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Abstracts

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Session I: Transplantation
Chairman: J. Taylor

PEDIATRIC HEART TRANSPLANTATION: SURGICAL PROBLEMS IN RECIPENTS WITH CONGENITAL HEART MALFORMATIONS.

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Heart transplantation in recipients with complex cardiac malformations may carry an increased risk because of the underlying anatomy. In addition, the potentially distorting anatomical effects of previous operations must be managed.
Out of 45 children who underwent heart transplantation, 19 (42%; mean age: 4.8±4.1 yrs; range: 13 dys - 13 yrs) had complex malformations including: single ventricle variants (4), hypoplastic left heart syndrome (3), corrected transposition (3), complex transposition of the great arteries (2), atrioventricular septal defect (2), left ventricular outflow tract obstruction (2), tetralogy of Fallot, Ebstein's anomaly, multiple ventricular septal defects (1 of each). Heart transplantation was performed as a primary procedure in 5 patients, after palliative surgery in 4, and after failed complete repair in 10. Additional surgical procedures included: takedown of mediastinal adhesions (13), repositioning of transposed great arteries (8), reconstruction of the pulmonary or the aortic pathway (5 and 4), correction of total anomalous pulmonary venous drainage (1). Reconstructive procedures were performed using donor and/or recipient tissue. There were 4 early and 5 late deaths, yielding survival rates of 60%±12% and 51%±13% at 1 and 2 years, but no deaths were related to surgical technique. Probability of survival was not significantly different in recipients with cardiomyopathy (26 patients, 72%±9% at 1 and 2 years) (p=0.27).
In conclusion, careful planning of both harvesting and transplantation techniques allow orthotopic heart transplantation in children with complex congenital heart malformations. The surgical technique may be demanding but the operative risk is not increased.

HEART TRANSPLANTATION IN CHILDREN : RESULTS IN COMPARISON WITH ADULTS.

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Heart transplantation, now an established therapeutic tool in end-stage cardiomyopathies in adult, is increasingly used in children. Between January 1988 and November 1991, 11 children aged from 5 days to 12.6 years (mean 4.2 years) and 65 adults underwent consecutively orthotopic heart transplantation in Marie-Lannelongue Hospital. Immunosuppressive therapy was initially the same for all the patients: thymoglobulin (3 days), azathioprine and prednisolone, and cyclosporine. Tritherapy was carried on in adults and children over 2 years of age. Steroids were discontinued after 3 weeks in younger children. Operative survival rate was similar in both groups (90.5% vs 90.3%). Rejection episodes were diagnosed by serial endomyocardial biopsies except for younger children in whom diagnosis was made only by echo-doppler in some patients. Rejection rate was 1.2 rejection/patient in children below 2 years of age, 3 in older children and 1.5 in adults. Refractory rejection requiring thymoglobulin or OKT3 therapy was more frequent in children over 2 years of age: 50%, vs 3.7% in adults (p=0.01). Infection was rare in both groups: no bacterial infection in children vs 3.7% in adults; no CMV infection in children vs 14.8% in adults; 1 viral bronchiolitis in children; no fungal or protozoarian infection. Systemic hypertension requiring therapy was present in 20% of children vs 41% of adults. No child had elevated blood creatinine levels, vs 52% of adults (p<0.01). Two- and 3-year actuarial survival rate are similar in both groups (69.2% vs 69.5%). In conclusion, short-term results in children are similar to those observed in adults; only minor side effects of immunosuppressive therapy are observed; however, refractory rejection occurs more frequently in children over 2 years of age than in adults and younger children.

NEW ASPECTS IN HEART TRANSPLANTATION IN CHILDREN

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Heart transplantation is a well accepted procedure for the treatment of endstage heart failure. In children, however, only recently acceptable results were achieved. Out of a total number of 496 heart transplants we analyzed our experiences in 22 children. Preoperative diagnoses was primary cardiomyopathy in 15 patients, congenital heart disease in 5, 2 further patients suffered from endocardial fibrosis. In 3 patients, the implantation of a mechanical support system (biventricular, univentricular, or ECMO system) as bridge to transplant was necessary. All children received postoperatively triple drug immunosuppressive therapy consisting of cyclosporine A, azathioprine, and prednisolone. Perioperative immunosuppression was supplemented by ATG which was administered for 4 days. Rejection diagnosis was based on noninvasive methods i.e. intramyocardial electrogram (IMEG) and echocardiography. Only in a few instances transvenous endomyocardial biopsy was necessary. At present 16 children (73%) are alive and well. The mean observation period was 22 months (6 weeks to 76 months). Six patients died from early postoperative graft failure, one other patient from rejection. One chronic kidney failure was treated successfully with kidney transplantation. The physical and psychological development of all children appeared to be normal.
The survival rate of children and young persons after heart transplantation is similar to that of adults.

Noninvasive methodes for diagnosis of rejection decrease postoperative morbidity and increase early rehabilitation. Triple drug immunosuppression appears not to affect physical and psychological development significantly. Mechanical circulatory support as a bridge to transplantation is possible with acceptable results.

MAINTENANCE IMMUNOSUPPRESSIVE THERAPY CANNOT ALWAYS AVOID STEROIDS AFTER HEART TRANSPLANTATION IN CHILDREN.

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As a rule, oral steroids should be excluded from long term therapeutic protocols in infants and children to avoid a great number of side effects, mainly those affecting the patient's growth. Is it possible to apply such a rule to maintenance immunosuppressive therapy? Between 1987 and 1991, 45 patients ranging from 13 days to 15 years in age, underwent heart transplantation. Post-operative immunosuppression consisted in a short course of anti-thymocyte globulins and IV methylprednisolone. Maintenance therapy included cyclosporine and azathioprine. Endomyocardial biopsies were performed both on a routine basis and as soon as rejection was clinically suspected. Patients with recurrent episodes of rejection received prednisone in addition to their maintenance protocol. Prednisone was given at least 1 month, and discontinued only when control biopsy returned to normal and remained so subsequently.
Out of our 23 early survivors followed-up for more than 6 months, 11 (48%) were given maintenance prednisone during 9.4+7.7 months (group P), whereas 12 were not (group NP). Only 2 needed maintenance prednisone beyond the second post-operative year. Mean cyclosporine dosages during post-operative months 1-3 and 4-6 were compared in both groups, and were respectively 11.7±4.8 mg/kg/d (P) vs 18.3±6.7 (NP) ($p<0.01$), and 11.8±4.6 (P) vs 16.3±5.7 (NP) ($p<0.05$). Age at transplantation did not differ.
These data suggest that: 1/ there is no age related difference regarding the need for maintenance prednisone therapy; 2/ decreasing cyclosporine dosage increases the need for steroids; 3/ maintenance prednisone can be discontinued before the end of the second post-operative year in most patients.

APPLICABILITY OF CROSS SECTIONAL ECHOCARDIOGRAPHY FOR DIAGNOSIS OF REJECTION FOLLOWING HEART TRANSPLANTATION

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Purpose of this study was to examine whether rejection following heart transplantation can be diagnosed by cross sectional echocardiography of the left ventricle (LV). Echo tapes from 34 patients with a median age of .5 years, who had undergone heart transplantation were analyzed . LV regional wall motion was assessed by calculating area change in 8 segments of the heart at mitral valve and papillary muscle level in the short axis view and in the four chamber view. The endocardium was manually traced in an enddiastolic and an endsystolic frame. The centres of mass of these frames were calculated and superimosed to correct for movement of the heart (floating system). Data were compared to normal values previously established from 55 controls. LV mass and volume were evaluated by echocardiography in the apical 2 and 4 chamber view. From these 2 perpendicular imaging planes we calculated mass as difference between epicardial and endocardial volume x 1.05 (specific gravity of heart muscle). Mass divided by volume at enddiastole yields the mass/volume index. Normal values had been established in 95 controls. Reproducibility of both methods had previously been demonstrated by a low interobserver variability. All studies were evaluated by one blinded observer (MV) who had no knowledge of the clinical status of the patient. Results: All 34 transplant recipients had normal regional wall motion. The mean endocardial volume was 99% of normal, the mean LV mass was 124%of normal and LV mass/volume index was 125 % of normal. One of the 34 transplant recipients had an acute rejection, which could be detected by a increase in LV mass to 221% of normal (185% of the LV mass of the other transplanted patients), while regional wall motion remained normal. These preliminary data suggest that cross sectional echocardiographic examination of LV mass may be helpful to prospectively detect acute rejection of heart transplants, while regional LV wall motion may remain normal during rejection.

REHABILITATION AND LONG-TERM PROGNOSIS AFTER HEART TRANSPLANTATION IN CHILDREN AND ADOLESCENTS

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Since the introduction of cyclosporine, orthotopic heart transplantation (OHTX) has become an accepted therapy of end-stage heart disease in adults. However, only limited experience is available with this procedure in the younger age group, especially with regard to long-term results.
From 09/85 to 10/91 we performed 338 OHTX in 225 patients (pts). Fifteen pts (6,7 %) were less than 18 years old (range 4 - 17 years, mean age : 12,5 ± 3,4 years). The underlying disease was end-stage congestive cardiomyopathy in all but 1 pt (endomyocardial fibrosis). Three pts died early postoperatively due to cerebral bleeding (3rd p.o.d.), gram-negative sepsis (14th p.o.d.) and servere acute rejection (68th p.o.d). Follow up in the survivor group ranged between 543 - 2182 days (median : 1394 days). Immunosuppressive regime early and late postoperatively was the same as for adults (cyclosporine A, prednisone, azathioprine). Three children, however, did not receive initial prophylaxis with ATG. The rate of rejection episodes requiring therapy revealed no significant differences at any time postoperatively compared to the adult group and kidney function was not impaired despite of the use of cyclosporine in the younger age group. Cardiac catheterization was performed yearly and showed excellent right and left ventricular function in all pts without any signs of graft arteriosclerosis up to 6 years postoperatively. One pt developed mild tricuspid insufficiency (I° - II°). At 6 months postoperatively, all children except for the youngest were back at school or work.
OHTX represents the treatment of choice for end-stage heart disease in adults and in an increasing number of children. Individualized immunosuppression of this younger patient group resulted in normal long term renal function and rejection frequency was the not elevated compared to adults. However, graft arteriosclerosis has not been found in children and adolescents up to 6 years postoperatively. Active physical rehabilitation and social life is not restricted.

ISOLATED LUNG TRANSPLANTATION VERSUS HEART-LUNG TRANSPLANTATION IN THE TREATMENT OF EISENMENGER'S DISEASE

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Aside of primary pulmonary hypertension, Eisenmenger's disease (EMD) represents a maior indication for heart-lung transplantation (HLTX). However, in selected cases of EMD with ASD, VSD or PDA isolated lung transplantation (ILTX) in combination with repair of the underlying defect may represent a more reasonable therapeutic alternative for better long term prognosis in this young patient group. This omittes early or late complications related to the transplanted heart and allows replacement of the contralateral lung in cases of obliterative bronchiolitis later on. Additionally, the contralateral lung and the heart of the donor can be transplanted to other recipients.

Between 5/88 and 9/91 a total of 13 pts underwent transplantation for EMD. In 3 pts (age : 16 - 29 (22,3 ± 6,5) years) ILTX and in 10 pts (age : 10 - 52 (27,2 ± 11,0) years) HLTX was performed. In the ILTX group ASD, VSD and PDA was present in one case each. The mean pulmonary artery pressure (PAP) ranged preoperatively between 62 - 99 (mean : 63,0 ± 35,5) mmHg in this group and there was no evidence of pulmonary valve insufficiency. In the postoperative course, 1 pt died of hypoxemia due to accidental airway decannulation One pt who underwent left lung transplantation required double lung retransplantation and closure of a previously undetected aorto-pulmonary window resulting in persistent pulmonary hypertension and early graft failure. The underlying diagnoses in the HLTX group were ASD (n=2), VSD (n=3), PDA (n=1), TGA (n=2) and AVC (n=2). In this group preoperative mean PAP ranged between 52 - 104 (mean : 77,2 ± 17,2) mmHg and in 4 of 6 pts with ASD, VSD or PDA severe pulmonary valve insufficiency (PVI) was present. There was no early death in the HLTX pts with one late (7 months) death.

In cases of EMD with correctable congenital heart disease, ILTX combined with the repair of the underlying defect may represent the less traumatic therapeutic option. To confirm the indication for ILTX versus HLTX in these pts invasive right heart catheterization or color doppler echocardiography with estimation of the PAP and especially the degree of PVI are required. While PAP showed no significant differences in between both groups the degree of PVI represents the most important point of decision making wether or not to replace the heart.

Session II: Heart Failure
Chairman: E. Shinebourne

CAPTORIL (ZORKAPTIL) IN TREATMENT OF HEART FAILURE DUE TO CHD WITH PULMONARY HYPERTENSION

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Twenty five children aged 3 mo to 7yrs (mean 20 mo) were studied. DAP+ASD+VSD had 3pts, VSD+ASD had 5, VSD had 12 and AVSD had 5. All of them were in chronic congestive heart failure, with tachycardia, tachypnea and hepatomegaly.

At the time of the study they have all been receiving optimal doses of digitalis and diuretics at least one month. All pts had biventricular or marked right ventricular hypertrophy on ECG, marked cardiomegaly and increased pulmonary vascularity on x-ray.

All children were by noninvesive and invesive methodes investigated. Zorkaptil (Captopril) was given oraly 1-2 mg/kg/24h, during 1 to 4 mo and then they were reexamined after 1/or 3 mo.

After zorkaptil (z) dyspnoa, tachycardia, hepatomegaly, desappeared in 42% of pts and in 50% was much less. The cardiac silhuette diminished in 88% of pts. The tolerance on effort was much better.

Before (Z) PA/Ao pressure ratio was 0,98±13 and after PA/Ao pressure ratio was 0,79±0,11 with regretion line y=-0,04x+0,83 (p < 0,05). Mean PA pressure decreased significantly after Z p < 0,001: before Z was 79,57±13,7 and after Z 47,14±9,4. Mean Ao pressure before Z was 73,6±10,4 and after 71,0±7,94 (NS). Mean ratio Pa/Ao pressure was significantly decreased p < 0,001 after Z: bifore 1,1 ±0,25, after 0,67±0,17 (y=0,2x+0,83).

Heart rate before Z was 131,2±18,2, after 108± 13,5 p < 0,05.

ACE inhibitore are important in treatment of heart failure and PAH especially in pts before the surgical intervention.

* *Intraduced*

ENALAPRIL IN CHILDREN: 5-YEAR EXPERIENCE

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The records of 60 children receiving enalapril (E) between 1986 and April 1991 were reviewed. There were 64 treatment periods, age range 8 days-16 years (mean 4.5 years, median 4.9 months). Eighty-three percent were in heart failure graded 3 or 4 of 4; all were on digoxin and/or diuretics. All patients received a test dose of E and if tolerated the dose was increased gradually. Assessments were made prior to treatment, at 1 week, at 1 month and at late follow-up using available clinical, laboratory, CXR and echo data.

Results: The maintenance dose of E was 0.04-0.91mg/kg/day (mean 0.28mg/kg/day). All haemodynamic groups improved except left-to-right shunts who were all under 3 months of age.

Haemodynamic group	n	Improved	No change	Side-effects*
Left-to-right shunt	15	5	5	5 renal
Cardiomyopathy	14	8	6	-
Postop: palliated	16	12	3	1 renal
Postop: "repair"	7	7	-	-
Valve regurgitation	12	7	4	1 neutropenia
	64	39	18	7

* required discontinuation of enalapril. Three others also under 3 months of age had renal impairment but E was not stopped. Renal impairment was not dose-related but was associated with fluid restriction. Hypotension requiring dose modification occurred in 8.

Conclusion: Enalapril appears clinically safe and effective in diverse cases of heart failure in children except in infants less than 3 months old with left-to-right shunts. Such infants are unlikely to have surgery deferred by ACE inhibitor therapy.

IBOPAMINE: A LAST RESORT TREATMENT FOR INTRACTABLE HEART FAILURE IN CHILDREN ?

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Heart failure resistant to conventional drug therapy is a difficult management problem in children. Ibopamine (oral Dopamine) is an attractive option for home therapy of such patients. The renal and cardiac effects of oral Ibopamine were studied in 8 patients aged 8 mo to 17 yrs (median 7 yrs) with severe heart failure since November 1990. All were in NYHA class IV. The cause of heart failure was dilated cardiomyopathy in 5; Rheumatic mitral valve disease in 1; post-op Fontan in 1; and broncho-pulmonary dysplasia in 1. All received diuretics, 5 had digoxin; and 4, captopril. Ibopamine 2 mg/kg/d in 2 doses was given for 2 weeks to 9 mo (median 6.5 mo).

Cardiac effects: Symptomatic improvement occured in 6 patients (to NYHA III in 2 & II in 4). Two patients showed no improvement and died 2 wks and 5 mo later. Left ventricular % fractional shortening improved from 11-32 % (mean, SD; 18.4, 8.1) to 15-36% (mean, SD; 24.4, 8.8), p < 0.1.

Renal effects: Glomerular filtration rate (Inulin clearance) and renal perfusion rate (Para amino hippuric acid clearance) showed a transient increase. Fractional Na

excretion and Lithium clearance (a marker of proximal tubular Na reabsorption) were unchanged.
No adverse effects were noted and no patient required discontinuation of Ibopamine.

Conclusions: Ibopamine appears to be safe in children. While no consistent measurable effect on cardiac or renal parameters were seen, most patients showed impressive symptomatic improvement.

DOBUTAMINE STRESS TEST TO ASSESS CARDIAC FUNCTION IN LONG-TERM CHILDHOOD CANCER SURVIVORS TREATED WITH DOXORUBICIN

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We studied the use of inotropic stimulation with dobutamine during echocardiography to detect differences in cardiac function between 21 asymptomatic doxorubicin treated pts and 12 healthy, normal control subjects of similar age. The experimental group (mean age = 16 yrs) was treated a median of 5.3 yrs (range 1.6-14.3 yrs) prior to this study with doxorubicin doses ranging from 27 to 532 mg/m2 (mean = 196 mg/m2). Echo-Doppler studies were performed prior to infusion and at the end of 20 min infusions of 0.5, 2.5, and 5.0 µg/kg/min of dobutamine. We measured HR, BP, cardiac output indexed (CI), stroke volume indexed (SVI), LV shortening fraction (SF), LV end systolic wall stress (LVESS), LV posterior wall thickness in systole and diastole (LVPWs and LVPWd), isovolumic relaxation time, deceleration time and the ratio of early to late diastolic mitral velocities. There were no significant differences between the groups at rest or during dobutamine stress for HR, BP, CI, SVI, LVPWd or diastolic function parameters. We found no significant correlation between any parameter and doxorubicin dose. LVPWs consistently differentiated the 2 groups. Data comparing the 2 groups are as shown:

dobutamine µg/kg/min	LVPWs(mm) doxo	control	SF doxo	control	LVESS doxo	control	
0	11.0**	13.1	0.34	0.37	68	56	
0.5	11.9**	14.8	0.37	0.39	58	46	
2.5	13.3**	17.1	0.41*	0.46	50*	36	*p<.05
5.0	14.1**	19.3	0.45**	0.51	47**	28	**p<.01

Conclusion: Our data suggest that asymptomatic doxorubicin treated pts may have cardiac damage undetected by the commonly used SF measurement. Decreased SF and increased LVESS were found in doxorubicin treated pts only during moderate dose dobutamine. LVPWs, the most sensitive indicator of doxorubicin treatment, was decreased in doxorubicin treated pts at rest, but differences were greater with inotropic stimulation.

COMPARATIVE HEMODYNAMIC EFFECTS OF DOPAMINE (DP) AND DOBUTAMINE (DB) FOR TREATMENT OF EARLY POSTOPERATIVE HEART FAILURE AFTER TOTAL REPAIR OF TETRALOGY OF FALLOT (TF)

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Dp and Db are used often for inotropic support in early postoperative heart failure. In order to compare their potency to increase cardiac output (CI) after total repair of TF, in a prospective study 11 children were administered i.v., in random order, increasing doses of Dp (3.3 - 8.3 - 16.6µg/kg.min) and Db (3.3 - 6.6 - 11 - 16.6µg/kg.min), each dose - for 15 min. At the end of each infusion period hemodynamic variables were recorded. CI was measured by thermodilution. For similar doses Db caused greater increase of CI, heart rate (HR), and mean arterial pressure (MAP); CI increments being 7.8%; 18.7%; 28.9% and 32% for respective doses of Db, and 3.9%; 14.6% and 23.5%, respectively, for Dp.

Table1 Dopamine (µg/kg.min)

	Control	3.3	8.3	16.6
HR (Beats/min)	118+/-8	120+/-11	125+/-18*	138+/-12***
MAP (mm Hg)	72+/-12	71+/-10	79+/-9*	85+/-8***
CI (l/min.m^2)	2.8+/-0.6	2.9+/-0.5	3.2+/-0.6**	3.5+/-0.5***

Table2 Dopamine (µg/kg.min)

	Control	3.3	6.6	11	16.6
HR	120+/-12	127+/-14*	143+/-13***	157+/-9***	165+/-7***
MAP	70+/-10	73+/-9*	81+/-10***	84+/-9***	86+/-12***
CI	2.9+/-0.6	3.1+/-0.6**	3.4+/-0.7***	3.7+/-0.7***	3.8+/-0.6***

mean+/-sd; * - p<0.05; ** - p<0.01; *** - p<0.001 vs control

Systemic vascular resistance, LAP and SVI did not change significantly. These results imply that after total repair of TF the increase of CI is achieved mainly by tachycardia and that Db is more effective for inotropic support in early postoperative heart failure as it causes greater increase of HR and, hence, of CI.

CONTINUOUS EXTRACORPORAEL FLUID REMOVAL BY ULTRAFILTRATION/HEMOFILTRATION IN CHILDREN WITH LOW CARDIAC OUTPUT AFTER CORRECTIVE CARDIAC SURGERY

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From June 1986 to October 1991 22 children developed drug resistent low cardiac output syndrome (LCO) after corrective cardiac surgery.They were hypervolemic and oliguric up to 12 hrs despite conventional medical therapy, and thus treated by slow continuous ultrafiltration (SCU) or continuous arterio(veno)venous hemofiltration (CA(V)VH).

Mean continuous extracorporeal fluid removal lasted 49±6.1 hrs. A mean negative fluid balance of 1.6±0.4 ml/kg/h decreased the mean body weight from 8.1±1.1 to 7.6±1.3 and significantly improved the hemodynamic status and oxygenation (Table 1).

Table 1:Hemodynamic data, oxygenation and pH

	pre	post-SCU/CA(V)VH	p-value
MAP (mmHg)	41.6±1.5	53±2.8	<0.001
CVP (mmHg)	16±0.8	10.2±0.7	<0.0001
HR (beats/min)	152±2.9	142±2.4	<0.01
PaO_2/FiO_2 (mmHg)	98±9.4	159±18.7	<0.005
pH	7.26±0.01	7.38±0.02	<0.0002

Hemodynamic improvement during SCU/CA(V)VH allowed a significant reduction of the mean infusion rate of dopamine, dobutamine and epinephrine from 18.9 to 13, 19 to 9 and 0.31 to 0.17 mcg/kg/min, respectively.

In 12 patients urinary output returned to normal. Due to persistent anuria CA(V)VH had to be continued up 19 days in 10 patients. Survival rate was 55%.

Continuous extracorporeal fluid removal by SCU and CA(V)VH in children with LCO after cardiac surgery improves acid-base balance, cardiovascular function by otpimizing preload conditions and oxygenation by reducing pulmonary edema.

Session III: Oral Presentation of Posters
Chairman: I. Oberhänsli

THE "END-ON" AORTOGRAM IN TETRALOGY OF FALLOT: IMPROVED DEFINITION OF CORONARY ARTERIES WITHOUT SELECTIVE ANGIOGRAPHY.

