Volume 4 • Supplement 1 • Pages 1-32

Cardiology in the Young

ISSN 1047-9511

Abstracts of the XXIX Annual Meeting of The Association of European Paediatric Cardiologists, Edinburgh, 11-14 May 1994

01

Intravascular stent implantation for the treatment of peripheral pulmonary stenosis—a multicenter study

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Stenosis of branch pulmonary arteries is a challenging therapeutic problem. Intravascular stent implantation was recently introduced as an alternative to surgery and balloon dilatation. We present our experience in 18 patients with mean age 10.1 yr (range 4-20) and mean weight 29.6 kg (range 12-65). The most frequent diagnosis was postoperative tetralogy of Fallot (13 pts); one patient had native pulmonary stenosis. Various operations had been performed in 16 patients; eight had previous surgery on the pulmonary branches and nine balloon angioplasty. Ten patients had bilateral stenoses. Twenty-eight stents were implanted in 25 branches (13 right, 12 left); seven patients had bilateral stents; one patient underwent intraoperative implantation. At angiography, the mean increment of the diameter of the lesion was 165% (range 59-600%; $p<0.01$); right ventricular-to-systolic pressure ratio decreased from 0.78 (range 0.36-1.09) to 0.64 (range 0.33-1; $p<0.05$). No major complications nor stent displacements occurred. In conclusion, intravascular stent implantation is a safe and effective procedure to relieve both postoperative and native pulmonary branch stenosis. The immediate hemodynamic results are superior to surgery and balloon dilatation; long-term follow-up needs to be evaluated.

02

Early results of stent implantation for pulmonary artery and right ventricular outflow tract stenoses

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Balloon dilation for acquired or congenital pulmonary artery stenoses is disappointing. Large balloons are required with the risk of pulmonary artery rupture, and these only produce a low success rate due to undilatable stenoses which recoil after the balloon is deflated. Metal stents which are deployed using high pressure balloons are more successful. We have implanted 20 Palmaz-Schatz stents (1.8, 2.0 or 3.0 cm long and 7-13 mm in diameter) in 12 patients with pulmonary artery stenoses or right ventricular outflow tract obstruction. Six patients had Fallot's tetralogy—three had previous multiple shunt procedures and three complete correction. One patient had pulmonary atresia/VSD with a shunt-related pulmonary artery stenosis. Five patients had congenital pulmonary artery stenoses—two had Alagille's syndrome, one had a VSD, one had infundibular stenosis and one had multiple peripheral pulmonary artery stenoses. Balloon dilation had been previously performed in four with little improvement. Stent implantation improved the stenosis diameter from 5±1.5 to 10±2 mm with a fall in the transstenotic gradient from 43±10 to 8±5 mm Hg. The right ventricular pressure fell in those without a ventricular septal defect and there was a rise in the distal pulmonary artery pressure in those with a systemic pressure right ventricle. In one patient, a stent was redilated from eight to 12 mm six months after the initial implant. In four patients with a single source of pulmonary artery blood flow, lung perfusion increased on the stented side from 36.5±23.3 to 54.3±24.6% of total lung perfusion. Two patients with Alagille's syndrome are now awaiting a liver transplant. Follow-up extends to 21 months (mean five months). Proximal displacement of the stent into an aneurysmal main pulmonary artery (repaired Fallot's tetralogy) occurred in one patient. The stent was removed at surgery 48 hours later. In one patient with multiple peripheral pulmonary artery stenoses, a distal pulmonary artery branch was perforated with the guide-wire and the vessel was occluded with coils at the same procedure. Stent implantation into pulmonary arteries is a useful method of relieving postoperative and congenital pulmonary artery stenoses.

03

European clinical trial of atrial septal defect closure with the buttoned device—early and mid-term results in 125 patients

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Transcatheter closure of atrial septal defect (ASD) was performed in 125 pts between 7/90 and 9/93 using second and third generation devices. The patient age varied between one and 77 years and the defect balloon diameter between three and 25 mm; the mean Qp/Qs was 2:1. Defect occlusion was achieved in 114/125 pts (91%). There were 46 residual shunts (37%) by color flow mapping (CFM) acutely (33 trivial). Complications included unbuttonings in 11 cases (surgery in seven); transient cerebrovascular episode in a non-implanted case; trivial mitral and tricuspid regurgitation in six and three, respectively; seven additional cases had surgery because of inadequate occlusion or device malposition. Wire distortion was noticed in a small bunch of third generation devices; in three cases it resulted in wire migration (two operated) and in two small atrial perforations (two operated). This problem was related to violent introduction of the device and wire connection and was corrected with the introduction of a new pusher and the fourth generation device. At six month follow-up, we observed 47/55 pts with full occlusion and eight patients with partial occlusion; at one year follow-up, 14/14 pts had full occlusion or minimal shunts by CFM. In conclusion, the buttoned device effectively closed 86% of ASDs. Complications should decrease with better selection of patients and device size, more experience and the recent improvements of the device.

