results: A total fo 12 pts underwent cardiac catheterization with intention to close CAF. In 5 pts, closure was not attempted because of small CAF size. Of the remaining 7, 6 pts had successful placement of the ADO. One pt. developed small pericardial effusion. There were no other complications. Complete closure was acheived at 6 months follow up.
Conclusions: ADO appears to be an excellent tool for transcatheter occlusion of large CAF. Its advantages include small delivery sheath size, easy retrievability after deployment, high complete closure rate, and no incidence of recanalization in this series. More studies and longer follow up will further help to determine the efficacy of the device.

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P301 (see Abstract 111)
Transvenous pacemaker implantation for sinus node dysfunction and arrhythmias after the Fontan operation
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P302
Comparison between echocardiograpic and angiographic-based McGoon ratios in tetralogy of Fallot
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Objectives: Study the McGoon ratio based on echocardiographic measurements, by comparison with the same ratio, classically based on angiographic measurements.
Introduction: The success of tetralogy of Fallot's (TF) corrective surgery depends, among other factors, on the capacity of the pulmonary vascular bed to receive the increased volume of pulmonary blood flow. Among the methods demonstrated to be useful for this purpose, the McGoon ratio (ratio of the sum of both pulmonary arteries diameters by the descending aorta diameter at the diaphragm) is one the more commonly used, its biggest caveat being that it requires an invasive procedure. Therefore, the present study was done to study the validity the McGoon ratio obtained non-invasively, by echocardiography, by comparing it to the classic angiographic-based ratio.
Methods: The echocardiographic and angiographic McGoon ratios were obtained in 20 patients (pts) with TF. The data were collected by retrospective chart analysis of pts with TF's that were observed between 1995 and 2001. The echocardiographic measurements included the largest diameter of the PAs prior to bifurcation and the aortic diameter between the diaphragm and the celiac trunk. A statistical analysis of the data was made.
Results: The medium age was 2,2 years (0,01–10,6). Four patients were female. The time gap between the catheterization and the echocardiography was 18 days (0–123). The mean echocardiographic and angiographic McGoon ratios were 1,85 (0,98–2,45) and 1,70 (0,97–2,35), respectively. The difference between both means was 0,15 (0,08–0,21, CI 95%, $p < 0,001$). The angiographic McGoon Ratio was in mean 8% larger than the echocardiographic counterpart. Only 15% of the pts (3/20) had an echocardiographic ration larger than the angiographic ration. The correlation between both ratios was 0,944 ($p < 0,001$) and the regression equation to relate both was angio = 1,1860*echo-0,1696.
Conclusions: In selected cases of TF, the McGoon ratio based on echocardiographic measurements represented a reliable and reproducible non-invasive method to evaluate the size of the pulmonary arteries and thus the possibility of corrective surgery.

P303
Effects of repeated chlamydia pneumoniae infection on coronary artery endothelial function in piglets
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Background: Earlier in vitro studies have furnished support to the concept that repeated rather than single infection with Chlamydia pneumoniae contributes to the early development of atherosclerosis. Arterial endothelial dysfunction is a primary mechanism in the pathogenesis of atherosclerosis. We investigated both in vivo and in vitro, the effects of repeated inoculation with C. pneumoniae on coronary artery endothelial function in piglets.
Methods: Nine piglets were given two intratracheal inoculations of either C. pneumoniae (n = 5) or saline (n = 4) at 2-week interval. At 4 weeks after the first inoculation, coronary flow velocity (CFV) was measured in the LAD coronary artery with a Doppler flow wire in response to bradykinin, an endothelium-dependent vasodilator, before and after infusion with L-arginine, a substrate to nitric oxide synthesis, and glutathione, an antioxidant. Endothelium-dependent relaxation of PGF2alpha-precontracted LAD coronary rings was studied in vitro in response to serotonin, in the presence of ketanserin. Plasma nitrate and the activity of endothelial nitric oxide synthase (eNOS) from LAD coronary samples were measured.
Results: CFV in response to bradykinin was less in infected than in noninfected animals ($p < 0.1$). Infusion with L-arginine improved CFV in infected animals, having no effect in noninfected animals. In vitro, serotonin caused further constriction of LAD coronary rings in infected animals, while a stepwise relaxation response to this agonist was noted in noninfected animals ($p < 0.0001$). Both plasma nitrate and eNOS activity were significantly decreased in infected animals, as compared to noninfected animals.
Conclusion: Repeated C. pneumoniae infection results in persistent endothelial dysfunction of the large coronary arteries while the resistance coronary vessels appear to be less affected. The coronary endothelial dysfunction appears to be due to a markedly diminished NO production, a consequence of eNOS down-regulation along with diminished substrate availability to NO synthesis. This pathological condition could initiate the cascade of further atherogenic events.

P304
3 years experience with bovine jugular vein grafts for RVOT-reconstruction
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Center for Congenital Heart Disease Amsterdam Leiden. Amsterdam, Leiden, The Netherlands

Objective: To assess the results of RVOT-reconstruction with bovine jugular vein (Contegra®) grafts.
Methods: 54 Contegra® prostheses were used in 48 patients. Median age was 11 (range: 0.1–334) months. 37 prostheses were used for biventricular correction of PA, VSD ± MAPCA's (15), absent pulmonary valve syndrome (2), truncus arteriosus (8), TGA/DORV, VSD and PS (7), aortic atresia with VSD (2) or for a Ross procedure (3). 11 grafts were implanted to replace obstructed homografts (8) or to correct pulmonary regurgitation following Fallot repair (3). 6 patients received a second Contegra® graft. Echocardiographic follow-up was performed at 3 months intervals and is complete in all patients. Mean follow-up is 15,4 ± 9,3 months.
Results: Non-graft related mortality occurred in 3 patients (6,2%). 6 patients were reoperated for PA branch stenosis (2), endocarditis (2) or distal endoluminal graft fibrosis (2). Another patient is awaiting reoperation for distal PA branch stenosis. Conduit stenosis did not occur in the remaining patients. More than mild graft regurgitation was observed in 5 patients and was due to endocarditis (2), PA branch stenosis (2) or pulmonary hypertension (1). Two patients with distal PA stenosis and pulmonary hypertension showed significantly dilated grafts.
Conclusions: 90% of the Contegra® conduits used for RVOT reconstruction remained free of obstruction or more than mild regurgitation. Calcific degeneration did not occur in 3 years follow-up and obstructive endoluminal graft fibrosis was observed in only 2 patients. The Contegra® conduit is a good alternative for the pulmonary homograft, especially in neonates and infants.

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Exhibition

Industry profiles

Baylis Medical

Stand B

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