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In Fallot's tetralogy the prevalence of significant coronary artery anomalies is 2-9%. Precise preoperative identification of either left anterior descending or right coronary artery crossing the right ventricular outflow tract will optimise surgical decision making. Preoperative aortography in anteroposterior and lateral projections is routine but often fails to show coronary arteries clearly enough to identify anomalies with confidence.

We have developed an angiographic view with steep caudal angulation to show the aortic root "end-on" and to show the origin and courses of the coronary arteries. The ability of this view to identify coronary arteries crossing the right ventricular outflow tract was studied prospectively.

Nineteen consecutive children (age 6-156 months, median 13 months) with Fallot's tetralogy underwent routine preoperative cardiac catheterisation. Two patients had a right sided arch, one had dextrocardia and two had functioning aorto-pulmonary shunts. A biplane aortogram was performed ("end-on" and lateral projections) using 1 ml/kg of contrast injected at 1 ml/kg/s. The "end-on" projection had caudal tilt of 35-45 degrees and left oblique angulation of 0-30 degrees. A wedge was placed under the dorsolumbar area to increase caudal tilt to approximately 50-60 degrees. The origins and routes of both coronary arteries were clearly seen in all cases. One patient was found to have an anomalous left anterior descending artery crossing the right ventricular outflow tract and underwent palliative surgery. All other findings were confirmed at subsequent operation.

The "end on" aortogram enables normal and anomalous coronary arteries to be identified reliably and obviates the need for selective angiography.

DISCREPANCY BETWEEN ASSUMED AND MEASURED OXYGENCONSUMPTION IN CHILDREN DURING CARDIAC CATHETERISATION

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In pediatric practice cardiac output during cardiac catheterisation is most frequently determined using the Fick-method with assumed oxygenconsumption (VO_2). However, important discrepancies between assumed and individual measured values of VO_2 are observed.
The object of this study is to determine the influence of intra-cardiac shunting on VO_2 to explain these observed differences.

VO_2 measurements were performed with a modified version of the system described by Stam et al., consisting of a ventilator (Siemens 900A/C) an O_2- and CO_2-analyser (Datex, Capnomac), two mixingbags, a one-way-valve and a three-channel-recorder. The system was validated by methanol-combustion tests and by comparing cardiac outputs, derived simultaneously by the Fick-method -using measured VO_2-and by the dye-dilution-method, in patients without shunts.
VO_2 was measured 36 times in 31 patients with different cardiac malformations. They were divided into three groups: I. patients with cyanotic heartdisease, II. patients with large left-to-right shunts and III. patients without intracardiac shunts.
Measured VO_2 values are compared to predicted values which are based on the tables of LaFarge and Miettinen.

group	n	VO_2 ml/min/m² (mean ± sd) assumed	measured	p	r
I	5	168,2 ± 20,0	150,3 ± 17,0	0,04	0,65
II	7	139,9 ± 18,2	166,8 ± 29,8	0,02	0,66
III	24	153,4 ± 25,3	150,0 ± 25,2	0,54	0,85

Conclusion
Intracardiac shunting, considerably affects oxygenconsumption. Therefore, cardiac output calculated with VO_2 values, obtained from predictive tables is over- or underestimated depending on the kind of heartdisease.

LEFT VENTRICULAR DIASTOLIC FUNCTION DURING ACUTE RIGHT VENTRICULAR AFTERLOAD INCREASE

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To evaluate the effects of right ventricular afterload increase on left ventricular (LV) filling dynamics we studied 8 dogs, in baseline conditions and after acute banding of the main pulmonary artery. Dogs, 13.3 ± 1.1 Kg, were studied under general anesthesia with open chest and pericardium. Pressures were measured in both ventricles and cardiac output was evaluated by thermodilution. LV diastolic function was measured by echo Doppler analysis of mitral flow. Peak modal velocities of E and A waves, E/A ratio, mean deceleration time (EDT) and isovolumic relaxation time (IRT) were all measured, in baseline and banding conditions:

LVDP mm Hg	E wave cm/s	A wave cm/s	E/A	EDT ms	IRT ms
1.99 ± 0.61	72.3 ± 1.9	55.2 ± 1.4	1.32 ± 0.05	75.1 ± 1.7	63.1 ± 2.5
3.75 ± 1.12	68.9 ± 1.7	59.6 ± 2.2	1.16 ± 0.03	68.0 ± 1.8	69.4 ± 2.9
NS	NS	NS	$p < 0.02$	$p < 0.01$	NS

An acute banding of the pulmonary artery, adjusted to produce a 100% increase in right ventricular systolic pressure, has indeed restricted LV filling. E/A ratio and EDT variations, together with normal relaxation, suggest a change in LV pressure - volume conditions with decreased distensibility.This pattern possibly due to the interventricular dependence has clinical implications and should be considered in children surgically palliated with a pulmonary banding.

PERSISTING LV HYPERTROPHY LATE AFTER COARCTATION REPAIR

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Left ventricular function was assessed by echocardiography in 28 normotensive patients 8.3 y after coarctation repair (residual gradient 7mmHg). Patients were grouped according to their age at the time of surgery: group 1 ≤ 1 y (G 1, n=14), group 2 > 1 y (G 2, n=14). Two groups of age- and sex-matched volunteers (C 1 & C 2) served as controls.

	C 1	G 1	C 2	G 2
age at study (y)	11	11	17	17
SP(mmHg)	111	116	117	125
SPex(mmHg)	153	168	168	189*
ΔPex(mmHg)	21	36*	23	37*
LMMI (g/m2)	59	81**	74	105**

SP: resting systolic blood pressure, SPex: SP at submax. exercise, ΔPex: postexercise gradient; *p < 0.05 and **p < 0.01 vs. C.

LV systolic function according to the wall stress - shortening relationship was normal in all patients.

Conclusions: Despite successful coarctation repair, residual LV hypertrophy persists irrespective of the age at operation. This can be explained by the abnormal SP response due to the increased pressure gradient under exercise conditions.

LV DIASTOLIC DYSFUNCTION LATE AFTER COARCTATION REPAIR ?

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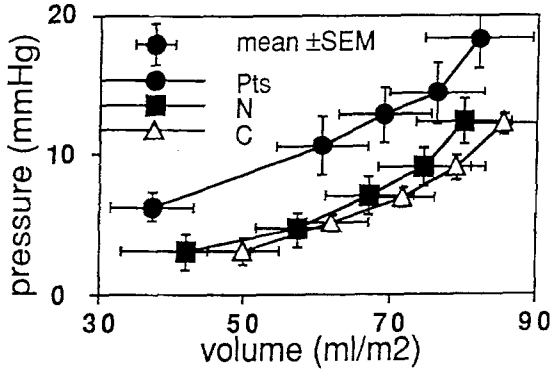
Systolic and diastolic LV function were evaluated angiographically in 7 normotensive patients (Pts) 8 years after coarctation repair (residual pressure gradient: 7 mmHg). Data at rest and after nitroprusside infusion (N, 1.8 µg/kg/min) were compared to 9 controls (C).
Systolic LV function (ejection fraction - mean systolic wall stress relationship) was normal in all Pts. However, angiographic muscle mass (118 vs. 88 g/m^2) and LV end-diastolic pressure (17 vs. 12 mmHg) were significantly elevated compared to C. There was a linear relationship between muscle mass and end-diastolic pressure ($r=0.66$, $p<0.01$).



The parallel downward shift of the pressure-volume curve after N suggests a pericardial constraint at rest which is probably due to residual LV hypertrophy.
Conclusions: Systolic function is normal late after coarctation repair. The upward shift of the diastolic pressure-volume-curve is probably caused by residual LV hypertrophy and can be modified in a beneficial way by the administration of a vasodilator.

MR-ANGIOGRAPHY AND 3D-RECONSTRUCTION OF THE CARDIOVASCULAR SYSTEM.

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Introduction: A wide variety of congenital and acquired heart diseases has been successfully investigated by MRI. In patients with severe vascular malformation e.g. hyopoplastic pulmonary arteries problems arise to clarify the relations and connections of arteries and veins. Respiratory and cardiac movement artifacts, however, may limit the potential value of MR-angiography (MRA) in these patients.

Methods: Two different techniques were used for MRA of the cardiovascular in 18 children (7 PA with VSD, 1 TOF, 2 Cor triatiatum, 2 Scimitar-syndrome, 1 TAPVR, 1 anomalous systemic vein connection, 4 D-TGA) and compared with conventional spin echo images and conventional angiocardiography. All MRA examinations were performed on a 1.5 T Magnetom (Siemens, FRG) using a standard circular polarized head coil (FOV 20 cm) or body coil (FOV 33 cm) with a 256 x 256 matrix. A 3D gradient echo sequence (TE 8 ms, slice thickness 16/32 mm, partitions 16/32) was applied without ECG-gating, further cardiac ECG-gated MRA was performed using a 2D-gradient echo sequence with a combination of the cinemode and multislice technique. After the selection of a region of interest the images were interpolated leading to 0.5 mm isotropic voxels. These images were then post-processed by a ray-tracer algorithm or used for surface reconstruction.

Results: Even without ECG-gating MRA of the cardiovascular system is possible. Compared to SE images pulmonary arteries and veins were clearly differentiated from bronchial structures and were traced to subsegmental levels. The course and connection of aortopulmonary collaterals were demonstrated, in contrast to the SE images. In two patients severe hypoplastic pulmonary arteries could be detected, what was confirmed by pulmonary vein wedge angiocardiography. Compared to the ECG-gated 2D-method the ungated 3D-technique is fast and small vessels are better visualized. The gated method, however, offers images of high quality without cardiac motion artifacts. Therefore reconstruction of cardiac structures could be done without blurring. Further on images corresponding to endiastole signal loss due to turbulent stenotic flow is minimized.

Conclusion: Despite of the exspected difficulties MRA of the cardiovascular system offers reasonable results and improve the diagnostic value of magnetic resonance imaging.

LIPOPROTEIN PROFILES IN HYPERCHOLESTEROLEMIC CHILDREN

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Atherosclerosis is a process that begins in childhood. Coronary heart disease is the result of complex interactions among a variety of risk factors of which hypercholesterolemia is but one.

A routine cholesterol screening of 6,500 children after 3 years of age was carried out in a private pediatric practice over a 2-year period in Cleveland, ohio. Five hundred children were identified to have total cholesterol levels above the 95th percentile of 5.2 mmol/L (200 mg/dL). Lipoprotein profiles were carried out on these children to confirm and delineate their lipid abnormalities.

A definable lipid disorder was present in 85% of this population. Abnormal lipoprotein patterns included 292 children with Type IIA, 99 children with Type IIB, and 25 children with Type IV Phenotypes. An abnormally low HDL cholesterol level of less than 0.9 mmol/L (35 mg/dL) was observed in 20 children.

Only 5% of patients were originally diagnosed as having hypercholesterolemia because they had HDL cholesterols above the 95th percentile of 1.8 mmol/L (70 mg/dL). Thirty-two percent of the children with total cholesterol levels above 5.2 mmol/L (200 mg/dL) had a family member (sibling, parent, aunt, uncle or grandparent) with a myocardial infarction prior to 55 years of age. Data from the study supports universal cholesterol screening after 3 years of age. Lipoprotein profiles are indicated for those children with levels above 5.2 mmol/L (200 mg/dL) or with a family history of premature heart attack or known hypercholesterolemia.

CONTINUOUS DETERMINATION OF OXYGEN UPTAKE AND CARBON DIOXIDE PRODUCTION AFTER OPEN HEART SURGERY IN INFANTS AND CHILDREN

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A new metabolic monitor (Datex® Deltatrac®) allows continuous monitoring of oxygen uptake (VO_2) and carbon dioxide production in the pediatric age group. It is based on a continuous in- and expired gas analysis using a paramagnetic O_2 and infrared CO_2 sensor. Minute ventilation is measured by gas dilution techniques with the same CO_2-sensor.

Purpose of this study was to validate VO_2 measurements of this system by calculating cardiac output using these VO_2 measurements and mixed venous and arterial oxygen saturations and comparing it with thermodilution derived cardiac output.

Patients and methods: Only patients without tube leakage were studied. 16 infants and children aged between 3 days and 16 years after cardiac surgery were studied under controlled, SIMV and CPAP ventilation with a volume controlled Siemens Servo 900C ventilator. VO_2 monitoring was done for 8 hours to 5 days. Arterial and mixed venous blood samples were analyzed with a Radiometer ABL 300 blood gas analyzer and an OSM 3 hemoximeter. Thermodilution cardiac output was measured using the Oximetrix 3 cardiac output computer.

Results: The results indicate a good correlation ($r > 0.95$). Our data did also indicate that frequently in the post operative period oxygen consumption can not be estimated from predictive equations. Continuous measurement of oxygen uptake permits observation of cardiovascular, ventilatory and metabolic adaptation after cardiac surgery.

DISCRETE SUBAORTIC STENOSIS: OPERATIVE AGE AND GRADIENT AS PREDICTORS OF LATE AORTIC VALVE INCOMPETENCE.

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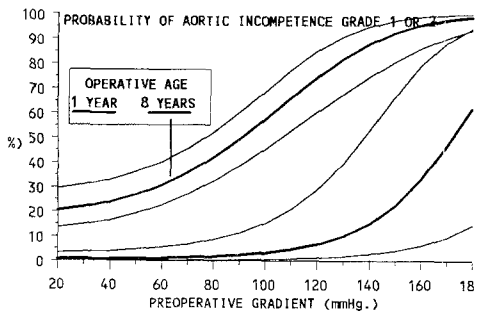
Cardiochirurgia Universita' di Padova.

Between January 1969 and May 1990, 100 patients were operated of discrete subaortic stenosis.



Three patients died perioperatively. We selected 67 surviving patients, with a median follow-up of 60 months, who had no evidence of preoperative or postoperative valvular stenosis and had a recent postoperative hemodynamic or echocardiographic examination. Preoperatively 51 patients had a mild aortic incompetence and 8 patients a moderate one. At follow-up 27 mild and 6 moderate incompetences persisted. The probability of aortic incompetence of the observed grade was analyzed with the multivariate ordinal logistic model. This probability was significantly and independently related to the operative age and gradient as shown in the figure. This finding suggest the opportunity of early repair.

CARDIAC MALFORMATIONS IN THE FETAL RAT ASSOCIATED WITH TRICHLOROETHYLENE ADMINISTRATION IN DRINKING WATER

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Trichloroethylene (TCE) is a common drinking water contaminant which has been implicated as a cardiac teratogen in an epidemiologic study and in a mammalian study in which TCE was delivered to fetuses *in utero*. Our purpose was to determine if TCE delivered in drinking water is a teratogen. Maternal Sprague-Dawley rats imbibed TCE in drinking water. The effect of high (1100 ppm) and low (1.5 ppm) dose TCE administered to 3 groups was examined under the following circumstances: 1) before pregnancy only (for an average of 2 months), 2) during prepregnancy and for the duration of pregnancy, and 3) during pregnancy only. Control maternal rats drank only uncontaminated water. On gestational day 22 (just prior to parturition), control and treated maternal rats were terminated. Fetuses were removed and carefully examined for morphological anomalies. The heart and great vessels were removed for microdissection after close scrutiny of venous and arterial connections *in situ*. The incidence of non-cardiac anomalies was 0.1% - not significantly different from random expected values in Sprague-Dawley rats. One thousand three hundred and ten blindly coded fetal hearts, 232 of which were controls, were studied employing a strict protocol. One hundred and four abnormal hearts were found with a wide array of lesions and with no particular pattern of defects emerging. The incidence of cardiac malformation (CMF) in controls was 3%. Fetal rats exposed to either dose of TCE before pregnancy had numbers of hearts with CMF similar to controls. Fetal rats exposed to both doses of TCE-contaminated water before pregnancy and during the whole duration of pregnancy had a significantly greater incidence of hearts with CMF than controls ($p<.02$). Fetal rats exposed to high dose TCE during pregnancy only had significantly greater numbers of hearts with CMF than controls ($p=.01$), but the group receiving low dose TCE during pregnancy only had a higher precentage of abnormal hearts with CMF than controls but the difference was not significant. These results show a dose response to TCE. This study demonstrates that exposure to TCE contaminated drinking water only in the prepregnancy period is not associated with increased incidence of CMF. However, fetal rats exposed during pregnancy to TCE in maternal drinking water had a significantly greater incidence of CMF than controls whose mothers drank uncontaminated water.

EDGE ENHANCEMENT ANGIOGRAPHY IN CONGENITAL HEART DISEASE

F. Maymone Martins, Isabel Menezes, Rui Anjos and Margarida Telo (Hospital de Santa Cruz, Carnaxide, Portugal)

An increasing number of catheterization laboratories is equipped with Digital Subtraction Angiography. However, the use of edge enhancement angiography for the study of congenital heart disease has seldom been reported. In our laboratory edge enhancement angiography has often been performed by the Sobel gradient in children undergoing cardiac catheterization both for diagnosis and intervention. This type of image processing eliminates the areas with a uniform degree of radiopacity and enhances those where abrupt changes are noted by the pixel values. The resulting filtered images clearly highlight the edges of the opacified chambers, including its sharpest inner changes in thickness and contour, as well as the boundaries of overlapping structures that would otherwise be missed. These are usually first quality, easy to read

images. The main drawback is the need for postprocessing, which, however is feasible while other parts of the catheterization are carried out.

In our experience this technique has proved to be particularly useful to depict vessel contour in specific lesions. These include the demonstration of the coronary arteries in children with Transposition of the Great Arteries being prepared for the Arterial Switch operation, Blalock-Taussig anastomosis with unnoticed stenoses subsequently treated by balloon dilatation, Coarctation and Recoarctation of the aorta, etc.

Edge enhancement angiography is a useful tool in the study of congenital heart disease by cardiac catheterization both for diagnosis and intervention.

ATRIAL SEPTOSTOMY BY PERCUTANEOUS BALLOON DILATION

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Atrial septostomy (AS) by percutaneous balloon dilation (PBD) was attempted instead of blade atrial septostomy (BLAS) in 13 patients (pts) with congenital heart deffects (CHD). Ages ranged from 10 days to 14 months and weights from 2.8 to 8.5 kg. Seven pts had transposition of the great arteries (TGA), 5 pts had left AV valve atresia or stenosis, and one suffered from pulmonary vascular obstructive disease. The size of balloons used varied from 13-15mm. Balloon catheter (7F) entered the left atrium through a patent foramen ovale (8pts) or via transeptal puncture (5 pts). The size of interatrial communication (IC) before AS by PBD was 1.7 $\pm$ 1.6 mm and it was increased to 8.8 $\pm$ 1.5 mm after the procedure (P< 0.001). Transatrial gradients (TAG) improved from 9 $\pm$ 4.3 mm Hg to 1.8 $\pm$ 2 mm Hg with AS by PBD. Arterial oxygenation improved from 37.5 $\pm$ 8.6 % to 73 $\pm$ 6.9% in pts with TGA. Two-dimensional + Doppler echocardiographic evaluation performed in 11 pts 4 to 12 months following AS by PBD showed no significant change in defect size (8.5 vs 8.8, P=NS) and good interatrial blood flow. No pt developed any complication as a result of the procedure. The findings demostrate that AS by PBD is an effective and safe technique from creating an adequate IC.This method may provide an effective alternative to BLAS for palliation of infants with certain types of CHD.

BALLOON VALVULOPLASTY IN INFANTS AND CHILDREN USING CARBON DIOXIDE.

* * *
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We used carbon dioxide gas (CO2) as the contrast agent for balloon valvuloplasty (BV) in 47 infants and children (25 male, 22 female). In addition to the peak systolic withdrawal gradient (PSG) at catheterization, the peak instantaneous gradient (PIG) was also derived using continuous wave Doppler ultrasound 24 hours prior to, and 24 hours after the procedure.

BV of the aortic valve was performed in 20 patients (including 4 neonates) at a median age of 43 months (range 0.07 - 264) using a balloon to anulus ratio (BAR) of 1.15 (SEM ±0.05). The PSG fell from 59 (±5) to 22 (±3) mm Hg (p<0.01). New or increased aortic regurgitation occurred in 13 patients, but was <grade 3 angiographically in all but 2 cases. The PIG fell from 78 (±5) to 34 (±3) mm Hg (p<0.01), and has remained virtually unchanged (mean 40 ±6 mm Hg) over a follow-up period of 5 (±1) months.

In 14 patients with pulmonary stenosis (median age 18 months; range 5-129) and with a BAR of 1.46 (±0.07), BV reduced the PSG from 54 (±9) to 20 (±4) mm Hg (p<0.01), and the PIG from 67 (±7) to 27 (±5) mm Hg (p<0.01). In 1 patient with predominant infundibular stenosis there was no change in the gradient. At follow-up (mean 5 ± 1 months) the mean PIG was 23 (±6) mm Hg.

In 11 patients with Fallot's tetralogy (median age 3 months; mean BAR 1.8 ±0.03) BV of the pulmonary valve produced an immediate increase in systemic arterial oxygen saturation from 75% (±16) to 87% (±12) (p<0.01). Over a follow-up period of 6.6 (±1.2) months 2 patients have required further palliation with an aortopulmonary shunt.

CONCLUSIONS: CO2 allows for rapid balloon inflation and deflation at low inflation pressures, with minimal disturbance of systemic blood pressure and cardiac rhythm. The markedly reduced inflation and deflation times enable the use of larger diameter balloons on smaller shafts (e.g. 8mm balloon on 3.4F shaft) and hence cause less vascular trauma.

AGE AT PRESENTATION IS AN IMPORTANT DETERMINANT OF THE OUTCOME OF BALLOON DILATION FOR AORTIC STENOSIS

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Balloon dilation (BD) of the aortic valve was attempted in 36 children presenting over a 5 year period. The age at presentation was < 1 week in 9, < 1 month in 5, 1 month - 1 year in 8 and > 1 year in 14.

Of the 9 patients presenting < 3 days of age, there were no survivors of BD. All presented with severe congestive cardiac failure and required a prostaglandin infusion. Endocardial fibroelastosis was present on echocardiography. In 3 of these, procedural complications contributed to the fatal outcome: arterial damage in 2 and left ventricular perforation in 1. The other 6 all died between 1 - 185 days after BD from left ventricular dysfunction despite "technically successful" dilations (further augmented by surgical valvotomy in 2). A successful outcome was obtained in 4 of the 5 patients treated between 1 and 4 weeks of age, and have Doppler gradients between 25 to 50 mmHg at a follow up of 1 - 2 years (1 required a surgical valvotomy). The fifth patient died of congestive failure 10 days after BD.

Of the 8 patients treated between 1 month and 1 year of age, 4 are well palliated (1 after a 2nd BD). There was 1 procedural death due to an anaesthetic complication. Surgery was performed in 3 patients: 2 after a 2nd BD and one urgently after valve disruption.

Of the 14 patients treated after the first year of life, 2 had previously undergone a surgical valvotomy. The immediate transvalvar gradients after BD fell from a mean of 66 mmHg (range 40 - 105) to a mean of 29 mmHg (range 10 -40). One patient required a further balloon dilation. There has been no mortality or need for surgery in any of these patients. The Doppler estimated gradients ranged from 30 - 60 mmHg after follow up of 4 months to 4 years.

Thus BD of critical aortic stenosis in neonates is accompanied by a high mortality due to arterial complications and associated left ventricular dysfunction. Presentation beyond the 1st week of life is associated with a better outcome and prognosis. In patients > 1 year of age balloon dilation is an effective and safe palliative procedure that can delay the need for, and supplement surgery.

Anaemia in children following cardiac transplantation - treatment with low dose human recombinant erythropoietin.

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Anaemia is common in cardiac transplant recipients. In a series of 5 children with anaemia outside of the immediate post-transplant period, 1 had a hypochromic, microcytic anaemia due to iron deficiency, which corrected with oral iron. The other 4 children, had normochromic, normocytic anaemias (Hb 8.3-10.0 g/dl), with normal levels of B12, folate and ferritin. All received standard immunosuppression with azathioprine and cyclosporin.