04

Early clinical experience with the fourth generation adjustable buttoned device

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The adjustable buttoned device (A-BD), was used in the repair of 42 patent ductus arteriosus (PDA), nine atrial septal defects (ASDs) and one ventricular septal defect (VSD). The A-BD includes a button loop which adjusts for ductal length and septal thickness. It incorporates strengthened wire connections and a new introduction (fourth generation) device. PDA pts received 15 and 20 mm devices through a 7 F sheath for ductal diameters from 2-10 mm. ASD and VSD patients received 25-50 mm devices through 8-9 F sheaths for defects 8-25 mm. Patient age varied between six months and 76 years. All patients had effective occlusions with only trivial residual shunts. The full occlusion rate averaged 60% acutely, with a tendency for more complete occlusions on follow-up. No unbuttonings, embolizations, wire fractures or migrations were noticed. The early clinical results with the A-BD are encouraging and superior to the results of the previous BD generations, mainly because of the lack of complications and the wider clinical application.

05

Retrievable coils for interventional treatment in congenital heart disease

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In 1864 Moore performed the first coil occlusion of an aortic aneurysm placing coiled metal wire percutaneously through a cannula. In order to enlarge the thrombogenic surface, Colt (1903) invented a transcutaneously implantable single or double umbrella system consisting of wisps of steel wires. With the modifications of Gianturco, coil occlusion gained clinical acceptance. These cylindrically shaped coils are not repositionable or retrievable, involving the risk of peripheral embolization in high velocity arterovenous shunts or congenital cardiovascular malformations like the patent ductus arteriosus (PDA). We have developed a new occlusion device with memory-shaped retrievable coils (double-disk shaped; pfm, Cologne, Germany). The varying size and shape of these coils allows adaptation to the individual vessel. The external windings serve for safe fixation and the small inner windings for complete occlusion, even if no thrombogenic fibers are added. A mechanical snap-in mechanism keeps the coil attached to the core wire before final detachment. This allows retrieval back into the low-friction implantation catheter size 3 to 5 F (1.0/1.7 mm). Experimental evaluation demonstrated its use even in small piglets (≥1.8 kg). Clinically, we have occluded bronchial and coronary fistulas (n=4) as well as 33 PDA in 37 patients (four months to 21 years, 4.5 to 64 kg). Only in the PDA group was residual shunting detected by color Doppler echocardiography in five patients (follow-up one to 12 months). No coil embolization, no hemolysis, no endocarditis occurred. Minor problems included tricuspid valve entanglement (one patient), and intracatheter coil disconnection (two patients). Surgical interventions were never necessary. Further studies will show whether these three-dimensionally adaptive coils are suitable in lesions with changing wall diameters.

06

Interventional closure of the persistent neonatal ductus arteriosus Botalli (PDA) using a new, very small device—an experimental study

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We report the development of a new device (pfm, Cologne, Germany) to close the ductus in newborns using memory-shaped double disk coils (stretched wire diameter 0.018 inch) which are mounted on a titanium/nickel core wire (diameter 0.25 mm). A snap-in mechanism keeps the coil attached to the delivery wire, and thus the coils can be retrieved and repositioned if inadequately selected or placed. The system is positioned through a 3 F Teflon or Nylon catheter. In order to overcome inadequate friction or thrombosis the catheters and the pusher-wires had to be heparin-coated. Additionally, the push-wires had to be Teflon-coated. The system was tested in six newborn piglets (12 hours to 10 days, 1.5 to 4.5 kg). Ductal patency was secured by stents (3.5 to 4.5 mm inner diameter). Under general anesthesia, a cut-down of the external jugular vein was used to deliver the system through the right heart and placing it into the PDA under fluoroscopy. In all cases, adequate placement was possible. Due to high blood flow velocities in the PDA, the coils had to be repositioned up to six times. In one instance it was necessary to place a balloon into the PDA in order to lower the shunt size before final placement of the coil. The PDA closure was evaluated by aortic angiogram or color Doppler echocardiography. In four cases, the PDA was completely closed within one hour after coil placement. In two cases with large PDA s (4.5 mm), residual shunting remained, even after placement of an additional coil; hemodynamic relevance, however, diminished in both. In conclusion, our new very small device enables interventional PDA closure in the neonatal period. Our experimental study demonstrates as well the obstacles to overcome while closing window or tube shaped PDA s with high shunting.