Serum erythropoietin levels were determined by radio-immunoassay and compared with a control group with dietary iron deficiency anaemia.

The 4 children with normochromic, normocytic anaemias had inappropriately low levels of erythropoietin relative to their degree of anaemia. Treatment with subcutaneously administered erythropoietin, 40U/Kg twice weekly, resulted in a significant rise in haemoglobin in all 4 patients, (mean 2.4 g/dl, 95% CI 0.7-4.0 g/dl). This was associated with subjective improvement in wellbeing. Stopping treatment in one patient and dosage reduction in a second, was followed by a fall in haemoglobin to pre-treatment levels and a further significant rise when full dosage was reinstated.

We postulate that cyclosporin is responsible for the low levels of erythropoietin and hence the anaemia. The effect is seen at therapeutic levels of cyclosporin and may be mediated by a toxic effect on erythropoietin synthesising cells in the kidney.

Our findings show that erythropoietin deficiency is an important cause of anaemia in transplant recipients. It should be sought in cases of anaemia refractory to conventional haematinic therapy, since treatment with erythropoietin is both effective and practicable.

(* introduced by DF Dickinson)

INFECTIONS IN PEDIATRIC HEART-TRANSPLANTATION

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Infections are a serious problem in patients undergoing heart transplantation (HTx). A simple transfer of experience from adult to pediatric heart recipients may neglect the properties of the immune system and the perioperative care in pediatric patients. The incidence and mode of infectious complications were analyzed in 13 pediatric patients with a mean age of 2.7 years; the follow-up time of surviving patients was 10-41 months. The immunosuppression consisted in cyclosporine, azathioprine and prednisolone; the latter were withdrawn between 1 and 12 months after HTx. 3 proven (positive cultures from normally sterile sites) and 3 probable (clinical and radiological signs of infection, positive C-reactive protein, negative cultures, no rejection) bacterial infections were observed during the first 30 days after HTx in 6 of 8 surviving patients. Pseudomonas aeruginosa (2) and Staph.aureus (1) were isolated in the proven infections. All bacterial infections affected the bronchopulmonary system, manifested within the first 4 days after HTx and required prolonged artificial ventilation. Prophylaxis with CMV-hyperimmune globulin and acyclovir resulted in 1 primary, but asymptomatic CMV-infection in 3 CMV-negative recipients of CMV-positive donor organs. In addition 1 secondary, symptomatic CMV-infection with hepatopathy was successfully treated with CMV-hyperimmune globulin. Both CMV-infections appeared after a trial of ATG during the first month after HTx. 2 primary and 1 secondary infections were due to Epstein-Barr virus, the former associated with mild mononucleosis-like symptoms. Until now there is no evidence of lymphoproliferative disease in these patients. The high incidence and the early onset of bacterial infections suggest a major influence of the preoperative colonization and the vulnerability of the lung to perioperative lesions beyond immunosupression. These results contrast to previous reports and underscore the pecularities of the pediatric age group.

HETEROTOPIC HEART TRANSPLANTATION IN INFANCY

Kececioglu, D., Konertz, W., Weyand, M.,Frank, T., Scheld, H.H.

Surgery of the Bland-White-Garland (BWG)-syndrome in infancy is associated with an operativ risk of 40-70%.

A 4 month old infant with BWG-syndrome was seen in frank left ventricular failure due to a large anterior myocardial infarction and extensive posterior ischemia evidenced on stress thallium scintigraphy. Surgery was performed when an appropriate donor heart became available. Revascularization of the heart was accomplished with a tunnel repair from the aorta to the left coronary ostium in the pulmonary artery and patch reconstruction of the main pulmonary artery. The donor heart was transplanted heterotopically as left ventricular assist. Recovery from surgery was uneventful. Repeat scintigraphic studies one month postoperatively documented the absence of any ischemia in the heart with an increase of LV- EF from formerly 30% to 54%. Repeat angiographic studies showed an increase of LV-EF from 30% to 60%, cardial index was 3,6 1/min.x m^2. Because there is evidence that myocardial reparative processes and remodelling will continue the assist is still in place, however, in case of further improvement of the native left ventricle the deliberate removal of the graft seems to be indicated.

To the best of our knowledge this is the first report of succesfull infant heterotopic heart transplantation.

PATTERNS OF DIASTOLIC VENTRICULAR DYSFUNCTION IN CHILDREN TREATED FOR ACUTE LYMPHOBLASTIC AND MYELOID LEUKAEMIAS. DOPPLER ECHOCARDIOGRAPHY IDENTIFIES DIFFERENCES BETWEEN TREATMENT GROUPS

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Changes in Doppler echocardiographic (echo) measures of left ventricular (LV) diastolic function occur with anthracycline chemotherapy; their clinical relevance has been unclear. We have therefore examined diastolic function in 2 groups of children with similar diseases but different treatment regimes.

Results from serial echo examinations of 38 children with acute lymphoblastic leukaemia (ALL) were compared with those of 12 children with acute myeloid leukaemia (AML) and to matched controls. The ALL patients were treated with up to 380(median 370)mg/m^2 of Daunorubicin (Dauno). The AML patients received up to 500(median 300)mg/m^2 of Dauno, usually followed by 50mg/m^2 of Mitoxantrone, +/- high dose cyclophosphamide and total body irradiation for bone marrow transplantation (BMT).

One child with ALL died from a dilated cardiomyopathy after 270 mg/m^2 of Dauno; one with AML developed similar problems after 450mg/m^2 but survives on 'anti-failure' medication. Two AML patients died from low output cardiac failure at 1 month after Mitoxantrone therapy. Asymptomatic systolic dysfunction (LV ejection fraction <60%) developed in 5 AML patients; systolic function of the rest of the ALL group was normal.

Pulsed wave Doppler echo measurements of transmitral early (E) and atrial (A) phase filling velocities, the EA ratio and the isovolumic relaxation time (corrected for heart rate, IVRTC)), were used to assess diastolic function. The previously described fall in (median(IQR) EA ratio at low anthracycline doses was seen in both groups, from 1.7(1.4-2.2) (normal) to 1.12(1.06-1.6) in ALL & 1.4(1.27-1.67) in AML groups respectively ($p = <0.05$ Mann-Whitney U Test). EA ratio then rose with increasing Dauno dose to 1.35(1.25-1.78) in the ALL patients ($p = 0.08$), but fell further to 1.2(1.07-1.43) ($p = 0.008$) following Mitoxantrone & BMT for AML. The initial fall in EA ratio was due both to a fall in E & rise in A velocities. The further fall seen in the AML group was from a further fall in E velocity, whereas E remained constant & A fell in the ALL group. Changes in IVRTC mirrored those of EA ratio. Thus the AML regime exacerbated the diastolic relaxation abnormality produced by lower dose anthracycline therapy, whereas the ALL patients developed a pattern of diastolic dysfunction more consistent with altered ventricular compliance.

Doppler echo assessment of diastolic function discriminates between the cardiotoxic effects of different antineoplastic regimes. The insights provided into underlying pathophysiology may give useful guidance for the prevention & treatment of systolic decompensation.

DIASTOLIC FUNCTION IN THE FETUS: A DOPPLER-ECHOCARDIOGRAPHIC STUDY

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To determine changes of ventricular diastolic function in the human fetus, velocity waveforms of tricuspid and mitral valves were

studied longitudinally in 48 fetuses from 14 weeks gestation to term. Doppler tracings were analyzed for: peak early (E) and peak late (A) inflow velocities, time velocity integral (TVI) of total inflow and A-wave velocity waveforms. Isovolemic relaxation time (IVRT) was measured from the outflow/inflow signal and presented as a proportion of the R-R interval.

E-velocity, A-velocity and E/A ratio of both valves increased significantly with gestational age ($p<0.001$). Heart rate decreased significantly with gestational age ($p<0.001$). Total and A-wave TVI increased across both valves ($p<0.001$). The ratio of A-wave TVI to total TVI of both valves was constant as well as heart rate corrected IVRT.

This study demonstrated that the passive and atrial portions of ventricular filling did not change as a percentage of total filling during second and third trimester, even as velocities increased. The increasing E-wave velocity with unchanged heart rate corrected IVRT could be the result of an increasing atrio-ventricular pressure gradient with increasing blood flow in the growing heart 2and does not necessarily imply changes in ventricular compliance.

TWO-D ECHOCARDIOGRAPHIC (ECHO) AND DOPPLER CHANGES IN THE MAIN PULMONARY ARTERY (MPA) AND ITS BRANCHES IN NEW BORNS WITH FUNCTIONAL SYSTOLIC MURMURS

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Transient systolic murmurs radiating to the axillae are common in neonates and premature infants. Mild left (LPA) and right (RPA) pulmonary branch stenosis has been demonstrated but follow-up studies are lacking. We studied 32 neonates (NN) with echo and Doppler in the first month of life. 21 NN (6 prematures) with a murmur (group I) were compared with 11 controls (no murmur) matched for age, weight, height and heart rate (group II).
2-D echo diameters of the MPA and the RPA were slightly smaller in group I (PA 7.2 ± 1.1 vs 7.8 ± 1.2 mm ; RPA 3.7 ± 0.7 vs 4.2 ± 0.8 mm, NS). LPA was significantly smaller in NN with a murmur (3.7 ± 0.8 vs 4.6 ± 0.9 mm, $p<0.05$). Doppler flow velocities were significantly higher in group I with a mean peak gradient of 15 ± 4 mmHg ($p<0.0001$) in LPA and 12 ± 5 mmHg ($p<0.0001$) in RPA. The follow-up study 3 months later in 15/21 (71 %) patients of group I showed absent or decreased murmur in 10 (67 %). 2-D echo diameters increased from 3.9 ± 0.6 to 4.9 ± 0.8 mm ($p<0.0001$) in LPA and from 3.7 ± 0.5 to 4.8 ± 0.7 mm ($p<0.0002$) in RPA. The ratio LPA/MPA increased significantly from 0.51 ± 0.07 to 0.60 ± 0.14 and the ratio RPA/MPA from 0.50 ± 0.06 to 0.60 ± 0.15 with a $p<0.05$, suggesting accelerated growth or dilatation of the PA branches compared to the trunk. Flow velocities decreased significantly from 1.8 ± 0.3 to 1.3 ± 0.3 m/s ($p<0.0002$) in LPA and from 1.7 ± 0.3 to 1.3 ± 0.3 m/s ($p<0.005$) in RPA. Turbulent flow was no longer present in the 10 patients who lost the murmur. Thus a functional murmur in NN is associated with a temporary relative hypoplasia of the PA branches leading to accelerated blood flow. Increased relative growth of these branches leads to disappearance of the murmur in most of the cases within 3 months of life.

ECHOCARDIOGRAPHIC SPECTRUM OF MITRAL VALVE ABNORMALITIES IN COARCTATION OF THE AORTA

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Purpose of this study was to examine the incidence of mitral valve abnormalities in patients with aortic coarctation (COA) with or without ventricular septal defect (VSD). Echocardiographic studies from 88 consecutive newborns with COA between January 1988 and October 1991 were reviewed. Mean age at presentation was 17 days in cases with and 85 days in cases without VSD. Mitral valve abnormalities were arbitrarily divided into 4 grades: grade 0: normal Grade 1: thickening of valve leaflets plus shortening of chordae with normal Doppler flow; grade 2: marked shortening of chordae with thickenend lateral and small anterior papillary muscle and grade 3: grade 2 changes plus stenotic Doppler flow and/or hypoplastic mitral valve ring or single papillary muscle with stenotic flow. There had been 46 cases with intact ventricular septum (group A) and 42 with VSD (group B). Results are listed in the table:

	Grade 0	Grade 1	Grade 2	Grade 3
Intact ventricular septum (n=46)	21	18	5	1
VSD (n=42)				
infundibular (n=7)	2	4	1	---
perimembranous (n=21)	2	13	5	---
muscular (n=5)	2	1	1	1
multiple VSDs (n=9)	1	4	2	2

One case with intact ventricular (and atrial) septum had a cleft mitral valve. In the VSD group 5 had additional subaortic stenosis (Shone complex) and 2 had valvar aortic stenosis. All had grade 2 or 3 mitral valve abnormalities. One patient with VSD and grade 2 changes died. He also had pulmonary vascular disease Changes grade 1 and 2 did not require surgery so far. Mitral valve surgery was done in 2 grade 3 cases and was deferred in 2 cases because of hypoplastic valve ring. We conclude that mitral valve in isolated COA is either normal or only slightly abnormal; severe mitral abnormalities are more prevalent in cases with VSD especially in cases with multiple defects or Shone complex and negatively influence morbidity.

BICUSPID AORTIC VALVE AS PART OF A MORE COMPLEX CONGENITAL CARDIOVASCULAR MALFORMATION

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Bicuspid aortic valve may be found as an isolated anomaly or in combination with other congenital cardovascular malformations. In the present study we evaluated the associated anomalies and the morphologic details of the semilunar valves in 64 post-mortem heart specimens with a bicuspid aortic valve.The patients' age of death ranged from 1 day to 24 years, with a median of 21 days. The sex ratio was 36 males vs 28 females. Raphes, observed in 38 cases, were situated predominantly 29/38 between the origins of the right and left coronary arteries in the sinus facing the pulmonary orifice. Dysplasia of the valve was found in 8 cases. With regard to the associated malformations there was a striking predominance of left-sided abnormalities. All hearts had left ventricular abnormalities (64) comprising abnormal muscular structures and mitral valve abnormalities (33). The latter anomalies (malattachment, and/or hypoplasia, displacement) were most often seen (30/33) concomitant with an anterolateral muscle (ALM) and/or anteroseptal twist (AST). In 16 cases the mitral valve anomalies were correlated also to a posteromedial muscle. AST and ALM caused left ventricular outflow tract narrowing in 30 cases. Right ventricular abnormalities were observed in all cases with pulmonary valve anomalies (10). Ventricular septal defects were present in 35 specimens. Seven cases showed multiple VSD's. Most defects (21) were related to the outflow tract. Other VSD's comprised central muscular defects (11), AVSD (2), muscular inlet defects (8).The bicuspid aortic valve was fre-

quently (45/64) associated with aortic arch anomalies such as coarctation, hypoplasia, atresia and interruption.Taken the frequent association of left sided anomalies into account, bicuspid aortic valve appears to be part of a more complex cardiovascular malformation.The classical view, that hemodynamics play a crucial role in development of bicuspid aortic valve is debatable. The role of a neural crest cell population which is essential for aortic arch and outflow tract formation, should be considered as a possible common denominator as well.

FRESH FROZEN VEIN HOMOGRAFT VERSUS GORE-TEX PROSTHESIS FOR SYSTEMIC PULMONARY SHUNTS IN NEWBORNS AND INFANTS. PRELIMINARY RESULTS.

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Two kinds of systemic-pulmonary shunts were compared in 16 babies aged from 6 days to 7 months. All had lung oligaemia due to either pulmonary atresia or severe pulmonary stenosis in complex congenital heart defects.

Nine children were operated on between January 88 and March 91, in 8 babies a Gore-Tex prosthesis was implanted, and the ninth had a classic Blalock-Taussig shunt. There were 2 early post-op deaths (22%) - one from shunt failure and the other from low output syndrome.

From March 91 till October 91 fresh frozen vein homografts were used for shunt creation in 7 babies with similar defects. There was neither shunt failure nor post-op death. The vein homograft, being more elastic than the Gore-Tex prosthesis, was technically easier to implant - particularly in the presence of very narrow pulmonary arteries.

Our preliminary results suggest that, in newborns and infants, fresh frozen vein homografts are superior to Gore-Tex prostheses in systemic pulmonary shunts.

FIXED SUBAORTIC STENOSIS (FSAS) - LONGTERM POSTOPERATIVE RESULTS

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Postoperative evolution of FSAS is freqently unfavourable and unpredictable. From 1980 to 1990 50 children aged 10,1+/-4(1,3-16 yrs), preop. peak LV/Ao gradient (GR) 81,5+/-28,5mm Hg (5-145) were operated. Accompaning CHD were present in 19 (38%). One pt aged 1,3 yrs with FSAS, MST, Ao hipoplasia and CoAo died in the operating theatre. 40 children were followed up for more that 6 mts-mean 3,4+/-2,3 yrs (0,5-10 yrs). They were devided into 3 groups according to the following criteria:

Results / Criteria	Favourable	Satisfactory	Unsatisfactory
Clin. symptoms ECG ghanges Ao insufficiency (AoI) LV/AoPG (Doppler)	Asymptom. +/- LV hyper trophy (LVH) No or NS <25 mmHg	Asymptom. LVH or LABE, LBBB mild to moderate 26 - 49 mmHg	Symptomatic LV strain or 3°AVblock significant >50 mmHg
No of pts (%)	16 (40%)	15 (37.5%)	9 (22.5%)

Postoperative results were affected mainly by
1. Development and progression of AoI;
2. Development of restenosis;
3. Progression of the residual stenosis.

Results were dependent on preoperative status, accompaning CHD (especialy valvular AoS and Ao hypoplasia), the operative procedure used and duration of the follow-up.

FSAS shows a tendency of unfavourable postoperative evolution and pts need careful and close postoperative follow-up. The results of this study make us reevaluate the indications for operation in FSAS and the surgical procedure used.

PERMANENT CARDIAC PACING IN CHILDREN

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Indications and results of permanent cardiac pacing in 72 children aged 4 mo to 17 y are presented.
Indications for cardiac pacing were a-v heart block / congenital or acquired/ in 59 pts , sick sinus syndrome in 9 pts, long QT syndrome in 3 pts.
The unipolar pacing systems were used in all cases , in 71 VVI and in 1 DDD pacemakers were implanted.
Epicardial leads were used in 48 pts , and endocardial in 24 pts.
Follow-up period ranged from 7 mo to 10 y 10 mo.
In 19 children 23 reoperations were done from different causes with exchange of pacing system in 8 pts.
Postpericardiotomy syndrome was observed in 14 pts with epicardial leads.

66 children are alive , doing well, 6 pts died in spite of effective stimulation.

DEATH IN INFANCY FROM UNRECOGNISED CONGENITAL HEART DISEASE

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Previous studies of the incidence of congenital heart disease have been limited by selection bias and by poorly defined catchment populations. Babies who died from undiagnosed congenital heart disease have necessarily not been included. This study was performed to identify deaths from unrecognised congenital heart disease in infancy and to determine how many of these deaths could be avoided.

In a retrospective study we identified all infant deaths in the Northern Region of England in 1985-1990. Data were obtained from the Regional Surveys of Perinatal, Late Neonatal, and Infant Mortality and of Fetal Abnormality and from hospital case notes and autopsy reports. Congenital heart defects were designated extremely severe (ESCHD) in the presence of an absent or atretic valve or an absent chamber, whilst hearts with four chambers and four valves were considered less severe (LSCHD).

The Northern Region has a population of 3,085,700. In 1985-90 there were 242,766 live births and the total

infant mortality was 2081 (8.6 per 1000). 158 babies (7.5%) died with congenital heart disease, of whom 109 (69%) had been seen by a cardiologist and 49 (31%) died undiagnosed. Of these 49, 15 had major non cardiac abnormalities which discouraged further investigation. Of the remaining 34, 13 had ESCHD whilst 21 had LSCHD. In the less severe group babies presenting with cyanosis (transposition, Fallot's tetralogy etc) died at a mean age of 12 hours whilst those with heart failure (coarctation etc) died after a mean of 9 days.

Our data suggest that in about a third of all infant deaths from congenital heart disease the lesion is unrecognised in life and at least half of these are babies are treatable. Earlier recognition of cardiac problems in the neonate could lead to a reduction in mortality.

INCIDENCE AND PREDICTIVE FACTORS OF RHYTHM DISTURBANCES AFTER FONTAN ANASTOMOSIS

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Clinical data, pre-op and all post-op electrocardiograms (ECG) along with 24 hour Holter monitoring of 37 consecutive patients surviving a Fontan anastomosis for more than 30 days were reviewed; 21 had tricuspid atresia and 16 other forms of univentricular heart. Age at surgery was 10.7±5.5 years; follow-up duration 4±2.6 years. Rhythm disturbances appeared in 20 patients (54%) during the follow-up period; early in 12 (< 30 days after surgery) relapsing later or persisting in 5, late in 8. Arrhythmias were: atrial flutter (AF) 10, atrial tachycardia 3, junctional tachycardia 3, sinus bradycardia 2, atrioventricular (AV) block 1. AF appeared early after surgery in 7 patients with relapse later in 2, it was associated with a sinus node disease in all of the 7 patients with AF after the early post-op period. A pacemaker was implanted in 10 patients (27%); 9 for sinus node disease (7 AF), bradycardia being exacerbated by antiarrhythmic treatment; 1 for AV block. Four patients (11%) died, 2 suddenly (1 with AF and implanted pacemaker, 1 without previous arrhythmia) and 2 from other causes. Type of cardiac malformation, age at surgery, arrhythmia in the immediate postop period did not predict long term rhythm disturbances. Follow-up duration was longer in patients with arrhythmia (5.6±2.8 vs 3.4±2.3 years, $p<0.05$). On the pre-op ECG, amplitude of negative deflection of the P wave in V1 was greater in patients with arrhythmia (1.8±0.13 vs 1.0±0.14 mv, $p<0.05$). Incidence of rhythm disturbances after Fontan anastomosis is high with sinus node disease (with or without AF) being by far the most frequent. Risk of sudden death always exists even whitout previously detected arrhythmia. Incidence of arrhythmia increases with follow-up duration. Anomaly of the P wave on the pre-op ECG can help predict patients at risk for post-op rhythm disturbances.

PULMONARY ARTERIAL SUPPLY IN PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT.STUDY WITH DIGITAL SUBTRACTION ANGIOGRAPHY

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To assess the accuracy of Digital Subtraction Angiography (DSA) in the demonstration of pulmo-arterial supply (PAS) in Pulmonary Atresia with Ventricular Septal Defect (PAVSD) 14 patients (pts) aged 0.5 to 174 months (mean 79.6) were studied. Clinical and 2D-Echo/Doppler investigations were performed prior to cardiac catheterization. In all children 0.3 to 1.8 ml/kg of contrast was injected in the aorta and in 8 pts. 0.1 to 0.6 ml/kg in major aortopulmonary collateral arteries (MAPCAS) or in surgical shunts (shunts) (total: 0.4 to 5.2ml/kg). DSA images were stored in video and cut films. Immediate replay of good quality images avoided unnecessary injections.

Results: 12 pts had confluent pulmonary arteries (PA) and two had only one branch PA. 10 pts had PA supplied by MAPCAS, 2 by Ductus Arteriosus and 2 by shunts. Detailed anatomic information was obtained in all cases, the quality of images being excellent in 12 and acceptable in two.

Conclusion: DSA is a good technique (more informa tive in video) for evalution of PAS in PAVSD.

SYMPTOMATIC HEART DISEASE IN INFANCY: COMPARISON OF 3 STUDIES PERFORMED DURING THE YEARS 1969-1987

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We compared three studies of symptomatic or hospitalized infants with congenital heart disease - the New England Regional Infant Cardiac Program (NER), 1967-1974 (2,251 infants), The Brompton Hospital series (BH), 1973-1982 (1,619 infants) and the Northern Great Plains Regional Cardiac Program (NGP), 1982-1987 (4,735 infants). Within the first 2 weeks of life, the percentage of infant admissions has decreased from 45.1% for NER to 34.1% and 34.6% respectively for BH and NGP ($p<0.001$). In both BH and NGP, the foremost cardiac anomalies during the 1st month of life were complete transposition (15.8% and 15.4%), coarctation of the aorta (13.4% and 12.3%), hypoplastic left ventricle (8.7% and 10%) and Tetralogy of Fallot (5.7% and 11%). For the period 0-6 months, the major conditions were ventricular septal defect, complete transposition, coarctation of the aorta and Tet. During the last 6 months of the 1st year of life, VSD and Tet were the most common conditions. Among infants in the NER< mortality was 40% for the years 1969-1974 and NGP was 20% for the period 1982-1987. About 1/3 of all infants included in these series were hospitalized during the 1st week of life, with at least 1/4 of the infants hospitalized within the 1st 2 days of life. Despite some differences in diagnostic categorization, the frequency of the different cardiac conditions among symptomatic infants is essentially the same over the 20 years encompassed by the 3 studies. Some changes have occurred in the specific age presentation, but these are most likely related to changes in the pattern of care.

MRI-FEATURES OF PULMONARY HYPERTENSION IN CHILDREN WITH CONGENITAL HEART DISEASE

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Refering to the wellknown radiological features of pulmonary hypertention - i. e. criteria of right ventricular pressureload and hypertrophy as well as dilated main pulmonary arteries in connection with decreased peripheral vascular markings - we investigated 12 children with pulmonary hypertension, proven by pressure-measurement, in a MRI study.

As cause of pulmonary hypertension there existed ventricular septal defects, frequently

in combination with other congenital heart defects, as were transposition of the great arteries, coarctation of the aorta and truncus arteriosus communis.

The follwing convincing MRI-findings we were able to demonstrate:
- **right atrial and ventricular hypertrophy and dilatation,**
- **strongly dilated main pulmonary arteries,**
- **pruning of the intrapulmonary arteries (calibre breaking off),**
- **pathologic relationship of the diameters truncus pulmonalis versus aorta ascendens**
- **reduced cross-sectional diameters of the peripheral pulmonary arteries.**

Medial hypertrophy and intimal fibrosis of the pulmonary vessels could be visualisatised by their marked wallthickness.

Moreover we succeeded in detailed representation of most associated heart malformations.

CLINICAL SIGNIFICANCE OF MATERNAL SS-A ANTIBODIES IN CHILDREN WITH ISOLATED HEART BLOCK.

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Isolated heartblock (HB) in children is a well known entity. In some HB patients an association with maternal SLE, especially with anti SSA-antibodies (ab) has been reported.
To assess the significance of this association all children (N=30) with isolated HB known in our hospital since 1975 were studied.
The medical history was reviewed and related to the ECG variables. All mothers were screened for the presence of anti SS-A ab by means of a RNA precipitation assay.
21 children, 2 being siblings, proved to have an anti SS-A positive mother (group A), 9 children an anti SS-A negative mother (group B). A summary of the results is given in the table.

	group A (n=21)	group B (n=9)	p
prenatal diagnosis	18 (86%)	0	<.001
mean age at postnatal diagnosis	5 mo.	4.3 yrs	<.001
Caesarian section or Vac. Extr.	11 (52%)	2 (22%)	<.05
mortality	3 (14%)	0	n.s.
3rd degree block initially	17 (81%)	6 (66%)	n.s.
pacemaker insertion	8 (40%)	6 (66%)	n.s.

In total 4 of the positive anti SS-A mothers had a pregnancy history with evidence of foetal heart block.

We conclude that isolated heart block in children is in 70% of the cases related to maternal anti SS-A ab and has a specific clinical presentation, of which the early onset with considerable mortality is the most striking.
In addition this study demonstrates that the recurrence risk for HB in the offspring of mothers with positive anti SS-A ab is increased.

Session IV: Interventional Cardiology
Chairman: S. Quereshi

DOES PULMONARY BALLOON VALVOPLASTY PRODUCE MORE VALVE REGURGITATION THAN SURGICAL VALVOTOMY?

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Continued use of balloon pulmonary valvoplasty (BPV) requires knowledge, not only that the procedure is equally effective to surgical valvotomy, but also that it produces no deleterious effects from valve regurgitation. The results of treatment of pulmonary valve stenosis by percutaneous balloon valvoplasty (30 patients) and surgical pulmonary valvotomy (28 patients) were compared using ultrasound ; to determine the residual transvalvar gradient and the degree of pulmonary regurgitation. Before intervention the gradient distribution was statistically comparable (two sample t-test, p = 0.92). There was a statistically greater reduction in the transvalvar gradient in the surgical group (from 65.2 ± 37.9 to 13.1 ± 6.5 mmHg after a follow up of 5.4 ± 2.0 years) than in the PBV group (from 64.3 ± 28.2 to 23.7 ± 10.9 mmHg after a follow up of 2.0 ± 0.9 years) (analysis of covariance, test of equality of adjusted means, p < 0.0005). The surgical group was found to have a greater degree of pulmonary regurgitation as assessed by diastolic backflow in the pulmonary artery and larger right ventricular size (X2 tests, p < 0.0001, p < 0.012 respectively).

Although PBV may produce a lesser degree of reduction of valve gradient this is not clinically significant. The lesser degree of residual regurgitation from PBV may be an advantage in the longterm. This study justifies the continued use of BPV for isolated pulmonary valve stenosis.

THERAPEUTIC EMBOLIZATIONS IN CONGENITAL HEART DISEASE

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Percutaneous transcatheter embolization of 37 cardiovascular anomalies was attempted in 29 patients, aged 8 months to 67 years (median 8 years).
Ten patients underwent embolization of 16 aorto-pulmonary colaterals. Embolization materials included steel coils (14 vessels), detachable balloon (1), and gelfoam (1).
Seven patients had Blalock Taussig shunts made redundant after surgery or balloon pulmonary valvoplasty. These were embolized with coils in 4 patients and detachable balloons in 3.
Eight patients had 10 coronary artery fistulas (9 congenital and 1 acquired). Embolization was attempted with standard steel coils in 2 cases, microcoils in 4 cases, detachable balloons in 3 and a combination of detachable balloon and coils in 1.
Other anomalies treated with embolization included pulmonary arteriovenous malformations (2 cases, embolized with coils), a patent arterial duct (treated with coils) and a fistula from an internal mammary artery to pulmonary veins (embolized with coils).
There were no major complications associated with the procedures. In one patient with a coronary fistula there was deflation of a detachable balloon after it had been well positioned. In the remaining patients complete occlusion of the vessels or shunts was obtained, except in two in whom substantial reduction of flow was achieved.

Therapeutic embolization is an effective alternative to surgery in patients with selected cardiovascular anomalies.

MORPHOLOGICAL DETERMINANTS OF SUCCESSFUL COMPLETE OCCLUSION OF ARTERIAL DUCTS USING THE RASHKIND UMBRELLA DEVICE.

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The shape and size of the patent arterial duct was assessed from lateral aortograms of 101 patients who have had attempted transcatheter duct occlusion with the Rashkind device. The shape of the duct was classified into one of the five categories below. Patients with a previous Rashkind device or previous ligation were excluded. The frequency of complete occlusion on Doppler examination at late follow-up was calculated for ducts of each category and for each occluder size as follows. The position of the occluder on the post implantation angiogram was assessed.

Duct Type	Overall	Complete occlusion 12mm device	17mm device
1)Broad Funnel	38/50 (76%)	30/37(81%)	8/13 (62%)
2)Short (no ampulla)	8/10 (80%)	6/8 (75%)	2/2 (100%)
3)Parallel tubular	12/19 (63%)	3/8 (38%)	9/11 (82%)
4)Complex	3/4 (75%)	2/2 (100%)	1/2 (50%)
5)Tubular with narrow pulmonary end	16/18 (89%)	15/17 (88%)	1/1 (100%)

Duct size	Overall	Complete occlusion 12 mm device	17mm device
$\leq$ 2mm	27/33 (82%)	27/33 (82%)	-
>2mm$\leq$3mm	33/41 (80%)	27/35 (77%)	6/6 (100%)
>3mm$\leq$4mm	14/22 (64%)	2/4 (50%)	12/18 (67%)
>4mm$\leq$5mm	2/3 (67%)	-	2/3 (67%)
>5mm	1/2 (50%)	-	1/2 (50%)

In 8 patients the final position of the occlusion device was sub optimal and 6 of these (75%) there was a residual leak on Doppler.

Complete occlusion with a single Rashkind device is most frequent in the smallest ducts. However the shape of the duct also influences outcome and ducts without a discrete narrowing (parallel tubular) are associated with a high incidence of residual flow. Malposition of the occlusion device is an important additional cause of a poor result.

SIX YEARS' EXPERIENCE OF TRANSCATHETER OCCLUSION OF PATENT ARTERIAL DUCTS WITH ONE OR TWO UMBRELLA DEVICES .

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Of 140 patients in whom transcatheter occlusion of a patent arterial duct with an umbrella device was attempted since 1986, a first device was successfully implanted in 127 patients (91%). In the remaining 13, the duct had unusual anatomy in 2, was too small in 6 (2 previously surgically ligated), and was too large in 3. Abnormal systemic venous anatomy precluded passage of the introducer sheath in 2.

The age at implantation ranged from 7 month to 67 years (median 2.87 years) and weight from 6.15 to 79.6Kg (median 13.7Kg). Two patients had had a previous Senning operation and 5 had had previous duct ligation. A 12 mm device was implanted in 88 patients and 17 mm device in 39 . A complete arterio-venous guidewire circuit was established in order to position the sheath and dilator in 7 patients.

One device was embolised to the femoral artery. One implanted device was removed from the duct surgically because of severe haemolysis. No case of infective endarteritis occured.

Eighty nine patients(70%) had complete occlusion confirmed by colour Doppler study after implantation of a single device and 38 (30%) did not. Eighteen (47%) of these 38 have a follow-up of less than 4 months and 20 (53%) of more than 4 months.

Seventeen patients with residual leak subsequentlly had attempted implantation of a second device and implantation was achieved in 16. The duct in the remaining patient could not be dilated sufficiently to allow passage of the delivery system. After a mean follow up of 8 months, 13 ducts are completely occluded on colour Doppler study. Three ducts still have a small residual leak one day, 6 weeks and 18 months after the second insertion.

Transcatheter occlusion of a patent arterial duct is feasible and implantation of a second umbrella device is sometimes needed to achieve complete occlusion .

ANGIOPLASTY OF THE ARTERIAL DUCT IN NEONATES WITH INTRACTABLE HEART FAILURE SECONDARY TO SEVERE LEFT HEART OBSTRUCTIVE LESIONS.

* * * *
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Five neonates (age 2 - 8 days; weight 2.2 - 3.9 kg) presented with intractable heart failure due to severe obstruction of the left heart. Three of them had aortic arch interruption or atresia, one had hypoplastic left heart syndrome with aortic valve atresia, and another had critical aortic valve stenosis with hypoplastic left ventricle. All patients had severe metabolic acidosis (pH <7.1) and oliguria (urine output <0.5ml/kg/hour) at initial assessment. Despite resuscitation with intravenous prostaglandin E2, sodium bicarbonate and dopamine all continued to have a low cardiac output with persistent acidosis, oliguria and absent femoral pulses. Cross sectional Doppler echocardiography showed a patent though restrictive arterial duct (all 5 patients) and poor right ventricular function (4 patients).

Angioplasty of the arterial duct was performed using a percutaneous femoral venous approach. A 10mm diameter balloon was passed through the arterial duct over a guide wire positioned in the descending aorta. Between 1 and 3 inflations with carbon dioxide gas were performed in each case. Following the procedure, the gradient between the pulmonary artery and descending aorta fell from 11.2 (SEM ±2) mm Hg to -7 (±2) mm Hg ($p<0.01$). Pulmonary arterial angiography showed a widely patent duct in all patients.

The patients' clinical condition improved dramatically after the procedure. Peripheral perfusion increased with strong femoral pulses, adequate diuresis, and disappearance of metabolic acidosis (arterial pH improved from 7.03 ±0.06 pre angioplasty to 7.48 ±0.3 post angioplasty - $p<0.01$, and the base deficit improved from -22 ±2 to 8 ±2; $p<0.01$). Cross sectional Doppler echocardiography showed improved right ventricular function with a widely patent and unrestrictive duct. It was thus possible for the patients to undergo surgical treatment in better condition than could have been achieved by conventional resuscitation.

CONCLUSIONS: Establishing wide patency of the arterial duct is the mainstay of resuscitation in neonates with obstructive left heart lesions. Where this has not been achieved with prostaglandin infusion, angioplasty offers an attractive alternative. It may also expand the therapeutic options available for neonates with the hypoplastic left heart syndrome.

DUCTAL STENTING AND PULMONARY ARTERY BANDING - A NEW APPROACH TO PALLIATION OF HYPOPLASTIC LEFT HEART SYNDROME

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Neonatal heart transplantation offers some hope for babies with hypoplastic left heart syndrome but is severely limited by the scarcity of donors. An extension of the waiting time would increase the chance of a transplant but conventional palliative surgery has a very high mortality. We have developed a new palliative programme involving open septectomy and bilateral pulmonary artery banding followed by transvenous stenting of the ductus to avoid long term prostaglandin infusion.

Two neonates with hypoplastic left heart syndrome presented at 3 days of age. Each deteriorated at 4 weeks of age and underwent bilateral pulmonary artery banding and open atrial septectomy. One to two weeks later cardiac catheterisation was performed. Pulmonary artery angiography was used to measure the position, diameter and length of the ductus. Palmaz-Schatz biliary stents (Johnson & Johnson) with an expanded diameter of 4-7 mm and length of 20 mm were used. Half of a 20 mm stent was mounted with a crimping tool on a catheter with a compliant balloon 7 mm diameter and 20 mm length (Cordis). The stent was positioned at the aortic end of the duct and deployed by inflating the balloon to 10 atmospheres pressure for 5-10 seconds, achieving a diameter of 7.7 mm.

One baby, with a 12 mm long duct, required 2 stents, overlapping by 1-2 mm. In both the prostaglandin was stopped and anticoagulation with heparin and warfarin was continued for one month.

This new combined medical and surgical approach to palliation of hypoplastic left heart syndrome greatly increases the chance of successful heart transplantation by extending the stable waiting period without drug treatment.

THE PALLIATIVE ROLE OF BALLOON DILATION IN CHILDREN UNDER TWO YEARS OF AGE WITH TETRALOGY OF FALLOT

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Since March 1988, balloon dilation (BD) of the right ventricular outflow tract (RVOT) was performed in 22 children with tetralogy of Fallot under 2 years of age, mean 8.1 months and weighing between 2.6 and 10.7 Kg, mean 6.45 Kg. Successful dilation was achieved in 21 patients. The balloon/annulus ratio ranged between 1.2 and 1.7, mean 1.5. The aortic oxygen saturation increased from mean(SD) 78(12)% before to 91(5)% after BD ($p<0.05$), whilst the pulmonary artery saturation increased from 65(10)% before to 80(7)% after BD ($p<0.05$). Infundibular perforation with a guidewire occurred in 1 patient. One patient with CHARGE association had a cardiac arrest before and died soon after a successful BD. During a follow-up of 1.5 to 20 months, palliation has been adequate in the survivors except two with associated atrioventricular septal defects, who required BT shunts 8 and 9 months after BD. Repeat angiography 1.5 - 10 months (mean 5) after BD showed an increase in pulmonary valve annulus diameter from 7.1(2.2)mm before to 9.8(2.4)mm after BD. Branch pulmonary artery diameters increased from 6.9(1.9)mm on the right and 6.7(2.6)mm on the left before to 8.9(1.7)mm and 8.0(2.6)mm respectively after BD ($p<0.05$). Eleven of the patients have undergone surgical correction 3-19 months after BD. Tearing of a pulmonary valve cusp was noted in 5 pts. There were no tears of the pulmonary artery or its branches. In 8, there was extensive scarring of the infundibulum.

Thus balloon dilation of RVOT results in an increase in pulmonary valve annulus and pulmonary artery size. It achieves adequate palliation, and is a good alternative to shunt operation in children with tetralogy of Fallot.

CATHETER ABLATION OF ACCESSORY ATRIO-VENTRICULAR PATHWAYS BY RADIOFREQUENCY CURRENT IN CHILDREN

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Radiofrequency current (RFC) catheter ablation of accessory atrioventricular pathways (AP) was performed in 4 children. All patients had frequent sustained episodes of drug-refractory supraventricular tachycardia. Mean age was 11.4 (6.7-14) years. Of the 4 patients, 2 children had overt preexcitation due to a left posterior and a left lateral AP, respectively. The remaining 2 patients exhibited only retrograde conduction over the AP, located parahissian and left posterior. RFC was applied through a 6F steerable catheter positioned against the mitral or tricuspid anulus. Criteria for RFC application were recording of an accessory pathway potential or an interval of the local atrial potential to the local ventricular potential <40 msec. RFC (temperature-guided at 70°C at the tip-electrode) was delivered during sinus rhythm in overt APs and during SVT in concealed APs. Conduction over the AP was completely eliminated in 3 patients. In the remaining patient, antegrade refractory period of the AP was significantly prolonged and supraventricular tachycardia was self-limited after the procedure. In the patient with the parahissian AP, induction of complete av-block was inevitable. No other complications occurred, serum creatine kinase activity remained within normal limits in all 4 patients. After a mean follow-up of 4.5 months, all patients remained free of symptoms.

Conclusion: RFC is highly effective in ablating accessory atrioventricular pathways in children.

Session V: Long-Term-Follow Up
Chairman: F.M. Picchio

PREDICTIVE FACTORS FOR SPONTANEOUS CLOSURE OF ATRIAL SEPTAL DEFECT DIAGNOSED DURING THE FIRST 3 MONTHS OF LIFE.

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The widespread use of echo-doppler in infants has shown interatrial shunts to be more common than previously thought. However, the incidence of spontaneous closure of secundum atrial septal defect (ASD) and disappearence of interatrial shunts has never been determined on a large series of patients. One hundred and one infants (age 0 to 3 months, mean 26 days) referred for non specific cardiac murmur in whom the presence of an interatrial shunt was confirmed by echo-doppler, were followed for an average of 265 ± 190 days. ASD diameter (D) defined by the two-dimensional image or by width of the color-flow jet whenever ASD was too small to allow clear visualization, was measured in the subcostal view. Right (RV) and left ventricular (LV) diameters and atrial septal curvature were also studied. There was no significant correlation between ASD diameter and RV/LV ratio, or type of septal curvature (vertical or concave towards the left atrium). The classic predominance of girls over boys was observed only for defects greater than 3 mm. An overall rate of spontaneous closure of 87% was observed. Frequency and timing of closure were inversely correlated to ASD diameter: 100% (32/32) in group 1 ($D<3$mm), 87% (39/45) in group 2 ($3\leq D<5$mm), 80% (16/20) in group 3 ($5\leq D<8$mm). Spontaneous closure did not occur in the 4 patients of group 4 ($D\geq 8$mm) after an average follow-up of 417 days (range 294 to 597). Kaplan-Meier curves were obtained to predict age of spontaneous closure in relation to initial ASD diameter. These results suggest that infants with ASD <3 mm need not be followed as 100% will be closed by 18 months of age, those of 3-5 mm and 5-8 mm should be evaluated respectively by the end of the 12th and 15th month when more than 80% will be closed. ASD ≥ 8 mm may have little chance of closing spontaneously and the possibility of a surgical correction should be entertained. Considering their natural evolution and sex distribution, defects < 3 mm probably do not constitute a cardiac malformation.

QUALITY OF LIFE: LONG TERM FOLLOW-UP AFTER OPEN HEART SURGERY FOR CONGENITAL HEART DISEASE

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To evaluate quality of life long time after open heart surgery in patients with congenital heart disease operated in our institution before the age of 15, between 1968 and 1980, an extensive psychological and cardiological follow-up study was started.
The cohort consisted of 712 patients. Diagnosis was atrial septal defect (ASD II) in 135, ventricular septal defect (VSD) in 176, tetralogy of Fallot (TOF) in 142, transposition of the great arteries (TGA) in 90 (Mustard-repair), pulmonary stenosis (PS) in 61, aortic stenosis (AS) in 22, and miscellaneous in 86. Age at operation ranged from 0.1 to 14.9 yrs, mean 4.7 yrs. Follow-up ranged from 9.3 to 22.9 yrs, mean 14.3 yrs.; 54% was male, 46% female. Of the 498 participants (82% of the survivors) 445 were seen by both the pediatric cardiologist and the psychologist.
Results: 84% judged their health as good, 14% as moderate, and 2% as poor. According to the Warnes/Somerville ability index 73% was in class I, 24% in class II, and 3% in class III. Bicycle ergometry revealed a decreased exercise tolerance in 25% of the ASD II patients, 30% in VSD, 39% in TOF and 56% in TGA. Echocardiographically right ventricular dilatation was present in 92% in TOF, 67% in PS and 26% in ASD II.
Holter registration demonstrated arrhythmias in 72% of the Mustard patients, 42% of the ASD II patients, and 24% of the VSD patients. According to Kugler's critera for sinus node dysfunction (SND), the percentage of SND in these patient groups were respectively 70%, 33% and 13%. Ventricular arrhythmias (Lown grade III) were present in 1% of the patients after Mustard repair, 1% of the ASD patients, and 5% of the VSD patients.
Conclusion: although in this unselected population a substantial number of patients have hemodynamical and electrophysiological sequelea, the clinical significance of these findings is often unclear, and the overall quality of life is good.

IDIOPATHIC DILATED CARDIOMYOPATHY IN THE YOUNG : NATURAL HISTORY AND PROGNOSIS FACTORS. A TWENTY YEARS EXPERIENCE.

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A retrospective study was done to assess the natural history of idiopathic dilated cardiomyopathy (IDC) in pediatric patients, and to identify prognostic factors .
From january 1970 to december 1988, 103 patients aged one day to 14 years (mean 37 months) were managed for IDC, ranging 52 males (50,5%) and 51 females (49,5%).
Patients presented predominantly with acute congestive heart failure (40,8% of cases); severe complication revealed the disease in 22,3% of the cases (neurological embolism, ventricular arrythmia, syncope, collapsus). All patients received medical treatment for congestive heart failure. Mean follow up is 61,9 months (one day to 19 years).
Fourty-one patients died (39,8%) mainly for refractory cardiac failure (48,8% of deaths) or sudden death (20% of deaths). Of these deaths, 70% occured during the first year of illness, and 56% within the first three months.
Sixty-two patients are still alive (60,2%) : 39 are completely cured (37,8% of the population), 2 received orthotopic transplants (1,9%) and 21 have variable degree of chronic invalidity (20,3%). The six months survival is 77,5%, the one year and five years survivals are respectively 70% and 60%.
As previously reported, mortality was greater for patients more than 2 years old at presentation, than for patients less than 2 years old (56,6% versus 32,8% : p=0,025).
Familial myocardial pathology was found in 15,5% of the cases; the incidence of death in this group was significatively increased, compared to the other patients (p=0,033).
No other clinical, radiological or echocardiographic parameter had predictive value.
Thus, tardive onset of the illness and familial context of myocardial disease are poor prognosis factors in idiopathic dilated cardiomyopathy of young people.

RESTRICTIVE CARDIOMYOPATHY IN CHILDHOOD

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Six children (1.4 to 9.7 years old, mean 5.3 ± 3.7 y) with restrictive cardiomyopathy were studied. All patients had marked left atrial dilatation on ECG. Echocardiografic examination showed gross left atrial enlargement on M-mode (186 ± 27% of BSA predicted values, p<0.001), along with normal left ventricular cavity dimensions (99 ± 6%, p:ns). Left ventricular wall thickness varied from normal to mild concentric hypertrophy (septum: 113 ± 11%, p<0.05). There was mild eccentric hypertrophy at the apex in 3 patients. Global left ventricular systolic function was normal or subnormal (fractional shortening 30 ± 3%, range 27% - 35%), but relaxation was significantly delayed throughout diastole. At 200 msec after the minimum end-systolic value, left ventricular dimension had increased by only 56 ± 7% (range 49 - 66%) of the total diastolic expansion (age-matched controls: 84 ± 4%, range 77 - 92%, p<0.001). Marked ventricular filling occurred in mid-diastole as could be deduced from a prominent mid-diastolic mitral flow velocity wave (L wave). All patients had a mild degree of retrograde pulmonary hypertension (mean 29 ± 10 mmHg). The left atrial pressure was 20 ± 4 mmHg with LVEDP 29 ± 5 mmHg. The left ventricular pressure curve showed a steady decline during mid-diastolic filling. This suggests the restrictive hemodynamics not to be caused by increased intrinsic stiffness of the ventricular wall, but to result from a serious dysfunction and delay of the active energy dependent relaxation of the cardiomyocytes. Light microscopic studies in 4 patients showed myocyte hypertrophy, mild loose, mesenchymal interstitial fibrosis and myofiber disarray. No specific ultrastructural lesions were observed on electron microscopic examination in 2 patients. Heart failure responded poorly to medical therapy. Progression of the disease was observed in all patients, resulting in death (2 pts) or heart transplantation (2 pts). None of the parents had evidence of a cardiomyopathy.
Restrictive cardiomyopathy in childhood appears to be a variant of hypertrophic cardiomyopathy. There is no marked macroscopic ventricular hypertrophy or evidence of arrhythmia, but the diastolic dysfunction with delayed relaxation reaches extreme values. Gross left atrial dilatation, delayed ventricular relaxation and a marked L wave due to mid-diastolic filling is typical for this clinical entity. This disease in childhood is associated with a progressive deterioration and a grave prognosis.