07

Is arterial duct angioplasty safe?

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We have shown that thermal angioplasty of the arterial duct (AD) results in 80% long-term patency in lambs. To determine the feasibility and safety of AD balloon angioplasty in neonates, we reviewed 12 (eight males, four females) who had 19 AD angioplasty procedures. Mean weight 3.4±0.7 kg. Diagnoses were hypoplastic left heart syndrome (HLHS) (n=4), interrupted aortic arch (IAA) (n=4), critical aortic stenosis (AS) (n=1), critical pulmonary stenosis (PS) (n=2) and severe coarctation of the aorta (n=1). Mean age at presentation was 3.8±2.4 days. All were receiving PGE_2 before angioplasty with poor peripheral pulses, except for the two PS who were markedly cyanosed. Mean arterial pH was 7.0±0.9. Median time from admission to first angioplasty was six hours. Angioplasty was performed via the right heart. Saline/contrast was used for inflation in 14 episodes and CO_2 gas in six (one had both). The number of inflations ranged from one to five (median two). Bradycardia occurred in six episodes, of which three required adrenaline, two calcium, two external cardiac massage and one recovered spontaneously, all with no sequels. One HLHS died on the catheter table but had presented with asystole. Hemodynamics improved immediately in all others with resolution of metabolic acidosis and increased arterial oxygen saturation. Six ADs were explanted patent at surgery, median 26 days (range 3-245 days) after angioplasty. One PS AD closed at six months, the other is patent but constricted at 22 months. The AS AD is patent at five months. Two HLHSs and one IAA died with patent ADs 0-22 days post-angioplasty. We conclude that angioplasty of the AD in critically ill neonates with AD dependent circulations is safe and valuable for resuscitation. A method such as thermal angioplasty or stenting is required for long-term wide patency of the AD.

08

Long-term follow-up after valvotomy for critical aortic stenosis (AS)—predictors of survival and reintervention

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This study of 43 consecutive pts (35 M, 8 F; median age under two wks, range 8-80 d) assesses the influence of preop variables on the early and late outcome after valvotomy for critical AS. Valvotomy was performed in 41 pts, excision of myxoid valve tissue in one and conduit interposition between LV and aorta in one at 1 d–2.9 yr, median 19 d. Sixteen pts (37%) died after the first op (2 d-3.4 yr, median two mo), 10 early, six late, none after 3.4 yr postop. Follow-up of the 27 survivors after first op ranged from 6.9-19.3 (median 14.3) yr. The incidence of reinterventions accumulated with increasing age, with 10 pts requiring a second and three pts a third procedure, e.g. three revalvotomies, two balloon dilations (BD), three aortic and one mitral valve replacement. Among the remaining 17 pts with one op only, nine are asymptomatic, four have moderate re-AS ± aortic insufficiency (AI) 2°, one mitral stenosis and PH, one AI 3°, and mitral insufficiency 3°, PH and LV-and RV failure, and two are severely mentally retarded. By univariate analysis, none of the preop clinical and cath data demonstrably influenced survival. Young age (<2 weeks) at presentation and first op, small LV (angio LVEDV<100% norm), small aortic orifice (<7 mm) and ascending aorta (<10 mm) and uni (9%)-or bicuspid (67%) valve were associated with higher mortality and reintervention. This combination, closely related to HLHS, was present in 8/16 nonsurvivors and 1/27 survivors, making it a risk factor. Irrespective of the complexity of critical AS, in the young infant, valvotomy or BD may be performed to enable survival. Infants with unfavorable anatomy, persistence of LV dysfunction, duct-dependence and PH, may be candidates for Norwood op or heart transplantation. With recurrent obstruction of a bicuspid aortic valve, autograft replacement of aortic valve instead of repeat valvotomy or BD is an option.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

09

Long-term follow-up of patients who survive initial intervention for critical aortic stenosis