How Prevalent is Right Ventricular Dysfunction After Atrial (Mustard) Repair? Are There Serious Clinical Implications?

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Advanced right (systemic) ventricular dysfunction (RVD) is an acknowledged post-atrial repair (ATR) complication. Concern has increased with recent surgical efforts to salvage such patients by heart transplantation (HTX) or conversion to arterial repair (ART).

We assessed prevalence of significant RVD reflected in 210 patient angiographic volume studies (ANGIO): a) before atrial or arterial repair (87); b) after atrial repair (98); and c) after arterial repair (33). Right ventricular ejection fraction (RVEF) measurements indicate that most ATR patients (84%) had significant RVD based on lower 2 standard deviation (-2SD) limits for normal systemic LVEF. Even with changing to (-2SD) for normal pulmonic RVEF, 44% had significant RVD. The mean postoperative RVEF for the ATR subset was significantly reduced compared to preoperative RVEF, normal RVEF and ART LVEF ($p < .001$); $t = -6.66$, -10.66 and -17.65 respectively).

For 85 ATL patients, we mapped long-term clinical follow-up status (2y to 20y) against the above earlier ANGIO. A "troubled" subset (27 patients) was identified: dead (11), tricuspid valve replacement (4), ART (1), HTX (3), or symptomatic (8). Of these 27 patients 26 were readily discriminated by their mapping outside of the ±2SD co-distribution of normal RVEF, RVEDV values and the distribution of the "untroubled" subset (chi-square = 13.42, $p<.001$).

In this ANGIO study abnormal RV volume parameters were prevalent across the spectrum of TGA patients having ATR (1966-1985), and almost the rule for patients in the "troubled" subset. The roles of operative era (myocardial preservation) and postoperative interval (very late deterioration) remain to be clarified.

LONG-TERM OUTCOME OF PATIENTS UNDERGOING COMPLETE REPAIR OF TETRALOGY OF FALLOT AT THE MAYO CLINIC: FOLLOW-UP AT 29-34 YEARS.

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Late follow-up on 160 out of 163 (98.5%) operative survivors undergoing complete repair of tetralogy of Fallot at the Mayo Clinic from 1955-1960 was obtained. Follow-up was a minimum of 29 years or until death, and a maximum of 34 years. Twenty-seven patients had a prior Blalock-Taussig shunt and 11 a Waterston or Pott's shunt with 125 having had no palliative surgery.

Kaplan-Meier estimates of actuarial survival of operative survivors were 95%, 92%, 92%, 91%, 87%, and 86% at 5, 10, 15, 20, 25, and 32 years postoperative respectively. Comparison with the expected survival curve from an age and sex matched population showed survival only slightly less than expected. Only those 56 patients in the age group of 12 years or greater at the time of operation showed a significantly worse actuarial survival curve than their matched control group (76% vs. 93%). Patients with no prior palliation or a Blalock-Taussig shunt had similiar actuarial survivals (87% and 86%), but a Waterston or a Pott's shunt patient had a significantly worse 30 year survival of 73%. The predictors of late mortality on multivariate analysis were operative age 12 years or greater ($p=0.035$), a high (≥ 0.5) RV/LV systolic pressure ratio post repair ($p=0.019$) and a history of preoperative heart failure ($p=0.013$). Transannular patching did not affect late survival to 30 years. The most common cause of late death (10 of 22 patients) was sudden unexpected death, presumably due to a cardiac dysrhythmia.

Long-term follow-up of tetralogy of Fallot patients corrected at the Mayo Clinic during the earliest era of cardiac surgery is most gratifying with excellent survival rates and a high quality of life reported.

Session VI: Imaging
Chairman: A. Bozio

INFLUENCE OF CHANGES IN PLACENTAL RESISTANCE ON FLOW DYNAMICS OF THE FETAL AORTIC ISTHMUS.

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During fetal life, the aortic isthmus (AoI) is the only vascular link between the two ventricles which work in parallel. This study investigated the effects of an elevated placental resistance (EPR) on both the flow velocity profile and the amount of blood passing through the AoI. Four levels of mechanical umbilical vein compression (UVC) were achieved on 7 exteriorized fetal lambs. Arterial blood gases and pressures were monitored through a femoral artery catheter. Blood flows in the AoI and the umbilical artery (UA) were measured by the echo Doppler technique. Three additional fetuses were submitted to the same protocol without UVC and were subsequently used as controls. In the presence of an EPR causing a fall in UA blood flow of 30%, retrograde diastolic flow was observed in the AoI; the net isthmic flow was still forward but dropped significantly (from 91 ± 26 to 47 ± 31 ml/kilo). At the same time, a forward diastolic flow was still being recorded in the descending aorta and UA. With a more severe increase in placental resistance (fall in UA blood flow of 75%), diastolic reverse flow increased and the net blood flow through the AoI was either zero or retrograde. A strong positive correlation ($r=0.89$) was found between AoI and UA blood flows. No change was observed in the control group during the entire period of observation. It is concluded that variations in Doppler blood flow velocity waveforms and integrals in the AoI could be used as sensitive indicators of the state of the umbilical circulation, especially in fetuses with intra-uterine growth retardation.

BIPLANE TRANSESOPHAGEAL ECHOCARDIOGRAPHY IN INFANTS AND CHILDREN, LONGITUDINAL VERSUS TRANSVERSE PLANE

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A pilot study was performed to determine the feasibility and additional diagnostic value of biplane transesophageal echocardiography (TEE) in 36 anaesthetized infants and children with congenital heart disease during cardiac surgery, diagnostic or interventional catheterization or only for diagnostic purposes. In 26 pts (bodyweight 12-65 kg) a 13x9 mm biplane probe was used and in remaining 10 pts (bodyweight 4,4 - 12 kg) 2 separate 7 mm single plane (longitudinal and transverse) probes were used sequentially.

Results

In 35/36 pts the probe(s) could be inserted. The longitudinal plane appeared to be superior in defining both LV and RV outflowtracts. Furthermore this plane was very helpful during balloonvalvuloplasty of the pulmonary valve in one patient. Previously unknown diagnostic information using this plane was obtained in 4/35 pts. The transverse plane, on the other hand, remained essential in assessment of size and position of ventricles, A-V valves and great arteries relative to each other.

Conclusion

1. Biplane TEE with currently available probes is feasible in a high percentage of paediatric pts. In infants the problem of transducer size can be circumvented by insertion of 2 separate longitudinal and transverse plane probes sequentially.
2. Although the longitudinal plane provides important additional diagnostic information, it cannot subtute the transverse plane, but is complementary.

(*) introduced

BIPLANE INTRAOPERATIVE TRANSESOPHAGEAL ECHOCARDIOGRAPHY IN CONGENITAL HEART DISEASE

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Intraoperative monoplane transesophageal echocardiography (TEE) is useful for evaluating the results of cardiac surgery. Biplane TEE may be more valuable in congenital heart disease (CHD). To assess its utility, we reviewed our first 60 patients (pts) who had biplane intraoperative TEE. Five MHz probes with diameters of 10 to 12 mm were used. The indication for biplane intraoperative TEE was to assess the integrity of the repair, usually of complex CHD, valvular regurgitation or intracardiac shunts. Biplane TEE findings were compared to all available pre- and postoperative diagnostic studies and to the surgeon's observations. Pts ranged in age and weight from 2 to 49 yrs (mean=9 yrs) and from 7 to 70 kg (mean=26 kg). Biplane TEE detected all significant anatomic abnormalities diagnosed by other methods. Preoperative biplane TEE findings modified the planned repair in approximately 10% of cases (6 pts). Postoperative biplane TEE revealed significant residual abnormalities in 4 pts (2 stenotic Fontan anastomoses, 1 iatrogenic VSD, 1 subaortic obstruction and 1 restrictive ASD). Availability of an orthogonal view simplified the definition of complex anatomy. The longitudinal plane was superior to the horizontal plane in imaging the ventricular outflow tracts, ascending aorta, cavopulmonary anastomoses and left pulmonary artery. Intraoperative biplane TEE did not consistently diagnose tiny (<3 mm) residual VSDs observed on a later postoperative echo (4 of 8 were correctly identified). There were no complications secondary to TEE. Biplane TEE allows rapid assessment of surgical repairs and immediate correction of significant residual defects. Current probes allow biplane intraoperative TEE in all but the smallest pts (7 < kg). Biplane TEE is an accurate, safe and valuable addition to the intraoperative care of pts with complex CHD, valvular regurgitation and VSD.

MEASUREMENT OF CARDIAC OUTPUT IN NEONATES WITH DUAL BEAM DOPPLER : COMPARAISON WITH CONVENTIONAL PULSED DOPPLER ECHOCARDIOGRAPHY

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The dual beam doppler (DBD) is a reliable method to measure the ascending aortic flow in adult. To evaluate this new technique in néonates we performed paired DBD and doppler echocardiography (DE) cardiac output (CO) measurements in 16 newborn babies. Commercially available paediatric equipment was employed : quantascope (vital science) with 4MHz annular array transducer for DBD measurements and 5MHz duplex doppler echocardiography transducer (ATL Lab.) for DE measurements.
Conditions remained steady with no differences of heart rate between both the measurements. The range of DBD measurements (212.5ml/kg/min. to 406ml/kg/min., mean 268.6ml/kg/min.) were not different from DE measurements (209ml/kg/min to 417ml/kg/min., mean 270.3ml/kg/min.).There was a significant correlation between the two methods (r = 0.96, p<0,01). The mean difference between paired measurements was 1.6ml/kg/min. with 95 % confidence limits from - 8 to 11.2ml/kg/min., being 7.1 % of mean CO. No patient had a discrepancy over 15 % between the two methods. We found that the DBD was a rapid and simple method in comparaison with the DE.
We conclude that DBD is a convenient instrument to determine CO in neonates and correlates well conventional pulsed DE technique.

ULTRASONOGRAPHIC BRAIN CHANGES IN INFANTS AFTER HEART SURGERY

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We prospectively assessed brain changes in infants after heart surgery using cranial ultrasonography.

The study group consisted of 27 full term neonates and infants (16 male,10 female) with congenital heart disease,without other malformations,aged 6 to 90 days (mean=30±22.6 days) when they were operated upon; 13 of them underwent open heart surgery.
One preoperative brain scan was performed no longer than one week before surgery; postoperative scans were obtained one week and 3 to 6 months after the operation. One infant died on the early postoperative period and was excluded from the study.

Of the 26 patients, 23 (88%) had preoperative normal brain scans but only 10 (38%) had postoperative examinations wich were classified as normal and 9 (34.6%) remained so up to 6 months. Combinations of hypoxic-ischemic lesions and intracranial hemorrhage were the most commonly displayed abnormalities and the surgical methodology did not influence their occurrence.

This preliminary study has shown that heart surgery in early life is related to either the appearence or further development of ultrasonographic brain changes. Clinical follow-up, currently in practice,is expected to clarify their long-term impact on the neurodevelopment of these patients.

MAGNETIC RESONANCE IMAGING IN SUPRAVALVULAR PULMONARY STENOSIS AFTER THE ARTERIAL SWITCH OPERATION.

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Magnetic Resonance Imaging (MRI) has been shown to be a valuable tool in demonstrating the anatomy of congenital heart disease. Since the delineation of the Main Pulmonary Artery (MPA) and its branches is frequently incomplete with Color Doppler Echo-cardiography (CDE), we evaluated the use of MRI to identify and localize pulmonary stenosis. MRI (cardiac triggered spin-echo sequence) was performed in 29 patients after the arterial switch operation (ASO), 6 had undergone the Jatene and 23 the Lecompte procedure. A narrowing of more than 75% was considered to be significant. In 4 patients this was found in the MPA, as predicted by CDE. In 4 patients, all after the Lecompte procedure, stenosis was found in the left pulmonary artery, of whom 2 had narrowing of the right pulmonary artery as well. In these patients CDE could not accurately localize the site of the stenosis. These findings were confirmed by cardiac catheterisation. However in one patient MRI missed a membrane-like narrowing of the MPA, which was suggested to be present by CDE and confirmed by cardiac catheterisation.
MRI is useful in non-invasively detecting and localizing supravalvular pulmonary stenosis after the arterial switch operation. MR-velocity flow mapping would probably have detected the membrane-like stenosis.

Session VII: Miscellaneous
Chairman: E. Harinck

TOWARDS THE IDENTIFICATION OF A GENE MUTATION RESPONSIBLE FOR CONGENITAL HEART DEFECTS AND SITUS INVERSUS IN A MOUSE STRAIN

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One route to understanding the etiology and pathogenesis of complex disorders such as congenital heart defects is to identify genes that predispose individuals to the disease. Studying animals models have been proven to be useful. Comparative genetic maps between human and other mammals have been used for prediction of locations of human disease genes on the basis of their locations in the animal model. We are trying to identify a gene mutation called "*iv*" responsible for a situs inversus with congenital heart defects in mouse. This mutation is autosomal recessive. Despite homogeneous genetic background (the mouse strain has been bred sister-brother for more than 20 generations) and standard environmental factors, the *iv* mutation is expressed with incomplete penetrance and variable expressivity:
- 50% of two *iv/iv* homozygote parents are phenotypically normal.
- Among the other 50% mice, a wide variety of situs inversus is observed associated with various heart defects : ASD, VSD, TGA, DORV.
We set up a cross to obtain backcross animals: *IV/IV* X *Mus musculus m.* --> F1; F1 X *IV/IV* -->backcross mice. 581 backcross animals were checked for situs inversus (situs of the heart, aortic arch, lung lobulation, azygos arch, liver, stomach and spleen situs, position of the superior mesenteric artery). 120 (20,7%) mice were phenotypically abnormal and presumed homozygote for the *iv* mutation. The mice were tested with polymorphic markers :
- pMI3 is a probe derived from the "Cystein Rich Intestinal Protein" gene. 5 out of 115 mice were heterozygotes (4,34%).
- A microsatellite sequence located within the heavy chain immunoglobulin genes detected 6 heterozygotes out of 111 (5,4%).
- pYNZ2 is a human probe containing repetitive sequences. It detects multiple bands on Taq 1 digested mouse genome DNA, one of which is specific for *Musculus m.* and segregates in 18 out 102 mice (17,6%).
With the *linkage* softwares, we can order these markers: *iv*, CRIP, IghC, YNZ2 and estimate their genetic distance. Additional markers are currently tested. These genetic markers will be useful to study human families with recurrent cases of situs inversus.

OXYGEN DEFICIT DURING CONSTANT-LOAD EXERCISE IN CHILDREN AFTER CARDIAC SURGERY IN COMPARISON WITH HEALTHY CHILDREN.

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The exponential increase in oxygen seen at the beginning of a constant-load exercise develops into a steady-state, where the oxygen uptake corresponds to the demands of the tissues. Thus, during the initial period, an oxygen deficit arises and energy must be supplied by means of oxygen stores, by phosphagens or anaerobically. The velocity of the increase in oxygen uptake at the beginning of a constant-load work determines the magnitude of the oxygen deficit and is characterized by the half-time $t_{1/2}$, i.e. the time needed until half of the final oxygen uptake is attained.

We determined $t_{1/2}$, oxygen deficit, maximal blood lactate and maximal oxygen uptake ($\dot{V}O_2$ max) in boys aged 11 - 12 years at exercise levels corresponding to between 80 and 90 % of their $\dot{V}O_2$ max. The same measurements were performed in 40 children after surgical closure of a VSD, after corrective surgery for Fallot's tetralogy and after Senning- or Fontan-procedures. Oxygen deficit and $t_{1/2}$ were nearest the values found in healthy children in patients after closure of a VSD, whereas children after Fontan-operation or with a transannular patch after correction of Fallot's tetralogy had the most unfavourable oxygen uptake transient kinetics. A negative correlation was evaluated between $\dot{V}O_2$ max and $t_{1/2}$, whereas VO_2 max and maximal blood lactate showed a positive correlation.

Conclusion: Many children with decreased $\dot{V}O_2$ max after cardiac surgery also demonstrate unfavourable oxygen uptake transient kinetics. In addition to being less qualified for endurance performance, they are also less prepared for those types of exercise where energy must be produced anaerobically.

THE USE OF PROPAFENONE IN TREATMENT OF TACHYARRHYTHMIAS IN CHILDREN.

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Propafenone was given to 60 children (mean age 4.5yrs) with paroxismal reentrant supraventricular tachycardia (rSVT:41 cases), postoperative automatic junctional tacycardia (JET: 8 cases), automatic atrial tachycardia (AT:4cases), ventricular tachycardia (VT: 4 cases), and atrial flutter (AF:3 cases). Acutely propafenone was administered intravenously (mean dose 1.3±0.5 mg/kg) to 29 patients, cronic oral treatment (mean dose of either 11±3.3 mg/kg/day or 265±78 mg/m2/day) was given to 48 children. Overall efficacy were: 76% for acute and 69% for chronic treatment, with best results in rSVT (effectiveness of 89% acutely and 69% chronically. We try to identify any factor leading to a lack of response. Multivariate analysis showed that only factor predictive of chronic inefficacy was age: propafenone was ineffective in 61% of children <1 year old, but only in 26% of older patients. It is worth noting that the younger children recived doses body surface area which were lower than those given to older children: 206.5 mg/m2 and 307 mg/m2 respectively (p 0.01). No difference was found when dose levels were based on body weight. Side effects were observed in 25% of patients: 6% required suspension of therapy due to a: proarrhythmic effect in 1 case on intravenous administration, peripheral neuropathy 1 case, hypotension in 2 postoperative JET patients. At ECG, we observed a lengthening of PQ interval, no significant lenthening of the QTC or QRS intervals were recorded. Propafenone also reduced mean heart rate as showed by Holter monitoring. In conclusion: its efficacy and limited side effects make propafenone a safe and very useful drug in the treatment of tachyarrhythmias

EFFECT OF TREATMENT WITH BETA-ADRENERGIC BLOCKERS ON ADRENORECEPTORS. ITS IMPACT ON POSTOPERATIVE CARE.

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Beta-adrenergic antagonists are administered in some infants with tetralogy of Fallot and hypoxemic spells. Their usage can influence the density of beta-adrenoreceptors. A positive correlation was found between the maximal binding capacity of adrenoreceptors and response to catecholamines.

Maximal binding capacity of the beta-adrenoreceptors was measured in right ventricular tissue of 17 children with the tetralogy operated on at the mean age of 4.3 (1-15) years, nine of them after chronic administration of metipranolol (MTA- a nonselective beta blocker). The binding capacity was measured in crude tissue homogenates using ^{125}I-pindolol, after subtracting the nonspecific binding.

Mean value of the maximal binding capacity in pts treated with MTP (13.8, SD ± 5.1 fmol/mg protein) did not differ significantly from the value measured in pts operated without previous treatment with MTP (13.7, SD ± 4.7 fmol/mg protein). Postoperative ventilatory support was slightly shorter in pts treated with MTP (29.6 hours) than in nontreated pts (32.3 h.). A low dose of catecholamines was administrated routinely for 39.9 postoperative hours in the MTP pts and for 39.4 hours in pts without MTP. It was no difference in the response to catecholamines between both groups. One patient died of intractable repeated bronchial bleeding on the 8th postoperative day.

In conclusion, chronic administration of a nonselective adrenoreceptor antagonist did not alter the maximal binding capacity of the beta-adrenoreceptors in the right ventricle of pts with tetralogy of Fallot. The postoperative course, response to catecholamines and outcome of surgery were not influenced by the treatment.

PULMONARY ARTERY SIZE AND THE MODIFIED FONTAN OPERATION

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139 Fontan patients had their clinical outcome correlated with the size of their pulmonary arteries as determined by preoperative angiograms. Followup information was available in all patients for at least six months. Clinical outcome was assessed using three endpoints: 1) hospital death or take-down of the Fontan repair within 30 days or before hospital discharge, 2) death or take-down within the first six months, and 3) pericardial and/or pleural effusions persisting 18 days or greater after surgery or requiring pericardiectomy or pleurodesis within six months of the Fontan operation. Hospital mortality and early take-down for the 139 patients was 12.9%. Six-month cumulative mortality and take-down was 16.5%. 17.3% of patients had a prolonged effusion. Although there was a trend toward better outcome with larger pulmonary artery indices, it was not statistically significant. However, in a low risk subset of 30 patients with tricuspid atresia, those with smaller pulmonary arteries (PAI 185±47 mm^2/m^2) had more prolonged effusions than those with larger pulmonary arteries (PAI 276±82 mm^2/m^2) (P=<0.01). Futhermore, in this low risk subset, patients with a pulmonary artery index of <250 mm^2/m^2 had a failure/effusion rate of 43% while those with pulmonary artery index >250 mm^2/m^2 had no prolonged effusions or failures (P<0.01).

REHABILITATION OF CHILDREN AFTER CARDIAC SURGERY:MULTINATIONAL METHODOLOGY AND RESULTS.

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Aim of the study:to verify the improvement in aerobic fitness in patients(P) with significant residual heart diseases (s/p Mustard or Fontan operation,total repair of Tetralogy of Fallot)after hospital-based programs of cardiac rehabilitation.Materials and methods:prior to initiation of training and at the end, all patients undergo:exercise test(E T),measuring maximal heart rate,exercise time, blood pressure and, if possible,oxygen consumption,carbon dioxide production,minute ventilation,and cardiac output by rebreathing,Holter monitoring, echocardiography and lung function tests at rest.In one center,submaximal stres test on teradmill(SET), neurological and psychological parameters are also measured.Training period:12 weeks,2-3 sessions/week, each lasting 1 to 1 1/2 hours (subdivided in:warm-up/stretching,submaximal aerobic exercise and cool-down).Intensity of exercise: 60-80% of the peak heart rate attained during the pertraining ET.Results:in Italy:22 P (range 5-16 y)had statistically significant increase in SET (p<0,002);increase in maximum ET not significant.P self-image and other psychological parameters show meaningful improvement.In USA :34 P(range 4-16y). There was statistically significant improvement in oxygen uptake(p<0,05),cardiac output(p<0,05),exercise time(p<0,05 and systolic blood pressure(p<0,05).In both centers compliance was greater than 86%,reflecting the good feelings that P and their families get form the program.These results confirm that in different centers with culturally different populations,cardiac rehabilitation can successfully improve the aerobic fitness of these patients.

Session VIII: Mitral Valve
Chairman: U. Sauer

DEVELOPMENTAL PATHOLOGY OF THE MITRAL VALVE.