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Factors which affect the long-term prognosis in patients with critical aortic stenosis (AS) who survive more than one month after initial intervention are undefined. Sixty-four patients presented with critical AS between 1979 and 1992. Forty-one patients (64%) survived for more than one month after initial intervention (surgery 39, valvuloplasty 2). The median duration of follow-up was three years (range one to 15 years). Twenty-eight patients had a satisfactory result with mild or moderate AS or regurgitation without further intervention. Thirteen had a poor result with either reintervention (n=6) or death (n=7). The aortic valve diameter at presentation was significantly smaller ($p<0.2$) in patients with a poor result (median 5.5 mm; range 5-15) compared with patients with a satisfactory result (median 8 mm; range 5-10). Only one patient with a satisfactory result had an aortic valve diameter of <6 mm. Significant residual AS was present from the time of initial intervention in nine patients (69%) with a poor result. There was no difference in the incidence of a duct-dependent systemic circulation, other cardiac lesions, mechanical ventilation, acidosis or the use of inotropes preoperatively between patients with a satisfactory or a poor outcome. There was no significant difference in the increase in the aortic valve diameter at follow-up between patients with a satisfactory and a poor outcome. A small aortic valve diameter at presentation and residual stenosis after intervention were important predictors of a poor prognosis.

10

The role of balloon valvuloplasty for congenital aortic stenosis as compared with contemporary surgical results

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The purpose of this study was to compare the results of balloon dilatation and surgery for valvar aortic stenosis in infants and children undergoing aortic valvotomy. Between August 1988 and October 1993, 34 children underwent attempted balloon valvuloplasty (age range one day to 15 years, weight 1.72 to 53 kg). Four had a previous surgical valvotomy prior to 1988. During the same period, 22 children underwent direct surgical valvotomy, five of whom had had a previous balloon procedure prior to 1988. Infants with critical aortic stenosis or older children in whom the LVOT systolic Doppler gradient was >60 mm Hg were included in the study. Their age break-down was: <2 mo (Balloon (B)=8, Surgery (S)=7), <5 yrs (B=11, S=6) and <16 yrs (B=15, S=9). For balloon valvuloplasty, the retrograde femoral arterial approach was used, with a balloon diameter not greater than the diameter of the aortic valve as assessed both by echo and angiography. Successful balloon valvuloplasty was achieved in 33 (97%) with immediate reduction in the instantaneous mean systolic pressure gradient from 79 to 36 mm Hg. There was no significant difference in mortality, morbidity, or need for a repeat procedure within one year. There were two deaths in each group from poor cardiac output complicated by sepsis, with a third child from the surgical group dying with incessant VT/VF after AoV replacement. The main complications in the balloon group were significant aortic regurgitation in two patients and femoral artery tear requiring reconstruction in one patient. Follow-up from four months to four years showed sustained results in most cases. One required AoV replacement and three underwent second balloon valvuloplasty for restenosis. Hospital stay was shorter ($p<0.001$) in the balloon group (2-14 d) than the surgical group (5-24/mean 8 d). We conclude that balloon dilatation for congenital valvar aortic stenosis is effective and safe.

11

Balloon aortic valvuloplasty in children following previous surgical valvotomy

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There are few data on the efficacy of balloon valvuloplasty (BV) for recurrent aortic valve stenosis following previous surgical valvotomy (SV). Of 209 patients who have had SV over a 20 year period, 22 patients (18 M, 4 F), median age 157.5 (range 12-254) months, underwent 25 BV procedures at a median of 72 (8-155) months after surgery. The median age at SV was 82.5 (0.5-230) months, and the systolic gradient across the aortic valve immediately post-surgery was 31 (0-49) mm Hg. The indication for BV was a Doppler-derived peak instantaneous gradient (PIG) of >60 mm Hg, with or without associated symptoms, and with grade 2 or less aortic regurgitation (AR). A single balloon was used, and the median balloon to annulus ratio was one (0.9-1). Following BV the catheter pullback gradient decreased acutely from 55 (35-75) to 30 (0-75) mm Hg ($p<0.01$) and the Doppler gradient from 74 (52-92) to 40.5 (30-96) mm Hg ($p<0.01$). In three patients who underwent a second BV eight months after the first procedure, the pullback gradient decreased from 50 (50-60) to 15 (15-16) mm Hg. AR grade increased from one (0-2) to two (1-3); only one patient had grade 3 AR. Over a median follow-up of 33 (2-67) months, seven patients have undergone aortic valve replacement for recurrent stenosis (six patients) or AR (one patient with grade 3 AR post-BV, who had partial detachment of one of the valve leaflets along the hinge point). On comparing the 12 patients with a 'good' result (not requiring further BV or surgery) and the 10 patients with a 'poor' result, there was no significant difference ($p>0.01$) in the pullback gradient post BV (median of 19 vs 32.5 mm Hg), Doppler gradient 24 hours post-BV (33.5 vs 50.5 mm Hg), or duration of follow-up (27.5 vs 18 months). BV is a safe and feasible option, but a significant proportion of patients have progression of stenosis and will require further interventions.