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In serial sections of 78 human embryos, age 3 to 10 weeks, and in scanning electron micrographs of 15 rat embryos, age 10 to 16 days, mitral valve development was studied. In the youngest stages, no indication of a valve-like structure was present, except a mass of cushion tissue which was plugged in the atrioventricular canal. Instead of a well-defined tension apparatus, the left ventricle contained an irregular trabecular network interconnecting the walls of the cavity. Any lengthwise orientation of the myocytes, as was clear in these trabeculations, was absent in the free wall which was still very thin. The contraction force was, therefore, expected to be generated by the trabecular network. Somewhat later, the free wall had already become thicker, anticipating its future contractile role. During development of the outflow tract ("leftward migration of the aorta") the trabeculations were seen to coalesce and to become more unidirectional, and at the same time subendocardial layers of myocardium were peeled off from the septal region to form a flap-like structure between the main ventricular cavity and the outflow tract. Numerous connections between the flap and the septum and free wall were present to show the delamination process on its way. The latest stages showed a smooth-walled outflow tract and a mitral valve and tension apparatus all arranged along the long axis of the ventricle, indicating a contraction pattern differing completely from the primitive stage.

In autopsy specimens, most forms of malattachment of the mitral valve could be correlated with the above developmental morphology: leaflets, chords and/or papillary muscles were attached to the septum and/or the anterior left ventricular wall. Mitral valve pathology was nearly always accompanied by abnormal ventricular architecture, i.e. anomalous muscle bundles and localized hypertrophy.

It is concluded that in congenital malformations of the mitral valve the morphology is reminiscent of developmental stages of the left ventricle and, therefore, is not optimally adapted to the mature contraction pattern which needs a general long axis arrangement.

CONGENITAL MITRAL VALVE AND LEFT VENTRICULAR MALFORMATIONS ASSOCIATED WITH OSTIUM SECUNDUM ATRIAL SEPTAL DEFECT AND/OR ABNORMAL PULMONARY VENOUS DRAINAGE.

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Association of mitral valve (MV) anomalies and ostium secundum atrial septal defect (ASD) as well as abnormal pulmonary venous drainage (APVD) are described in the literature. The possible pathogenetic mechanism underlying this combination is not known. In this study it was hypothesized that the left to right shunt at the atrial level might influence the development of the MV and the left ventricle (LV).

43 autopsy specimens were examined (13 with ASD, 16 with APVD and 14 with ASD and APVD). MV and LV morphology was analyzed and compared to 20 normal hearts. Only 3 specimens had normal MV and normal LV, 9 had normal MV but abnormal LV. The remaining 31 showed abnormal MV as well as abnormal LV.

The MV was abnormal in 9/13 hearts with ASD (69 %), in 11/16 hearts with APVD (69%) and in 12/14 hearts with both ASD and APVD (86%). The LV was abnormal in all hearts with ASD, with or without APVD. Among 16 hearts with APVD, only 3 had a normal LV.

The spectrum of main malformations comprised LV abnormalities: antero-septal twist (AST), abnormal outflow trabeculations, thickened anterior wall, LV hypoplasia and/or hypertrophy; MV abnormalities: compression ("pinching") by surrounding LV structures ,particularly AST, abnormal tendinous chords, malattachment, underdevelopment of interpapillary space. In some cases the mitral leaflets were dysplastic.

Thus, hearts with ASD and/or APVD show LV and/or MV malformations. The distribution of these anomalies suggests that the interatrial shunt might play a role in the maldevelopment. Although the incidence of malformations in this autopsy study is not necessarily similar to clinical series, in patients with ASD and/or APVD particular attention is warranted to associated MV and/or LV anomalies.

CONGENITAL MITRAL VALVE STENOSIS.
FUNCTIONAL CLASSIFICATION AND CONSERVATIVE SURGICAL MANAGEMENT

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Lesions' complexity and associated anomalies make surgical treatment of congenital mitral stenosis difficult.
Between 1976 and 1991, 34 patients were operated on for congenital mitral stenosis in our institution. 29 patients underwent mitral valve repair using Carpentier's techniques. Mean age at operation was 5 years (range 5 months to 12 years). A functional classification based on echocardiography and intraoperative observations was as follows.
Type A Normal papillary muscle (N = 20)
1) Commissure to papillary muscle fusion,
2) Excess valvular tissue,
3) Valvular ring.
Type B Abnormal papillary muscle (N = 14)
1) Parachute mitral valve,
2) Hammock valve.
Eight patients died during hospital stay. Hospital mortality was correlated to mitral valve replacement and association to ventricular septal defect or patent ductus.
Follow up ranges from 1 to15 years (mean 7 years). One patient died. Two patients reqiered reoperation for residual mitral stenosis and/or insufficiency. 75% are in NYHA class I. Three patients were lost to the follow up. 10% thromboembolic events occured after valve replacement.
Surgical results for congenital mitral stenosis have improved. But high mortality remains, partly due to pulmonary artery hypertension and associated anomalies.

CONGENITAL MITRAL VALVE INSUFFICIENCY.
FUNCTIONAL CLASSIFICATION AND CONSERVATIVE SURGICAL MANAGEMENT

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Congenital mitral valve insufficiency is a difficult surgical problem because of lesions complexity.
Between 1970 and 1991, 108 patients were operated on for congenital mitral insufficiency in our institution. 93 patients underwent mitral valve repair using Carpentier's techniques.
Mean age at operation was 5.7 years (range 8 months to 12 years). Functional classification based on echocardiography and intraoperative data was as follows:
Type I : Normal leaflet motion (N=25).
1) anulus dilatation,
2) cleft leaflet,
3) leaflet defect
Type II : Prolapsed leaflet (N=36)
1) elongated chordae,
2) elongated papillary muscle,
3) absent chordae.
Type III : Restricted leaflet motion,
A) Normal papillary muscle (N=19).
1) commissural fusion,
2) short chordae
B) Abnormal papillary muscle (N=28).
1) parachute mitral valve,
2) hammock valve .
Hospital mortality was 12 patients. Follow up ranges from 1 to 20 years (mean 8.5), 7 patients died and 10 requiered reoperation. 14 patients were lost to the follow up. Thromboembolism is 2% at 8 years.
Conservative surgery has excellent results. Predictability of mitral valve repair can be assessed by echocardiography using the functional classification.

Session IX: Mitral Valve
Chairman: M. Querez-Jimenez

ECHOCARDIOGRAPHIC ANALYSIS OF MITRAL VALVE IN CHILDREN WITH THE COARCTATION OF THE AORTA

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The aim of this study was to analyze morphological and functional abnormalities of the mitral valve (MV) in children suffering from the coarctation of the aorta (CoAo). Study group consisted of 99 pts, 54 boys and 45 girls, aged 2 days to 14 yrs (mean 6,18 + 3,08 yrs). In all pts mean pressure gradient at the site of Co was 56,6+20,6 mmHg. The following additional abnormalities were found : LVOT obstruction in 78,8% pts, VSD in 13,23%, PAD in 6,1% and ASD in 6,1% pts.

Mitral valve abnormalities were recognized in 30 pts (30,3%). Dominant changes at papillary muscle (PM) level had 6 pts (single PM, asymmetric PM size or PM displacement). Thickness and shortening of chordae tendineae had 16/30 pts. Leaflet and MV annulus abnormalities (narrow MV orifice, accessory anterior MV leaflet) were recognized in 8/30 pts. One of these had double MV orifice. Supravalvular MV ring had 1 boy. Increasing diastolic flow velocity across the MV was found in all pts with MV abnormalities

(mean velocity 1,7 + 2,4m/s). Hemodinamically significant MV stenosis was diagnosed in only 2 children. Mitral regurgitation was recorded by Doppler echocardiography in 11 pts (semiquantitatively assessed as +1 in 7 pts, as +2 in 3 and as +3 in 1). Analyzing mean pressure gradient at Co site in groups with MV abnormalities (61,3+22,9 mmHg) and with normal MV (48,+19,8 mmHg) statistically significant difference (p< 0,001) was found.

Mitral valve abnormalities are often present in pts with CoAo (30,3%), and are in correlation with the severity of the disease. However, hemodinamically significant lesions of MV are rare (6,1%).

MITRAL ORIFICE DISEASE IN INFANTS AND CHILDREN

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During the years 1977-1991 we have studied 81 patients (pts) with disease of the mitral orifice: 78 congenital and 3 of rheumatic origine. The diagnosis was obtained on the basis of echo and/or angiocardiography. The mitral malformation was isolated en 29 pts and associated with other malformative complexes in 52. The most common isolated malformations were valve prolapse, hammock mitral valve, parachute mitral valve, arcade malformation of the mitral valve, congenital perforation and double mitral orifice. The malformative complexes to which mitral disease was most frequently associated were left ventricular and aortic tract pathology, ventricular septal defect, patent ductus + aortic coarctation, Tetralogy of Fallot and double outlet right ventricle. From the pathophysiological viewpoint, the lesions were considered as mainly stenotic (st,36 pts), mainly insufficient (insf, 27) and combined (comb, 18). The degrees of severity were slight (15+18+7), moderate (9+5+6) and severe (12+4+5); = (st+insf+comb). The diagnostic analysis referred to the annulus, leaflets, chordae tendinae and papillary muscles. A surgical treatment was applied to 22 pts; it was reconstructive in 7 and it involved the insertion of a prosthesis in 15. The survivors were 19 and they are asymptomatic after a period of follow-up ranging between 4 months and 14 years. Three of them are under medical treatment.

CONCLUSIONS: Repair shoud be performed whenever possible. Longtime prognosis remains unknown. Pts with slight - moderate degrees of severity should have surgery as late as possible. A reconstructive surgery may be intended in cases associated with malformative complexes when the are submitted to bypass surgery.

MITRAL VALVE REPLACEMENT IN 155 CHILDREN. LONG-TERM FOLLOW-UP WITH FOUR DIFFERENT PROSTHESES.

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We followed 155 children aged 6 to 16 years who underwent mitral valve replacement for rheumatic heart disease. Group I included 74 patients with Starr-Edwards prostheses and the mean follow-up period was 13 years. Group II was formed by 36 patients with Björk-Shiley prostheses and a mean follow-up of 7.5 years. Group III consisted of 31 children with dura mater bioprostheses and a mean follow-up of 5.1 years. Finally, group IV included 14 patients followed a mean period of 4.9 years with Hancock xenografts. The four groups of patients were comparable before surgery. The criteria of the New York Heart Association (NYHA) were used to evaluate all patients before and 12 months after surgery. All patients with mechanical prostheses received anticoagulant therapy with acenocumarol from the first postoperatory day. The incidence of complications such as thromboembolism, infective endocarditis, prosthetic dysfunction and death were determined at the end of the follow-up. The Fisher's exact test was used for nonparametric analysis and the two-tailed Student's T test for parametric results.

All but two patients improved their NYHA clinical status after surgery ($p<0.0001$). One hundred and ten patients were catetherized 1 to 7 years after the operation. The mean pulmonary arterial systolic pressure was reduced from 58 mmHg to 30 mmHg ($p<0.001$) and the mean pulmonary arterial wedge pressure decreased from 22 mmHg to 11 mmHg ($p<0.001$). There were no significant differences between the groups with mechanical and tissue prostheses in the incidence of thromboembolism and infective endocarditis. The incidence of both dysfunction and death were higher in the groups with bioprostheses than in patients with mechanical prostheses ($p<0.0001$ for both). There were no significant differences between Hancock and dura mater valves for all complications. Finally, we found a higher incidence ($p=0.037$) of thromboembolic episodes in patients with Starr-Edwards prostheses when compared to those with Björk-Shiley prostheses when analyzed in the mean follow-up of 5 years.

We conclude that in the pediatric age group, mechanical prostheses offer an excellent choice when conservative surgery of the mitral valve cannot be accomplished. In this group, Björk-Shiley prostheses have a lower incidence of thromboembolic episodes than Starr-Edwards prostheses.

MITRAL VALVE RECONSTRUCTION IN CHILDREN. LONG-TERM FOLLOW-UP.

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Between June 1985 and June 1991, 63 patients underwent mitral surgical valvuloplasty for rheumatic (46 cases), congenital (12 cases) and mixoid (5 cases) valve disease. Ages varied from 2 to 18 years with a mean of 14.1 ± 3.7 years. Mitral valve dysfunction was classified according to its pathophisiologic abnormalities. Group I was formed by 4 cases with mitral regurgitation secondary to lesions located in the valvular structures and with normal leaflet motion. Group II included 14 cases with mitral regurgitation with prolapsed leaflets due to lesions located mainly in the subvalvular structures. Group III was formed by 35 patients with mitral regurgitation with restricted leaflet motion due to lesions in the valvular and subvalvular structures. Finally group IV was formed by 10 cases with mitral stenosis due to lesion in the valvular and subvalvular structures. The surgical mortality rate was 4.7% (3/63) and the follow-up data are available in all the survivors from 1 to 96 months (mean 33.4 ± 25.4). Five patients underwent reoperation due to residual mitral incompetence (4 cases) and bacterial endocarditis (one case). One case died in the follow-up period during mitral valve replacement. Thromboembolism occurred in 4 cases in the absence of anticoagulant therapy; three of them were in atrial fibrilation. Before surgery, 28 cases were in functional class II of the New York Heart Association (NYHA), 34 patients were in class III and one patient in class IV. At the end of the follow-up period, 49 patients were in class I, seven in class II and four in NYHA funcional class IV ($p<0.0001$). Cardiothoracic ratio before surgery ranged from 0.40 to 0.81 (mean 0.60 ± 0.07), after surgery the values ranged from 0.40 to 0.79 (mean 0.55 ± 0.07) ($p<0.0001$). Randomized late echocardioghraphic evaluation in 24 cases revealed residual mild mitral regurgitation in 20 cases, moderate in 2 and severe in 2 more cases; the latter are waiting for mitral replacement. There were no significant differences in the surgical results between the four groups.

Mitral valve reconstruction provides stable functional results with low surgical and late mortality as well as an acceptable rate of reoperation despite the type of lesion in the mitral apparatus.

"MITRAL" VALVE CONFIGURATION AFTER REPAIR OF COMPLETE ATRIOVENTRICULAR SEPTAL DEFECT: EFFECT ON EARLY AND MEDIUM-TERM RESULTS

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Between 1982 and 1990, 82 patients with complete atrioventricular septal defect (AVSD) underwent surgery with a double-patch technique and

two types of left AV valve repair: complete closure of the "cleft" (57 pts, group 1) or partial/no closure , with preservation of a tricuspid configuration (25 pts, group 2). Mean age and weight at surgery, prevalence of Down's syndrome were similar; the AVSD anatomic type with undivided bridging leaflet was prevalent in group 2 (P = .05). In the "historical" part of the series (1982-1987), both repairs were employed. A tricuspid valve shape was elected along with Carpentier's principle in 20 cases; in 5, the cleft was left open or closed partially to prevent causing valve stenosis. In the modern era the cleft was routinely closed. The hospital mortality was higher in group 2 (52% vs 25%, P = .033); this may be due to different time of experience and a dissimilar use of a porous ASD patch, previously shown to be a risk factor. The mean follow-up interval extends to 79 months (group 2) and 35 months (group 1, P .001). Four reoperations were required (2 in each group), with a survival proportion, free from reintervention, of 76% (group 1) and 80% (group 2) at 6 years.

Functional class and need for cardiac medication of survivors did not differ. Trivial left AV valve incompetence was found in 64% of pts in group 1 and 50% in group 2 (P = NS); severe insufficiency was more common in group 2, but not significantly. Stenosis also prevailed in group 2, again not significantly. CONCLUSIONS: in this non-randomized study, the left AV valve function was slightly better when the "cleft" had been sutured, not up to statistical significance; a shorter follow-up period may be the cause of this difference. A definite conclusion can not be drawn on the superiority of one repair over the other and an individualized approach is indicated.

MITRAL VALVE RECONSTRUCTION IN TOTAL AV-CANAL

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Long-term results after operation for complete AV-canal mostly depend on atrioventricular valve function. Therefore we analysed our results of correction of total AV-canal with the two-patch technique in 42 patients operated between November 1988 and September 1991. Age at operation ranged between 3 months and 20 years (mean 26.7±6.3 months). At the time of operation 24 patients (57%) were 12 months old or younger. 32% of the patients were in NYHA class III or IV. All patients underwent VSD and ASD closure with two separate patches (autologous pericardium or dacron). Care was taken not to dissect papillary muscles or valvular cusps. After closure of VSD the newly created mitral cleft was repaired with a mean of 4 stitches. AV-valve function was then examined by saline injection to both ventricles.
Results: Follow-up ranged between 1 and 35 months. Operative mortality was 4.8%. Immediately after operation 76% showed no impairement of valvular function, 3 had mild and 4 moderate mitral valve insufficiency. One patient required mitral valve reconstruction 8 months after surgery. At the time of discharge from hospital all patinets were in NYHA class I or II. Two patients showed mild insufficiency of the tricuspid valve at the time of reexamination.
Mitral valve repair in total AV-canal with two patch technique without dissection of the cusps has excellent functional results and can be performed in the first yeare of life with low morbidity and mortality.

Posters not Orally Introduced

CARDIAC INVOLVEMENT IN TUBEROUS SCLEROSIS

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Twenty patients with tuberous sclerosis (TS) aged 1 week to 16 years (mean: 6,7 years) were investigated prospectively including clinical examination, standard-ECG, Holter-ECG and 2d-echocardiography in order to detect cardiac involvement.
Cardiac rhabdomyomas in the right (19) and left (13) ventricle as well as in the right atrium (1) were demonstrated in 13/20 patients. Two of them had tumor related obstruction of the left ventricular inflow and outflow tract, respectively.
Standard ECG (n = 19) was normal in 12 cases and showed right ventricular hypertrophy (1), premature atrial (1) and ventricular (1) contractions and repolarisation disturbances (3) in the remaining patients. Standard-ECG was normal in all patients with normal echocardiogram, in contrast 7/13 patients with rhabdomyomas showed an abnormal standard-ECG.
24-hours-Holter-ECG (n = 18) revealed frequent premature atrial (2) and ventricular (2) contractations, the latter in the presence of rhabdomyomas. No arrhythmias requiring medical therapy were observed.
The study encluded four neonates with extensive rhabdomyomatosis. One of them died after cardiac surgery. Tumor regression or complete involution could be demonstrated in the remaining three.
In conclusion, cardiac rhabdomyomas, although frequently present in our patients with TS, rarely caused hemodynamic obstruction; relevant arrhythmias were not observed. Therefore, surgical therapy is rarely indicated.

MORPHOMETRIC TECHNIQUES IN THE EVALUATION OF PULMONARY VASCULAR DISEASE

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The role of open lung biopsy as a means to predict the time course of pulmonary hypertension in patients with congenital heart disease and increased pulmonary vascular resistance is uncertain. The aim of the present retrospective study was 1.) to study the influence of technical aspects of morphometric analysis in the assessment of biopsy specimens with two different techniques (method of Cook & Yates versus method of Davies) 2.) to study the correlation of preoperative hemodynamic data to the degree of pathologic changes according to Heath & Edwards and Rabinovitch. 3.) to study whether the time course of pulmonary hypertension in the postoperative period evaluated with cardiac catheterization (20 of 61 patients; re-catheterization up to 4.5 years p.op.) could be predicted by the initial lung biopsy result.
8 controls (autopsy cases) and 61 patients (ASD II: n = 7; ASD I: n = 3; VSD: N = 22; complete ECD: n = 23 (with (n = 13) or without (n = 10) Down syndrome); persistent truncus arteriosus type I: n = 2; PAPVR: n = 2; PDA: n = 2) were included in this study (age 1 month to 75 years). 31 of the patients showed an increased Rp:Rs ratio (> 0.25). The pathologic changes according to Heath & Edwards in lung biopsy specimens obtained at the time of the corrective operation varied (Normal n = 23; Grade I: n = 22; Grade II: n = 12; Grade III: n = 3; Grade IV: n = 2).

Statistical analysis of the data showed that 1.) Grading of the biopsy was independent of the morphometric technique applied. However, vessel diameter was measured generally 1.64 times larger with the method of Cook & Cates than with the method of Davies. 2.) Preoperative hemodynamic data did not correlate to the degree of pulmonary vascular changes with neither the classification of Heath and Edwards nor the classification of Rabinovitch (Kruskall-Wallis test; $p>0.05$ %). 3.) With the limitations given by the small number of patients the time course of pulmonary hypertension cannot be predicted by the initial lung biopsy.

AORTIC VALVULOPLASTY FOR AORTIC REGURGITATION SECONDARY TO LEAFLET PROLAPSE

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At present there are only few encouraging reports describing the effectiveness of aortic valvuloplasty (AVP) in pts with significant aortic regurgitation (AR) caused by leaflet prolapse. We investigated a group of 7 pts, age range $6^{1}/12$ to $14^{10}/12$ yrs (average age $10^{1}/12$ yrs), with moderate to severe AR (average cardio-thoracic ratio (CTR) 0.59; average width of pulse pressure 58 mmHg). Four pts had a VSD (3 subpulmonic, 1 perimembranous). In 2 pts AR was associated with fibrous subaortic stenosis; 2 pts had had a correction of truncus arteriosus (TAC) and DORV, respectively, 6 and 12 yrs ago. The preoperative severity of AR was graded semiquantitatively by colour-flow Doppler examination and aortogram (5 moderate, 2 severe AR). The AVP for aortic leaflet prolapse (Trusler-plasty) was performed unicommissural in 5 pts, bicommissural in 2 pts (at the right- (6) and non-coronary cusp (1)). There was no operative death. Postoperative follow-up period ranged from 3 to 53 mo (mean 25 mo). Five children improved clinically (CTR 0.54, width of pulse pressure 38 mmHg) and showed only trivial or mild residual AR on colour-flow Doppler examination without any evidence of increase at the most recent examination. There were 2 valvuloplasty failures: 1 pt after TAC-correction still in NYHA I; 1 pt with subsequent homograft aortic valve replacement 19 mo later.
The AVP - carefully performed by cusp plication at the commissures - offers the possibility to conserve the aortic valve in pts with significant AR secondary to leaflet prolapse. With an encouraging rate of improvement of valve competence AVP can avoid or at least postpone valve replacement to adulthood.

HEART RATE AND RHYTHM IN PREMATURE INFANTS

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The aim of this study is to evaluate heart rate (HR) and rhythm in a group of preterm infants. Our study is prospectic. we report the data of the first 26 infants. Inclusion criterion are: gestational age < 34 weeks, absence of malformation and/or heart disease and/or arrhythmias. Arrhythmological evaluation is obteined with 2 Holter recordings: the first at 7 days of life, and the second at the age corresponding to the term of pregnancy. The mean gestational age of our 26 infants was 32±17 weeks(28-34), their mean wheight of 1733±450g.(920-2780). Mean, min., and max. HR were: 1°Holter 127±12, 79±13, 187±22 beats/min., 2°Holter 138±7, 85±9. 198±8b/min. HR Variability (an index of sympathetic tone) remained low in both Holter: 52±12 and 59±15ms. Supraventricular ectopies had an incidence of 58% in 1° and 27% in 2° recordings. Ventricular ectopies had an incidence of 19% in both Holter. Idioventricular rhythm was found in 8% and 4% respectively in 1° and 2°Holter. Junctional rhythm had an incidence of 34% and 15% in 1° and 2° controll. Max.RR interval was 1003±300 in 1°Holter and 843±257 in 2° Holter. PQ and QTc were normal in both recordings. We found different behaviour if neonatal wheight is more or less than 2kg. Under 2kg, HR varaibility was shorter (48±11 versus69±13ms), mean HR higher (133±13 v. 122±12b/m) and max. RR interval shorter (941±230 v. 1132±442ms) than in infants with wheight >2kg. In conclusion these data suggest that preterm infants have high sympathetic tone. more pronunced under 2 kg of wheight; and this high sympathetic tone influences more HR than incidence of arrhythmias.

THE BIDIRECTIONAL GLENN ANASTOMOSIS AS AN ADJUNCT TO REPAIR OF ATRIOVENTRICULAR SEPTAL DEFECT AND HYPOPLASTIC RIGHT VENTRICLE.