12

Mid-term results of transcatheter aortic valvotomy in 25 neonates with critical aortic stenosis

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At a mean age of 10.6±10.9 days (range 1-37), a transcatheter valvotomy was attempted in 27 patients presenting early after birth with a stenotic aortic valve and severe cardiac failure. It failed in two cases. The mean LV end-diastolic dimension (EDD) was 19±4.6 mm, but five patients had a very small LV (EDD <12 mm) with other features belonging to the hypoplastic left heart syndrome and all of these either died (3) or had to be transplanted (2) shortly after a successful dilation. Among the 20 dilated infants with a large LV, three died two to 26 weeks after the procedure because of associated endocardial fibroelastosis with mitral valve (MV) lesions; and another two had to undergo quick surgical valvotomy—one is still alive and doing well but the other subsequently suffered from MV dysfunction, had a MV replacement at three years and died from thrombosis of the prosthesis. The remaining 15 patients are alive at a mean age of 31±27 months with a mean LV-aortic systolic gradient of 39±11 mm Hg (range 25-60). Aortic regurgitation is present in six children, mild in three, moderate in one, but severe in two, leading in one to a successful Ross operation at 18 months. Severe MV regurgitation progressed in two infants who required MV replacement at seven and eight months. In conclusion, transcatheter aortic valvotomy is a quite safe and efficient procedure in the neonate with critical aortic stenosis. The mid-term prognosis is mainly linked to the size of the LV and to associated lesions such as endocardial fibroelastosis and MV abnormalities.

13

Pulmonary autograft root replacement for aortic valve disease in children

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Aortic root replacement with pulmonary autograft offers an advantageous surgical alternative for aortic valve disease in children because the autograft is autologous, needs no anticoagulation and has growth potential. From 1988 until 1993, this procedure was performed in 15 children (mean age 10.8 yr, range 0.3-15.0; 11 males). Aortic stenosis (AS) was the indication for operation in three pts, aortic regurgitation (AR) in five and both in seven. Twelve pts initially had congenital AS, one pt had juvenile rheumatoid arthritis (JRA) with AR, one pt had rheumatic fever (RF) with AR. Ten pts were previously operated (valvotomy in nine pts, including one with enucleation of a discrete subaortic stenosis (DSAS) and only enucleation of DSAS in one pt). Pulmonary autograft replacement was done as root replacement with reimplantation of the coronary arteries in the pulmonary autograft and orthotopical reconstruction of the pulmonary root with a pulmonary allograft (12 pts) or aortic allograft (three pts). There was no early mortality. One patient had postoperative AV-block needing pacemaker therapy. Mean follow-up time is 3.0 yrs. Late mortality concerned two pts. One pt died after nine months due to persistent fibroelastosis of the left ventricle, severe regurgitation of the pulmonary homograft being present as well. The pt with JRA died after six months due to a recurrence of JRA with neoaortic valve involvement. In the pt with RF, a recurrence with neoaortic valve involvement led to prosthetic valve replacement of the neoaortic valve after 22 months. The 12 functioning autografts show no stenosis. No regurgitation is present in one, trivial in 10, mild in one. The homografts show no stenosis. No regurgitation is present in 10, trivial in three. In conclusion, pulmonary autograft aortic root replacement is perhaps the best surgical alternative for aortic valve disease in children, with good mid-term results.

14

Percutaneous commissurotomy for rheumatic mitral stenosis in children—efficacy and limits

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The application of percutaneous mitral commissurotomy (PMC) for rheumatic mitral stenosis in children remains controversial. Fifty-six children (age 7-18 years, mean 14 yrs) underwent PMC as a common experience in two centers. All were in sinus rhythm. Four were in NYHA class IV, 29 in class III and 23 in class II. All but two had pulmonary hypertension. Procedure was performed without general anesthesia through transseptal approach by femoral vein puncture. Inoué balloon was systematically used since 1990; in other children, single or double balloon was employed. There was no death, no cardiac tamponade, no systemic embolism. One technical failure occurred at the beginning of the experience, with successful procedure two days later. LA/LV hemodynamic gradient decreased from 21 to 7 mm Hg ($p<0.001$). Mean pulmonary artery pressure decreased from 40 to 26 mm Hg ($p<0.001$). Cardiac index remained stable (3.1 to 3.3 l/min/m^2). Non-indexed mitral area increased from 0.82 to 1.86 cm^2 ($p<0.001$). The major problem was the occurrence of severe mitral regurgitation in three cases requiring early valve replacement. No correlation could be found between valvular and subvalvular anatomy, and result of the procedure in terms of residual stenosis and acquired regurgitation. During the follow-up period, five other patients needed mitral valve replacement for restenosis, without anatomic possibility of redilatation or open heart commissurotomy. In conclusion, children with rheumatic mitral stenosis are good candidates for PMC, without any particular difficulties or complications. Anatomic evolution is more rapid than in adults, with higher risk of premature restenosis, without anatomic possibility of valvular preservation. However, PMC may be considered as the first choice therapy in children with rheumatic mitral stenosis.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

15

Balloon valvuloplasty of the aortic valve in young infants

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The aim of this study was to prove safety and feasibility of balloon valvuloplasty of the aortic valve in young infants who presented with congestive heart failure. Between January 1986 and June 1993, 49 newborns and infants with significant aortic stenosis were scheduled for balloon valvuloplasty. The age ranged from one to 235 days (median age 24 days). In 45 cases the procedure could be performed, and in four patients the balloon catheter could not be placed correctly. In 12 patients a percutaneous access was used, whereas in the other patients a transcutaneous approach from the right axillary was chosen. There was no mortality related to the procedure. Major complications occurred in three patients. There was no major aortic regurgitation following the procedure. Follow-up ranges from 1.5 months to 7.3 years. Patency of the peripheral vessels are controlled by clinical examination and pulse Doppler studies. Twelve children did not survive the first year of life. Six of them had endocardial fibroelastosis with small mitral valves and left ventricles. In conclusion, balloon valvuloplasty can be performed at low risk in very sick young infants. Only those patients with a normal sized left ventricle will clearly benefit from this procedure.

16

Early and intermediate results of balloon dilation of aortic stenosis in the first three months of life

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Balloon dilation of the aortic valve was performed in 24 infants (15 males and nine females), using a right carotid artery approach. Age ranged from one to 75 days (19 pts aged less than 30 days) and weight from 2.1 to 6.3 kg (mean 3.46 kg). Associated lesions were present in eight pts (aortic coarctation 3, ventricular septal defect 1, mitral incompetence 2, mitral stenosis 1, pulmonary stenosis 1, left ventricular and/or aortic hypoplasia 1, patent ductus arteriosus with persistent pulmonary hypertension 1). Seventeen pts were in critical condition, 15 of them in the first month of life. Balloon dilation was accomplished in all pts. In three we had complications—two perforations of the left ventricle by the guide-wire, with pericardial effusion immediately drained without consequences and one undetected aortic dissection with death for aortic rupture 24 days later. Immediate results showed dramatic improvement in clinical conditions with a mean percent reduction of gradient of 66% and mean increase of left ventricular ejection fraction of 30%. Moderate aortic incompetence developed in seven pts. Seven pts (29%) died, five for associated lesions, one for aortic rupture and one for severe aortic regurgitation and refractory cardiac failure. The 17 surviving pts are doing well. During a mean follow-up of 29 months (2-74 months), five pts developed restenosis, three were successfully redilated and two underwent surgery. Two other pts still have a significant peak gradient and three have a moderate aortic regurgitation. In our opinion, balloon dilation is a simple and effective procedure for the treatment of aortic stenosis in infants.