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Seven consecutive children (median age 5.5 years) underwent repair of atrioventricular septal defect (AVSD) with tetralogy of Fallot (6 patients) or ventricular septal defect with hypoplasia of the right ventricle (1 patient). Intracardiac repair consisted of patch closure of the atrial and ventricular septal defect, suture of the "cleft" between the anterior and posterior bridging leaflets (5 patients), relief of muscular right ventricular (RV) outflow tract obstruction, and placement of a valved homograft conduit between the RV and pulmonary artery (3 patients). In all cases, the diameter of the right AV valve measured intraoperatively was >2SD below the mean expected value, following intracardiac repair. Additionally a bidirectional Glenn procedure (superior vena cava anastomosed end to side with the right pulmonary artery) was therefore performed. There were no operative deaths. One patient required early reoperation for a residual VSD. The early postoperative period was complicated by pleural effusions in all (chylous effusion in 4), which required insertion of a drain. Serial follow-up cross sectional and pulsed Doppler (PWD) echocardiography was performed in all (median duration of follow-up 4 months). All 6 patients with an AVSD had evidence of mild left AV valve regurgitation; the left ventricular shortening fraction was within the normal range in all. There was markedly diminished RV compliance immediately postoperatively, manifested by a prominent atrial contribution to ventricular filling, and atrial augmentation of pulmonary flow on PWD, which disppeared within 4 weeks of operation. In the presence of a patent pulmonary valve, systolic flow reversal in the superior vena cava (SVC) on PWD was a prominent feature early post surgery. This became less marked in all, and eventually disappeared in 4 of the 6 patients. In 5 patients, a diastolic contribution to pulmonary flow through the SVC to both branch pulmonary arteries was seen. **CONCLUSIONS:** Early follow-up data suggest that the hemi-Fontan procedure is a useful adjunct in repair of hearts with AVSD and RV hypoplasia. Pleural effusions are common, and reflect the high venous pressure in the presence of a patent pulmonary valve. However, both RV and SVC compliance improved in all, with the SVC contributing to flow to both lungs in the majority of patients.

ATRIAL SEPTAL DEFECT: CLINICAL PRESENTATION AND SURGICAL OUTCOME IN THE PEDIATRIC VERSUS ADULT POPULATIONS

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Between January, 1953 and August, 1986 455 patients with atrial septal defect (ASD) were evaluated at the Cleveland Clinic. Population consisted of 158 pts. in the peds age group (age

range 0.10-20.9 yrs.; mean of 10.9 yrs.; 74 males, 84 females) and 287 pts. in the adult age group (age range 21.3-70.3 yrs.; mean 41.3 yrs; 73 males, 214 females). Follow-up was complete for 76.3% of the pts. (range 0.01-35.1 yrs; mean of 11.4 yrs.).

74.8% of peds pts. were NYHA F.C.I while most adults were F.C.II (60.6%)($p<0.001$). Atrial fibrillation was found at presentation only in the adults (12.2%)($p<0.001$). Characteristic murmur of ASD was present in 98.1% of peds. and 96.9% of adults. EKG demonstrated RVH in 76.9% of the peds., but only 51.2% of adults ($p<0.001$). Chest radiography demonstrated cardiomegaly in 59.6% of peds. and 70.6% of adults ($p<0.020$). Mean systolic pulmonary artery pressure (PAP) was 30.25 mmHg for the peds. and 40.24 for the adult ($p<0.001$). Of adults, 93% had no mitral regurgitation (MR), with only 3 adults having severe MR.

Primary closure was performed in 72.3% of peds. and 66.2% of the adults. 3 peds. (2%) and 5 adults (1.7%) died in the operative/peri-operative period. The most common peri-op complication was atrial fibrillation, which occurred in 5.2% of peds and 24.1% of adults. Follow-up demonstrated that 92.5% of peds and 85.4% of adults were NYHA F.C.I post-op. Atrial fib. developed in 6.0% peds and 8.1% of adults after discharge. 10-yr survival was 99.5% in peds and 99% in adults ($p=0.007$).

COR TRIATRIATUM SINISTRUM - EARLIER RECOGNITION THAN IN THE PAST!

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This demand we have to make according to the excellent prognosis after early surgical removal of bloodflow obstruction in cases of cor triatriatum sinistrum. Merely the clinical manifestation with relapsing pulmonary symptoms should be suspicious for pulmonary venous obstruction and initiate cardiological investigation.

This report summarizes 8 children, 3 to 5 years of age, with cor triatriatum sinistrum. Cardiological attendance of them has been so much delayed, because of misinterpretation of their clinical symptoms as to be pulmonary caused. In all patients we were able to achieve and to evaluate all decisive findings by non-invasive investigation (Echo, Doppler, MRT):
- **a membraneous, sometimes windsock-shaped structure with a distal opening, 3 to 8 mm in diameter, separating the proximal accessory atrial chamber from the true left atrium,**
- **congested lungveins and indirect signs of severe pulmonary hypertesion.**

Additional invasive procedures are only necessary in cases of contradictory findings or the impossibility of complete assessment.

Conclusion: Modern noninvasive imaging procedures should be used more frequently for early recognition of pulmonary venous obstruction, cor triatriatum and mitral stenosis in all infants and children with adequate clinical symptoms of lasting lung disease in order to prevent pulmonary damage and rightventricular failure by surgical correction.

LONG-TERM FOLLOW-UP OF 65 CHILDREN WITH CONGENITAL CARDIOVASCULAR ANOMALIES SUGGESTIVE OF ASPLENIA AND POLYSPLENIA SYNDROME

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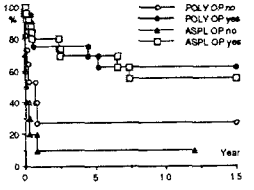
The natural history, as well as the effect of op were assessed in 65 pts (39♀, 26♂) with heterotaxy (1974 - Oc 91). 50 pts were recruited from the files of 218 pts (23%) with dextrocardia and/or single ventricle (SV). Cath/angio (0-4.5y, 22d) was compatible with asplenia in 34 pts (G1) and polysplenia in 31 pts (G2). The diagnosis was confirmed at autopsy in 18 and at op in 10 pts. Besides SV in 47%, other common lesions were: hypoplastic ventricle in biventricular hearts in 20%, common atrium - most frequently with common AV valve - in 64%, atretic or hypoplastic AV valve in 15%, anomalous return of systemic veins in 85%, including azygos continuation in 30 pts (G2), anomalous connection of pulmonary veins (PV) in 56%, PS in 44% and PA in 29% and noncardiac-nonsplenic anomalies in 34%.



For percentages of survival probability (Kaplan, Meier 1958) see diagram: Without op, only 10% (1/9) in G1 and 20% (2/11) in G2 survived > 1y. With palliation in infancy, survival was improved significantly (22/29♀) ($p< 0.001$). 7 of 8 pts survived Fontan op (FO) (4m-7.1y, median 2.5y) and were in NYHA class I-II to III-IV. 9 of the remaining 21 surviving pts (17 after palliation) appeared suitable for future FO.

Conclusions: In pts with asplenia and polysplenia the high risk for infant mortality necessitates extension of the indication for early op. Improved outcome after palliation and definitive repair by commonly used modification of FO may be accomplished by bilateral cavopulmonary anastomosis or total cavopulmonary connection. The specific type of cardiac and venous anomalies seems to be in favour for these newer techniques.

MANAGEMENT OF FIXED SUBAORTIC STENOSIS, A RETROSPECTIVE STUDY IN 57 CHILDREN.

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Though recommended by several authors, the benefit of early surgery in patients with Fixed Subaortic Stenosis (FSAS) has not been proven. We therefore reviewed both operated and non-operated patients with isolated FSAS (n=57), with special emphasis on the occurrence of Aortic Regurgitation (AR), during a mean follow up period of 6.7 years.

The course of FSAS after diagnosis was unpredictable. In a considerable number of patients (n=30) there was hardly any progression of subvalvular obstruction or left ventricular hypertrophy (LVH) and AR was of no hemodynamic significance.

In both, operated patients (n = 27) and non-operated patients (n=30) there was an increasing number of patients developing AR (resp. 37% at diagnosis to 81% postoperatively after 7.7 years and 23% to 30% after 4.4 years in the non-surgical group). Six patients (10%) developed moderate to severe AR. One patient died in follow up.

After surgery 15 patients (55%) showed a relapse redeveloping subvalvular pressure gradients of more than 30 mmHg and discrete subvalvular ridges in 11 cases (range 1 to 24 years after surgery, with a mean of 7 years).

In this population the follow up in FSAS and the number of relapses after resection or enucleation do not reveal any profit of early surgery. The unpredictable course and sometimes very severe progression makes frequent and careful follow up necessary.

INTIMAL THICKENING OF THE CORONARY ARTERIES IN RELATION TO FAMILY HISTORY OF CORONARY HEART DISEASE.

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Already in newborn children appear massive intimal thickenings composed mainly of smooth muscle cell proliferations. An important question is whether thickening of intima in children younger than one year of age reflects individuals suspectibility for later atherosclerosis.
The determination of luminal narrowing of the coronary arteries is based on measurement of the length of internal elastic lamina of the artery and the areas of arterial layers in cross-section. The degree of narrowing is the proportion of inner vascular layers to arterial lumen. The luminal narrowing in 141 infants varied between 0- 57 %. If a child had an infection at its death the degree of narrowing was significantly greater than in those without an infection ($p<0.01$). A total of 281 grandparents had died of whom 108 from coronary artery death (CAD). Positive family history for CAD was 3.1 times more usual in infants belonging to the highest tertile as determined by the degree of narrowing (18-58 %) as compared to those belonging to the lowest tertile (0-7 %). In male infants belonging to the highest narrowing tertile of the left coronary artery the CAD rate of the grandparents was 6.9 times higher than in those belonging to the lowest tertile. After adjusting for gender and age, positive family history and infection both had an increasing effect on the degree of narrowing. The correlation of the degree of intimal thickenings with positive family history of CAD suggests that intimal thickenings in infants are preatherosclerotic.

USE OF ASPIRIN IN SUBCLAVIAN TO PULMONARY ARTERY GORETEX SHUNTS.

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During the last decade, subclavian to pulmonary artery Goretex shunts (GS) have almost completely replaced the classic Blalock-Taussig operation for palliation of patients with decreased pulmonary blood flow. Partial or complete occlusion of GS remains however a major concern and a few authors have advocated the preventive use of antiplatelet agents such as acetylsalicylic acid (ASA). The influence of ASA on function and patency of 62 GS (61 patients) installed by the same surgical team, was assessed. Group I: 31 GS done between 1983-1986, no ASA; group II: 31 GS, 1987-1990 with ASA (mean dose 4.5 mg/kg/day). Functional status of GS was evaluated on follow-up angiocardiogram an average of 2.8 yrs after surgery. Patients of both groups were comparable for sex, weight, cardiac malformation and duration of follow-up. Patients of group I were older at surgery (mean: 581 vs 303 days, $p < 0.05$). The following indices were calculated: GS Patency Index (PAGI): GS minimal diameter at angio/GS diameter at surgery, and Pulmonary Artery Growth Index (PGAI): PA diameter at angio - PA diameter at surgery/PA diameter at surgery. There were 2 GS thrombosis in each group. Operative and angiographic (localized GS stenosis, PA kinking, PA stenosis) features were similar in both groups except for GS diameter installed at surgery (5.6 vs 5.2 mm, $p < 0.05$) probably due to the fact that the 2 smallest GS (4 mm) were in group II consisting of younger patients. GSPI was similar in both groups (68.5±24.9% vs 69.7±27.2%). PAGI was smaller in group I (57±43% vs 91±75%); this difference almost reaching significance. ASA does not appear to influence the rate of thrombosis, patency index or stenosis of PA or GS. However, it could affect the rate of growth of the pulmonary arteries and a prospective study is recommanded before drawing any final conclusion.

BALLOON DILATION OF CONGENITAL AORTIC VALVE STENOSIS: CAROTID ARTERY APPROACH IN THE FIRST MONTH OF LIFE.

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Critical aortic stenosis in neonates is a life threatening malformation. Operative mortality is high and restenosis or regurgitation are not uncommon. Balloon dilation is an alternative means of treatment but may be limited by the caliber of arteries in neonates. We performed balloon dilation of aortic valve in 7 consecutive unselected neonates, using a right carotid cutdown approach. All patients had congestive heart failure; their age ranged from 2 to 20 days and weight from to 2.1 to 4.0 Kg. Patients were evaluated before cardiac catheterization with Bidimensional and Color Doppler Echocardiography. In order to avoid femoral artery complications and difficulties in crossing the aortic valve, we used right carotid artery approach. Significant improvement in transvalvular pressure gradient was achieved with a mean percent reduction of 67.9% (70.4 mmHg before and 23.6 after dilation). None of the patients manifested complications related to the procedure and in all cases the procedure was safe, easy and successful. Two patients died because of associated lesions; all the surviving patients are followed (6-30 months) by means of Echocardiography.
In our opinion balloon dilation is a valid alternative to surgery in neonates. We recommend carotid artery approach for its technical advantages with good results and no vascular complications.

BALLOON AORTIC VALVOPLASTY FOR SEVERE SYMPTOMATIC AORTIC STENOSIS IN EARLY INFANCY. THE TREATMENT OF CHOICE ?

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Severe aortic stenosis (AS) in the neonatal period has a high mortality, even with prompt surgical treatment. During a 2.5 year period, 9 consecutive infants with severe AS have been treated by balloon aortic valvoplasty. All patients (pts) were symptomatic at < 6 weeks of age, 7 in the first week. Associated abnormalities included coarctation of the aorta in 5 pts (severe in 3, mild/moderate in 2), complete atrioventricular septal defect (AVSD) 1 pt, severe mitral regurgitation (MR) 1 pt and endocardial fibroelastosis (EFE) in 6 pts (severe in 3).

A percutaneous approach was used via the femoral artery and a valvoplasty balloon was used with a diameter equal to or 1mm less than the aortic annulus diameter measured echocardiographically. Co-existing severe coarctation was treated by balloon aortoplasty in 2pts and surgical repair in 1pt.

Symptomatic improvement with return of peripheral pulses and improvement of heart failure occured in 8(88%)pts. One pt with severe left ventricular hypoplasia died despite adequate valvoplasty shown at post mortem examination. Later death occured in 3pts with associated cardiac abnormalities (1-AVSD, 1-MR, 1-EFE + coarctation). An autopsy was performed on two patients and showed relief of commissural fusion in both. Complications of valvoplasty included guide wire perforation of the left ventricle in 1 pt and loss of femoral pulse in 4 pts. The presence of associated abnormalities and the degree of left ventricular hypoplasia were the main determinants of survival. Only 1 of 5 patients with a

left ventricular diastolic diameter (LVDD) of <2.0cm survives. The 4 patients with LVDD >2.0cm are alive & well.

The 5 survivors (56%) have been followed up for 2 - 24(mean 13) months. Four pts are asymptomatic and have mild/moderate residual AS, 1 with severe EFE is receiving diuretic therapy.

Balloon valvoplasty offers a promising alternative to surgical valvotomy in infants with severe aortic stenosis. Aortic coarctation frequently co-exists and may also be treated by balloon dilatation.

STENT IMPLANTATION IS SUPERIOR TO BALLOON DILATION FOR MAINTENANCE OF ARTERIAL DUCT PATENCY

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Prolonged patency of the arterial duct is of importance in children with duct dependent congenital heart disease. We have evaluated the relative efficacy of balloon dilation and stent implantation for this purpose in 26 newborn lambs (age 4.38 ± 1.7 days; weight 4.56 ± 1.2 kg). In 2 lambs it was not possible to recanalise the duct. In 7 only balloon dilation with balloon diameters of 4 - 10 mm was performed. 6 had stent implantation alone (3 with additional balloon dilation) using balloon-expandable or self-expanding stents of 4 - 8 mm in diameter. 11 underwent sequential balloon dilation and stent implantation with the same sized device (4 - 6 mm).

Within 5 minutes of balloon dilation the duct had either closed or constricted to a diameter of less than 1 mm in 4 and less than 2 mm in a further 2 lambs. The only inflation with a 10 mm balloon - after a poor result at 8 mm ruptured the duct producing tamponade.

Duct calibre is shown as mean +/- SD (number).

	4 mm	6 mm	8 mm
Balloon dilation	1.8 +/- 1.0 (5)	2.2 +/- 1.3 (9)	3.8 +/- 0.7 (5)
Stent implantation	3.6 +/- 0.7 (4)	5.2 +/- 1.0 (9)	7.1 +/- 1.3 (3)

Comparison of those undergoing sequential study showed:

	Pulmonary/ Systemic ratio	QPQS	Duct size (mm)
Baseline	0.31 +/- 0.1	1.1 +/- 0.4	0
Balloon dilation	0.41 +/- 0.1	2.2 +/- 1.0	2.1 +/- 1.3
Stent implantation	0.57 +/- 0.2	3.5 +/- 2.1	4.7 +/- 1.0

In 6 who have entered into a chronic study with follow up between 2 - 8 months (current weight 24 - 48 kg), patency by stenting (3 Medinvent - Wallstent and 3 Numed - Towers stent) has been well maintained.

We conclude that implantation of 4 - 6 mm stents produces superior haemodynamic and angiographic results to balloon dilation.

COMPARISON OF FOUR DIFFERENT TYPES OF STENT FOR MAINTAINING ARTERIAL DUCT PATENCY

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Stent implantation into the arterial duct is potentially of therapeutic value in some forms of duct dependent congenital heart disease. In atherosclerotic disease, stents are usually considerably longer than the lesion itself, so that perfect alignment of the centre of the lesion with the centre of the stent is not critical. Ostial lesions require more accurate placement to avoid protrusion into the main vessel. Stenting of the arterial duct, however, requires even more precision in order to encompass the whole vessel and its two ostia.

We have compared the ease of implantation and effectivness of 4 different types of stents [Tower (Numed), Wallstent (Medinvent), Palmaz-Schatz (Johnson & Johnson) and Gianturco-Roubin (Cook)] of various diameters and lengths in 19 newborn lambs (age 2 - 9 days, weight 2.4 - 7.3 kg, duct length 1.3 - 1.7 cm).

Stent	Tower	Wallstent	Palmaz Schatz	Gianturco Roubin
number	17	7	5	4
Diameter (mm)	4 - 6	4 - 8	4 - 6	4
Length (cm)	0.7 - 1.3	1.4 - 2.1	1.3 - 1.4	2
Radio-opacity	+ + + +	+ +	+/-	+ +
Retrievability	+ + +	-	-	+ + +
Expansion	balloon	self expansion	balloon	balloon
Mounted	yes	yes	no	yes
Deployment				
Ease	+ + +	+ +	+	+
Effectiveness	+ + +	+ +	+	-

Continuous strand stents had the advantage of being retrievable with a simple snare if misplaced. Stents shorter than the duct itself were effective when additional stents could be placed adjacent to them, but longer stents protruded into the aorta and/or pulmonary artery. Stents exhibiting significant shortening during expansion were difficult to position as were stents with poor radio-opacity.

For accurate stent placement into the arterial duct a well defined length and good radio-opacity are essential.

PERCUTANEOUS DUCTAL STENTING FOR PULMONARY ATRESIA: EARLY EXPERIENCE.

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Ductal patency was maintained by implantation of balloon expandable steel stents (Johnson & Johnson) in two neonates with pulmonary atresia. Both had a right sided aortic arch and left sided duct. One had hypoplasia of the right heart and a failed shunt operation and the other had a complete atrioventricular septal defect. The stents were mounted upon 3.5 or 4mm balloon diameter coronary angioplasty catheters and were delivered via the axillary or femoral arteries under general anaesthetic. Repeated procedures were necessary before successful stenting of the full length of the ducts; two stents (7mm and 15mm length) were required in one case and four (one 15mm and three overlapping 7mm stents) in the other. Complications included guide wire perforation of a pulmonary artery (treated conservatively) and cardiac tamponade (treated by pericardiocentesis and autotransfusion). After ductal stenting pulse oximetry showed both babies to be greater than 80% saturated without prostaglandin treatment and one required diuretic therapy. The first child died suddenly after 5 weeks of uncertain cause and the second died of pneumococcal septicaemia after 9 days. At autopsy both ducts were patent and endothelialised. Post mortem angiography confirmed ductal patency with flow to both branch pulmonary arteries in both babies. Percutaneous stenting of the arterial duct in pulmonary atresia is technically feasible and provides balanced perfusion of both lungs. More experience is necessary, but our results suggest that stenting of the duct is a promising alternative to aortopulmonary shunt operation in neonates with pulmonary atresia.

(* Introduced by D F Dickinson)

CATHETER OCCLUSION OF PATENT ARTERIAL DUCT - ECHOCARDIOGRAPHIC FOLLOW-UP

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Arterial duct occlusion using the Rashkind technique has been employed in 68 patients over the past 2½ years.

The patient ages ranged from 7 months to 29 years and 53 were female. Fifty-four patients had a small device implanted and the remaining 14 a large one. Five devices embolised to the pulmonary arteries either during or immediately after implantation, two of whom were referred for surgical retrieval of the device and ductus ligation. The other three were retrieved via catheter, two of whom had a device successfully implanted at a later date, but one opted for surgery.

Only 2 patients have clinically evident shunts with a continuous murmur. Doppler studies, however, showed residual shunts, usually very small, in 16 patients, although only 6 of these have had the device implanted more than 6 months. There is turbulence on colour flow mapping in the left pulmonary artery in 2 patients but no high velocity jet is detected. There was no obstruction to the descending aorta on Doppler.

Many residual shunts seen on Doppler early after implantation usually disappear. The absence of left pulmonary artery stenosis or aortic obstruction probably relates to the deliberate policy of using small devices where practically possible.

STATUS OF INNERVATION IN PEDIATRIC HEART RECIPIENTS

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To determine the status of autonomic innervation after pediatric heart transplantation (HTx) sinus node recovery time (SNRT), post-pacing cycles lengths and spontaneous heart rate fluctuations were examined in seven long term surviving pediatric patients. Mean age at the time of HTx was 2,6 years (range 3 weeks - 15,4 years), the follow-up time was 11 - 42 months. Heart rate variability (HRV) was assessed by spectral analysis based on a fast Fourier transform using a commercial Holter-ECG analysis program . In all except one patients corrected SNRT continuously increased at progressively rapid atrial pacing rates as previously described in denervated hearts. One patient showed a continuous decrease of corrected SNRT at higher stimulation rates without any signs of impaired sinoatrial conduction or incomplete sinus node suppression. Significant reduction of the post-pacing cycles was only observed in this patient; pre-pacing cycle length was reached within the first ten post-pacing cycles. Spectral analysis of HRV in this patient showed discrete peaks in the low-frequency=LF (0,04-0,15 Hz) and the high-frequency=HF (0,25-0,4 Hz) area, as typically seen in innervated hearts. Follow up of 3 other patients showed a discrete peak at a frequency of 0,22-0,24 Hz appearing 6-12 months after HTx. All these patients were younger than four years at the time of HTx; there was no evidence of acute graft rejection at the time of HRV analysis.
Electrophysiologic substrate and clinical relevance of modified HRV have to be determined in long-term surviving pediatric patients after HTx.

HEART TRANSPLANTATION IN THE YOUNG. IMMEDIATE RESULTS AND FOLLOW-UP.

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From december 1984 to july 1991, 23 young patients had 24 orthotopic heart transplantations (mean age 10,7 years, ranged 2 days to 21 years); three patients were younger than 2 years, 9 were 2 to 14 years old and 11 were over 14 years of age.
Pre-operative diagnosis were dilated or restrictive cardiomyopathy (n=10), congenital heart diseases with congestive heart failure after palliative or corrective surgery (n=10), hypoplastic left heart syndrome (1), symptomatic fibroma of the interventricular septum (1), and Uhl disease with severe and refractory ventricular arythmia (1); one teenager had retransplantation for acute rejection.
Four patients required external ventricular assistance during 1 to 15 days awaiting emergency transplantation: three of them died in the immediate postoperative period , and the last one died one month after transplantation with toxoplasmose and CMV infection.One adolescent died during the first month with necrotic pancreatitis and acute rejection, and another was lost immediately post operatively for technical anatomic complication. Mortality was 100% in the group of emergency indication compared with 10% for the others.Total mortality was 26 % (6 patients). Immunosuppression was achieved with combination of prednisolone (0,2 mg/kg/day), azathioprine (1 mg/kg/day) and ciclosporin. There were 2,6 episodes of acute rejection per patient (0 to 4) during the first year, and 82% of them occured in the first 3 postoperative months. Intravenous corticotherapy was given for acute rejection : only 1 patient needed antithymocyte globulin, and another had retransplantation at 34 months. Renal function was moderatly impaired in all patients but one. Linear growth improved after transplantation in the 7 patients who received transplants before age 14 years, but remains moderatly impaired for 5 infants. Follow up study (3 months to 6,8 years, mean = 25 months) demonstrates that 17 patients (74%) are living at home with a good quality of life. Psychomotor development, exercice tolerance and school results remain within normal range.

ENOXIMONE FOR THE TREATMENT OF REFRACTORY LOW-CARDIAC-OUTPUT.

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The effectivness of enoximone (E) was evaluated in pts with low-cardiac-output refractory to conventional treatment.

Newborns (N=16): Bolus 1 mg/kg followed by an infusion of 10 mcg/kg/min. E resulted in 12/16 pts ("responder") in a dramatic hemodynamic improvement, 9 of these survived. All non-responders died subsequently. E resulted in reduced filling pressures (p<.005) and improved cardiac index (before E: 0.96 l/min/sqm; after E: 3.05 l/min/sqm; p<.001).

Pharmacological bridging for transplant: In 96 pts with end-stage dilative cardiomyopathy jugded to be candidates for an assist device E was given. In 60% prolonged stabilization occured ("responder"), in 40% no or transient hemodynamic improvement was achieved. 32 of the responders were successfully transplanted. 7 of the non-responders died subsequentelly, in 30 a biventricular assist device had to be implanted.

Conclusion: E is an effective pharmacological approach for the management of end-stage low-cardiac-output.

ABOUT THE ORIGIN OF THE VIBRATORY INNOCENT HEART MURMUR.
Incidence, ECG and Doppler-echocardiography

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The vibratory innocent heart murmur (VIHM) is the most common one of the innocent heart murmurs in childhood. Genesis and exact site of origin of the VIHM is not known yet.

Methods: In co-operation with school medical officers, up to now, 691 children underwent heart auscultation by one pediatric cardiologist. Each VIHM-child is matched on lenght, bodyweight, sexe and the absence of any heart murmur. VIHM-children (n=62), non-caucasian race excluded) and paired controls (n=62) were examined by ECG, phonocardiography and Doppler-echocardiography.

Results: The incidence of the VIHM related to age is: 5-6 yrs 16.9%, 7-10 yrs 10.5%, 11-14 yrs 10.7%. ECG-axis, ECG precordial left ventricle (LV) voltages and carotid pulse waves are not significant different in the match-control study. LV-bands were seen in more than 40%, both in VIHM-children and controls.

The diameter of LV outflow tract and aorta is significant smaller in VIHM-children ($p<0.001$). Also maximal velocity and maximal acceleration in the ascending aorta are significantly increased in VIHM-children ($p<0.001$). Systolic vibrations on the aortic valve leaflets are seen in 89%(VIHM) and 70%(controls). In the paired study frequency and amplitude are significantly higher in the VIHM-children ($p<0.001$). Systolic vibrations on LV-bands or mitral chordea are not present.

Characteristic mean values:

	VIHM-children	case-controls
LV-outflow tract	15.7 mm	16.3 mm
Aorta	19.1 mm	20.2 mm
max. velocity Aorta	126 cm/s	108 cm/s
max. acceleration Ao	38 m/s^2	27 m/s^2
Vibration Ao-valve	96 Herz	76 Herz
	1.3 mm amplitude	1.1 mm amplitude

Conclusion: The VIHM is associated with higher aortic flow velocity and acceleration, probably due to a smaller diameter of the LV outflow tract and the aorta, or higher contractility of the myocardium. Vibrations of the aortic valve leaflets, with higher frequency and amplitude in VIHM-children, suggest that the site of origin of the VIHM is around the aortic valve. LV-bands are not related to a VIHM.

CARDIOTOXICITY OF BONE MARROW TRANSPLANTATION FOR ACUTE LEUKAEMIA; EFFECT OF PRETRANSPLANT TREATMENT, CONDITIONING REGIME OR BOTH?

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Bone marrow transplantation (BMT) can cause significant cardiovascular compromise. The relative contributions to this of prior treatment for malignant disease and of the ablative conditioning therapy needed for BMT are unclear. To elucidate this 52 echocardiographic (echo) examinations were performed serially on 20 patients (aged 1.9-19years) as they underwent BMT. Twenty age matched (1.8-18 years) controls were also studied.

The 14 patients with acute lymphoblastic leukaemia (ALL) or relapsed ALL had received 190-470(median 380)mg/m² of anthracycline (Daunorubicin +/- Epirubicin) prior to BMT conditioning. The 6 patients with acute myeloid leukaemia (AML) had received 3-500(median 300)mg/m² of Daunorubicin, plus 50mg/m² of Mitoxantrone. All patients received conditioning with high dose cyclophosphamide and total body irradiation.

Before conditioning, left ventricular ejection fraction (LVEF,median(IQR)) was lower in both ALL (70(67-74)%) & AML(70(58-74)%) patients than controls (75(72-78)%) ($p = <0.03$ Mann Whitney U Test). Peak LV early (E) to atrial (A) phase filling velocity ratio (EA ratio, measured by pulsed Doppler echo) was significantly lower in AML patients (1.5(1.3 -1.6) than controls (1.8(1.6-2.12) ($p=0.03$), but not in the ALL patients. This was largely due to peak E velocity differences; ALL (0.78(0.78-0.85)m/sec) and AML (0.63(0.55-0.71)m/sec) ($p=0.02$).

Scans were repeated at engraftment and at 3 months post BMT. One child with AML died from cardiac decompensation before engraftment took place. By engraftment, group LVEF had fallen only in the AML group, to 57(49-60)% ($p = 0.08$). EA ratio fell for both ALL and AML groups (to 1.3(1.1-1.5) & 1.02(0.8-1.2) respectively ($p = <0.03$). This is largely due to a rise in A. At 3 months post BMT, all study parameters had returned to pretransplant values for the AML group, but remained abnormal compared to controls. In the ALL group LVEF had fallen to 66(51-68)% ($p=0.09$) & EA remained lower ($p=0.04$) than pre-BMT, (1.3(1.2-1.5), with A higher than in the AML group.

Both pretransplant treatment & the BMT conditioning regime have profound effects on LV function in children. Previous treatment with Mitoxantrone is particularly associated with acute deterioration of ventricular function following conditioning. If this is survived, some improvement in ventricular function may occur. However, significant abnormalities of both systolic and diastolic function remain detectable, related both to the cumulative effect of cardiotoxic drugs and to myocardial irradiation.

ECHOCARDIOGRAPHIC DIAGNOSIS OF DOUBLE-CHAMBERED RIGHT VENTRICLE.

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In double-chambered right ventricle (DCRV) there is an obstruction in the mid-portion of the right ventricle due to aberrant muscle. Echocardiographic examination of this region is difficult, because of its short distance to the thoracic wall. Two-D echo and conventional Doppler may miss the anomaly. Colour Doppler imaging (CDI) allows improved non-invasive diagnosis of DCRV.

From January 1985 till October 1991 7 children were observed (age between 4 months and 12 years) with DCRV. The anomaly was not detected before surgery in the first two patients. Preoperative diagnoses were TOF and DORV with pulmonary stenosis, resp. After CDI had become available the following 5 cases were diagnosed correctly.

Associated lesions were subaortic VSD in 4 cases and subpulmonary VSD in one. Two patients had R-L shunts, one had a bidirectional, and two showed L-R shunting. So far the diagnosis has been confirmed in 4 cases by cardiac catheterization and surgery.

Conclusions:

1. Anomalous flow pattern (turbulence, increase of velocity) in the mid-portion of the RV suggests DCRV. It is detected best with CDI.
2. CDI guided pw Doppler (HPRF) allows estimation of the obstruction despite nonoptimal angle.
3. Careful 2 D-examination (5 or 7,5 MHz) can visualize details of the obstructed region.

RIGHT VENTRICULAR RELAXATION AND DIASTOLIC VENTRICULAR FILLING IN NEONATES: AN ECHO DOPPLER STUDY

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Our purpose was to serially study right ventricular filling using echo and Doppler in full term neonates after birth. Echo and pulsed Doppler studies were performed for 21 newborns during the first 36 hours of life and subsequent serial studies were performed in the second (n = 21) and third (n = 14) weeks of life. Evaluation included measurement of right ventricular isovolumic relaxation time (RIVRT), and peak E and A velocities of tricuspid valve. Pulmonary velocities were studied for evidence of patent ductus and to determine pulmonary artery acceleration time (PAT). Mean RIVRT decreased significantly from 68±9ms to 39±14ms ($p < 0.001$)

between the first and second week. Mean tricuspid peak E increased significantly between the second (48±7cm/s) and the third week of life (62±8cms; $p < 0.001$). Mean peak A wave velocity did not change significantly during the time of study. PAT increased from the initial measurement (68±10ms) to week 2 (85±9ms; $p < 0.001$). The changes in RIVRT were significantly related to the changes in PAT ($R^2 = 0.99$, $p = 0.03$), suggesting a significant influence of afterload on diastolic filling. These data demonstrate the normal progression of right ventricular filling indices in neonates.

INTRAOPERATIVE AND POSTOPERATIVE TRANSESOPHAGEAL ECHOCARDIOGRAPHY (TEE) IN CHILDREN.

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102 examinations were performed (ages 9 mo - 16 y, weight 6-50 kg): 77 of these were done intraoperatively, 20 in the cardiac catheter lab, and 5 postoperatively. 15 patients were followed up at 1 h intervals over 4-23 h. All 20 atrial septal defects (ASD) were closed completely.In 8/20 closures of ventricular septal defects (VSD), colour coded Doppler showed residual VSD, 2 of wich were directly visible in 2-d-echo. Residual VSD was also found after 2/11 operations for tetralogy of Fallot (TOF); the defect was pronounced in 1 pat. who needs re-surgery. 11 pat. with closure of ASD showed a marked reduction of the right atrial and the right ventricular area. We observed a significant reduction in surface area of both ventricles in 11 pat. after VSD surgery, but no significant change in surface area of any of the four cardiac cavities after surgery for TOF.
These changes in area (measured intraoperatively) were confirmed by subsequent TEE examinations in 15 pat. TEE also served to monitor ventricular function in these pat.
Conclusions: Perioperative TEE
(1) is the only imaging procedure to evaluate the result of surgical intervention intraoperatively.
(2) serves to determine quantitatively changes in surface area and volume of cardiac sections after surgical correction.
(3) serves to evaluate ventricular function peri- and postoperatively.

ECHOCARDIOGRAPHIC ASPECTS OF TETRALOGY OF FALLOT IN THE FIRST WEEK OF LIFE.

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With the purpose of establishing the validity of currently accepted echocardiographic criteria in patients with tetralogy of Fallot (TOF) below one week of age, 37 newborns aged 0-7 days with a large unrestrictive perimembranous ventricular septal defect (VSD) were studied with echo-doppler. Twenty-three of these were later confirmed as having TOF, 11 VSD only and 3 double chambered right ventricle (DCRV). VSD and TOF groups were compared for the following criteria: deviation of the infundibular septum, right ventricle to pulmonary artery pressure gradient (RV/PA grad.), aortic overriding, pulmonary artery (PA) and aortic (AO) anulus diameter. Anterior deviation of the infundibular septum was difficult to assess, especially in mild forms of TOF. There was no significant difference in the size of the VSD; degree of aortic overriding was different: 12±18% vs 45±9% ($p<0.001$); but with overlapping of results between the two groups. Doppler PA velocities were different: 1.23±0.40m/sec vs 2.63±0.73m/sec ($p<0.001$), but 3 TOF had velocities within the range of patients with VSD alone. Most significant differences were found in the diameter of the PA (10.3±1.6mm vs 5.4±1.2mm), AO (8.2±1.2mm vs 10.4±1.3mm) and PA/AO ratio 1.26±0.20 vs 0.52±0.11 ($p<0.001$) with no overlapping between the two groups for this last measurement. In patients with DCRV the PA/AO ratio was not significantly different from that of patients with VSD alone. On follow-up all patients with TOF developed a significant RV/PA gradient by the end of the first month of life. Most generally accepted criteria for diagnosis of TOF in the first week of life may be unreliable and subject to interpretation by the physician. PA/AO anulus ratio appears to be a reliable criterion, easy to obtain and less dependent on observer's interpretation.

SECUNDUM ATRIAL SEPTAL DEFECTS: CORRELATION BETWEEN ECHOCARDIOGRAPHIC AND SURGICAL FINDINGS.

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A prospective study of 28 children undergoing surgical closure of a secundum atrial septal defect (ASD) was undertaken to verify the value of echocardiography in providing anatomical definition of the defect by comparing ultrasonic data to findings at surgery. This could also help to select candidates for closure with an endovascular device. Mean age was 6 years (1-14 years). Echo were reviewed by 2 independent observers blinded to the surgical description. On each echo, in the subxiphoid view, were measured the size of the defect, length of the septum and the diameter of the transatrial color doppler jet. The right ventricular end-diastolic diameter was measured in the parasternal view on M-mode tracing. A defect was classified as superior or inferior when its center was located more than 10% above or below the center of the septum, otherwise it was labelled central. At surgery, major and minor axes of the ASD were measured, the defect was localized in the septum and the presence or absence of a rim wide enough for prosthetic implantation (at least 5 mm) was noted. Echo correctly diagnosed position in all superior 8/8, all inferior 6/6 and in 10/14 central ASD. At surgery a rim was never present in inferior ASD and present in all cases but 1 in superior and 1 in central ASD. There was no correlation however, with echo findings. At surgery, the defect was always oval in shape short axis averaging 65% (46 to 87%) of long axis, asymetry was never extreme. Major axis correlated better ($r=0.87$, $p<0.001$) than minor axis ($r=0.70$, $p<0.005$) with echo diameter. Width of color doppler jet overestimated diameter measured at echo and at surgery respectively by 13.4% and 11.3%, p=ns. Right ventricular end-diastolic diameter correlated well with size of the defect at surgery ($r=0.82$, $p<0.001$) or at echo ($r=0.72$, $p<0.001$). Echocardiography allows excellent anatomic description of secundum atrial septal defects, it can only predict indirectly (by locating the defect) the presence or absence of a rim wide enough for prosthetic implantation.

VALIDATION OF RIGHT HEART PRESSURE ESTIMATION BY DOPPLER IN INFANTS WITH TRICUSPID REGURGITATION

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The Doppler method of measuring maximal tricuspid regurgitant (TR) jet velocity and applying the modified Bernoulli equation to determine systolic pulmonary

arterial pressure has, surprisingly, not been validated for use in neonates and infants. Simultaneous Doppler-catheterisation studies were performed on 28 occasions in 26 infants with congenital heart disease (CHD) less than one year of age. Maximal TR jet velocity was recorded using continuous wave Doppler. Results were compared with systolic RV-mean RA pressure difference. Bland-Altman statistical analysis was used.

Results The 28 values were obtained from over 60 examinations; many cases did not have TR, even with RV pressure at systemic levels. Doppler values slightly underestimated the true pressure drop (mean -2 mmHg). Limits of agreement (95% confidence intervals) were -11.8 mmHg to +7.8 mmHg. Limits of agreement for velocity were -0.41m/s to +0.26m/s.

Conclusion: Peak TR velocity and the Bernoulli equation give a valuable and reasonably accurate assessment of systolic right ventricular pressure in infants with CHD less than 1 year of age.

MITRAL VALVE PROLAPS,OUR EXPERIENCES IN THE CLINICAL AND ECHOCARDIOGRAPHIC DIAGNOSIS

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The 9 year(1982-1991)retrospective analysis of 6838 first clinical and echocardiographic checkups showed that isolated MVP was diagnosed in 257(3,7%)children(2-18 years). The diagnosis is based on standard clinical and echocardiographic checkups by M-mode and 2D-techniques.Prevalence in girls is obvious(200-77,8%)compared to boys(57)and usually present in lo-13 year old (142).

In a large number of children cardiac murmur was revealeded accidently.Some heart troubles were revealed in 51 children (19,8%).Isolated diopatic MVP was diagnosed in 200(81,7%) children;(16/6,9%) had signs of Sy Marfan, 1(0,7%) Sy Down. 30 children had clinical basis for Rheumatic ethiology in MVP(11,6%).

Objective checkup revealed typical heart murmur of MVP in 183(71,2%), nontypical in 60(23,3%)and stronger murmur of mitrial insuficienci in 14(5,4%).A kind of a heart arrythmia was diagnosed in 16 children(6,2%).Clear body Asthenis was diagnosed in 70children (27,2), in 19 children this was very high. 19 children(7,4%) had isolated theracic skeletal abnormality. ECG was usually normal but vertical AQRS was often present (100 children-40%).13 children had repolarization disturbances(5%).X-ray chest checkup showed normal heart in 180 children(70%),small juvenile heart in 52(20,2%) flat spinal column in 21(8,2%).

Echocardiographic clear presenation of a cholosystolic MVP was diagnosed in 129(50,2%)children and telesistelic in 126 (49%)children.Two cases with typical murmur had no echo presentation of MVP.The clinical and echo follow-up of our patients showed that 19(7,4%)children had hemodynamic significant mitral regurgitation and valpuloplastic was performed in one.

Conclusion:Analysis made showed the characterstics of MVP.The use of Echo facillitated the diagnosis and follow-up of children with this anomaly.

SMALL MITRAL VALVE DIMENSIONS IN CHILDREN WITH COARCTATION OF AORTA.

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Children with coarctation of the aorta and morphologically normal mitral valves were studied to establish whether mitral valve dimensions are smaller than the normal range and whether this accounts for the apical diastolic murmur (MDM) heard in some cases. 41 children with coarctation (cases) were compared with 41 normal children (controls) matched for BSA.

Two dimensional/Doppler echocardiography was employed to measure mitral valve diameters in the parasternal long axis (MVD.LA), parasternal short axis (MVD.SA) and apical four chamber (MVD.4C) views; mitral valve area was measured in the parasternal short axis view (MVA.SA) and by Doppler flow velocity using the pressure half time method (MVA.DOP). The results in 14 children with coarctation plus MDMs were compared with 14 matched cases without murmurs.

Mitral valve dimensions were significantly smaller in cases than in controls in respect of MVD.LA (1.7 vs 1.9 cms; p <0.0003), MVD.SA (2.19 vs 2.28 cms; p <0.017), and MVA.SA (2.32 vs 3.15 cm2; p <0.008). The differences were just short of significant levels in respect of MVD.4C (1.99 vs 2.18 cms; p = 0.07), and not significant for MVA.DOP (6.23 vs 7.03 cm2; p = 0.38). There were no significant differences in mitral valve dimensions between cases with or those without MDMs.

Coarctation of the aorta is associated with relative hypoplasia of the left heart as reflected by reduced dimensions of the otherwise normal mitral valve.

EARLY AND MEDIUM-TERM RESULTS OF PERCUTANEOUS BALLOON MITRAL COMMISSUROTOMY. REPORT OF 210 PROCEDURES.

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Percutaneous mitral commissurotomy (PMC) has recently been developed as alternative for surgery in rheumatic mitral stenosis (MS). In 210 PMC were performed in a 3 year period. We intended to determine early and medium-term results and identify factors influencing the results. Ages ranged from 9 to 66 years (mean 29 years, 24 patients between 9 and 16 years of age).

The mitral valve area (MVA), measured by Gorlin's formula, increased from 1 $\pm$ 0.3 to 2.11 $\pm$ 0,5 cm^2 (P 0.001). Suboptimal results as defined by a post procedure MVA less than 1.5 cm^2, an increase in MVA less than 25% and/or creation of 2/4 or more grade mitral regurgitation, occurred in 21 of 205 patients. Multiple regression analysis of clinical, echographic and hemodynamic variables showed that the result of PMC was predicted by the anatomic status of the mitral apparatus especially subvalvular, as defined by echocardiography. Thickened, retracted and/or fused chordae tendineae were predictive for suboptimal results. Optimal results were seen in pliable mitral leaflets and none or only moderately diseased subvalvular mitral apparatus.

The main complications comprised: 3 Haemopericardium, 4 severe mitral regurgitation. No death occurred. In 5 cases PMC failed due to technical problems. The follow up ranged from 6 to 41 months (mean 17 months) and showed maintenance of the early results as a rule. Mitral "restenosis" as defined

by loss of more than 50% of the mitral increase in MVA and a MVA estimated less than 1,5 cm^2, occurred in 5 of 184 optimal results (2.7 %). We conclude that PMC is an excellent alternative for surgical repair, effective and safe. The results are predicted essentially by the anatomy of the mitral apparatus. The medium-term results are encouraging but close follow-up remains necessary to determine long term results.

LATE RESULTS OF COMMISSUROTOMY FOR RHEUMATIC MITRAL STENOSIS IN CHILDREN AND YOUNG ADULTS.

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Balloon dilation of rheumatic mitral stenosis has emerged as a new therapeutic procedure during the last few years and has been reported to give satisfactory immediate results. As reference for future follow up of balloon dilated patients and also to assess factors influencing the long term results we have analysed the outcome of surgical valvotomy of rheumatic mitral stenosis performed in children and young adults.
During a 7 year period ending Jan-85, 38 patients, 14 boys and 24 girls, age range 9-20 years (median 17) with rheumatic mitral stenosis as dominating hemodynamic feature undervent mitral commissurotomy. There was no operative mortality. The mean follow up was 5.0 years (range 0.1-9.4) and apart from three patients lost for follow up 0.1-1.2 years post operatively, all patients were followed regularly and were given penicillin prophylaxis monthly. Actuarial freedom from the need of reoperation was at 5 years after surgery 83% for the entire group. The long term results were unaffected by factors as age, preoperative pulmonary hypertension or preoperative functional state. The only factor that seems to influence the need for reoperation was the presence of mitral insufficiency in association with mitral stenosis. The actuarial freedom from reoperation 5 years after surgery was 95% in patients without and 76% in patients with associated mitral regurgitation. The corresponding values 8 years after surgery were 95% vs 66%.

In conclusion we find that mitral comissurotomy for isolated mitral stenosis has excellent long term effect. Patients having associated mitral regurgitation are likely to need reoperation early.

PERITONEAL DIALYSIS FOLLOWING OPEN HEART SURGERY IN CHILDREN

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Renal insufficiency requiring dialysis is a serious complication following open heart surgery with poor prognosis. Children developing oliguria, fluid overlaod, hyperkaliaemia or increasing uraemia were defined as having acute renal failiure and were dialysed. This study reviews our experience with 51 children requiring peritoneal dialysis following open. heart surgery in 1981-1991. Peritoneal dialysis was performed using bicarbonate dialysis solutions. Gentamycine was routinely added to the dialysate. Three passes were performed per hour with 10 cc/kg bodyweight as standard protocol.

	1981-1985	1986-09/1991
OP with HLM	1535	1787
OP-mortality	75 (4.9%)	68 (3.81%)
Dialyses	20	31
D.-mortality	17 (85%)	16 (51.6%)

Mean duration of dialysis was 169±314 hours (7-1800), the interval between operation and onset of dialysis was 38±30 hours (0-120). An onset of dialysis within the first 24 hours was predictive of a poor outcome. Time between onset of renal insufficiency and start of dialysis had no influence on the prognosis. Age at time of operation is an important predictor of death. Under one year of age mortality was 74.1% (20 out of 27), compared to 54.2 % (13 out of 24) in older children. Duration of bypass was not predictive.