17

Balloon expandable stents in the treatment of branch pulmonary artery stenosis

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Eleven children underwent endovascular placement of a balloon expandable metallic stent (Palmaz) in the treatment of branch pulmonary artery stenosis. Age ranged from seven to 15 yrs (mean 10) and average weight was 35 kg. Seven patients had tetralogy of Fallot, three had congenital pulmonary artery stenosis and one had left pulmonary artery stenosis following a Fontan procedure for tricuspid atresia. Nine of the patients had undergone previous cardiac surgery for the stenosis. From a femoral venous or right internal jugular approach, a long 11 French gauge curved sheath was positioned across the area of stenosis and either a 30 mm (nine pts) or 18 mm (two pts) long stent was deployed using a heavy-duty 12 mm diameter balloon angioplasty catheter. The stent was further dilated with a 15 mm angioplasty catheter in four patients. Hemodynamic and angiographic data were obtained before and after stent placement. An increase in pulmonary artery diameter was seen in all patients. Mean diameter before of 4.9 mm (range 2-8) increased to 11.2 mm (7-13) after stenting. The average increase in vessel caliber was 145% (range 63-300). Peak-to-peak systolic gradient across the stenosis decreased from a mean of 41 to 25 mm Hg. Average x-ray screening time was 41 minutes. Complications included balloon puncture/bursting in three patients, initial eccentric stent dilatation in two patients and blood loss requiring acute transfusion in two patients. Endovascular stenting is a safe and efficacious treatment for branch pulmonary artery stenosis.

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Use of self-expanding stents in congenital heart disease

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Self-expanding stents have potential advantages compared to balloon-expandable stents such as smaller delivery system, their length and their flexibility both prior to and after deployment. We report our prelimimary experience of their use in patients with congenital heart disease. We have inserted 10 self-expanding stents in 10 patients (age range 15 days-32 years, weight range 3-63 kg). The diagnoses were stenosis after a Fontan-type-operation (six patients), stenosed systemic-to-pulmonary collaterals in two patients with pulmonary atresia with VSD, idiopathic pulmonary vein stenosis (one patient) and coarctation of aorta in one patient after palliation for hypoplastic left heart syndrome. All patients had dilatable but unexpandable stenotic lesions. We used self-expanding Wall stents in nine patients (diameter 8-16 mm, length 15-48 mm) and Cook Z-stent in one patient (diameter 20 mm, length 80 mm). There were no procedural complications and the final position of the stent was felt to be optimal in all cases. In patients after a Fontan-type operation, the mean pressure gradient ranged between 1-10 mm Hg (median 5) before stent and was completely abolished in all ($p<0.001$). The three youngest patients subsequently died due to events unrelated to the stenting procedure. Follow-up in the remainders (3-15 months) demonstrated no stenosis across the stent. We conclude that the use of self-expanding stents should be considered for the treatment of tortuous and long or multiple sequential stenoses.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

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Balloon valvuloplasty of pulmonary valve in tetralogy of Fallot—influence on pulmonary artery size and oxygen saturation

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We retrospectively studied 24 patients (age three days-22 months, BSA 0.2-0.8 m^2) with tetralogy of Fallot in whom balloon valvuloplasty was performed between 1990 and 1993. Indications were severe cyanosis (n=24), hypoxic attacks (n=10) and coronary artery anomalies (n=2). Diameters of pulmonary valve, right and left pulmonary artery were determined by cineangiography before and after the procedure. Pulmonary artery index (PA-I mm^2/m^2) was calculated as ratio between area of right and left pulmonary artery and body surface area. Mean pulmonary annulus had a diameter of 5.7 mm (range 3.5-7.9). The balloon/annulus ratio was 120%. Systemic O_2 saturation improved from 75-85% after dilatation ($p<0.001$). Five patients were excluded from further measurements because of preceding palliative operations or other morphologic anomalies. Mean PA-I before dilatation was 177.5 mm^2/m^2 (range 98-317). In 11 patients angiographic control at the end of the procedure showed a PA-I of 209 mm^2/m^2 (range 111-333, $p<0.05$), reflecting dilatation of the vessels by increased flow. Catheterization before corrective surgery revealed a PA-I of 200.7 mm^2/m^2 (one pt with a PA-I of 507 mm^2/m^2 one year after the procedure was excluded). In six patients a second successful valvuloplasty was performed. Severe complications were hypoxic attacks during procedure (n=3) and femoral vein thrombosis (n=3). Emergency implantation of Blalock-Taussig anastomosis was necessary in one. Corrective surgery was performed after a mean period of 9.6 months after palliation. In conclusion, balloon valvuloplasty in tetralogy of Fallot is a safe procedure to improve systemic oxygen saturation of the patients. Hypoxic events can be avoided and optimal timing for corrective surgery is possible. Relevant growth of the pulmonary vessels is not achieved.

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Transthoracic real-time three-dimensional echocardiography by echo-CT provides new information about cardiac anatomy

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The purpose of the study was to determine in which specific heart defects three-dimensional (3D) can provide additional information over conventional 2D-echocardiography or angiography. 159 patients aged one day to 19 years (mean age 4.3 years) with normal hearts (n=3) or a variety of congenital heart defects (n=156) were examined with echo-CT. The tomographic ultrasound probes of 3.25, 5 or 7.5 MHz frequency acquire parallel images of the heart. The probe is steered by a stepper motor, which moves in 0.5 mm steps and acquires an image of the heart at each step with ECG and respiration-gating. Between 50 and 130 slices of the heart were thus acquired, which took 2-6 minutes. 3D reconstructions were possible in all and took 20-120 minutes. Information not detected by other imaging modes and confirmed at subsequent surgery was found in eight patients. Two had complete transposition of the great arteries (TGA) with LV outflow obstruction caused in both cases by abnormal insertion of the mitral valve, one of those had a ventricular septal defect (VSD). In one patient with TGA and subpulmonary VSD, the extent of straddling of the mitral valve could be better seen than with 2D echocardiography. One infant with VSD and coarctation had a supravalvar mitral membrane, which was only seen by Echo-CT. In two cases with subaortic obstruction (SAS), the extension of the obstruction to the mitral valve could be demonstrated. One developed severe mitral regurgitation following SAS resection and Echo-CT could precisely define a localized hole in the aortic mitral valve leaflet, which the surgeon had created on resection of the SAS. Two with AVSD were found to have a double orifice mitral valve. We conclude Echo-CT yields useful information unavailable by other imaging techniques especially on the morphology of the LVOT in patients with normally related great arteries or TGA and on the morphology of AV valves.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

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'White blood' imaging of the cardiovascular system using a rapid echo flow-rephased spin echo technique

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Flow rephased gradient echo techniques provide images with high signal from flowing blood. Disadvantages are the sensitivity to static field inhomogeneities (metallic implants, lung vessels), the lack of T1-contrast and saturation effects, which lead to signal loss or diminished separation of adjacent vessels. This however is crucial, especially in patients with pulmonary atresia, where imaging of severe hypoplastic vessels is required. Therefore we developed a flow rephased spin echo technique (REFRESH), which offers 'white blood images' of high quality almost free of artifacts. This technique was applied in 40 children with congenital heart diseases (18 PA with palliative shunts, one aplasia of left pulmonary artery, two PS, six D-TGA, three TAPVC, one DORV, four coronary artery aneurysms, four vascular rings, one RV-diverticulum) and compared with 2D-gradient echo images. As on gradient echo images, even small pulmonary arteries (8) were clearly differentiated from bronchial structures and adequately functioning shunts caused signal loss in the pulmonary artery due to turbulent flow (15). As the pulmonary vessels were traced to subsegmental levels, the perfusion of the lungs could be better estimated on the REFRESH-images (13) and in one patient peripheral pulmonary artery stenosis could be excluded. Four coronary aneurysms were better demarcated. Signal loss due to metallic implants was reduced in three patients, as saturation effects in four patients, where this technique was tested for angiographic projections. REFRESH is a new MR imaging modality, which combines and improves the positive features of conventional spin echo technique and gradient echo technique.

22

Value of combined transesophageal echocardiography and acoustic quantification for the detection of abnormal elastic properties of the aorta in patients with Marfan's syndrome

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Prognosis in Marfan syndrome (MAS) is strongly influenced by complications due to diseases of the aorta. Evaluation of the elastic properties (EP) allows early detection of these abnormalities even in morphologically normal vessels without significant dilatation. In 10/17 patients (pts) with MAS (mean 31 years), and in a control group of 12 normal pts (mean 27 years, p=ns), transesophageal echocardiography was performed. Descending aorta (AOD) was visualized in a short-axis view, acoustic quantification (AQ) was used for the on-line evaluation of maximal area changes of AOD over time (dA/dt). Maximal and minimal diameter (dmax, dmin in cm) were also obtained from 2D stop frame images. The following parameters were calculated using noninvasively measured blood pressure: compliance (C; 10^3 cm^2dyn^{-1}), distensibility (D; cm^2dyn^{-1}) and stiffness index (SI). There were highly significant differences between the two groups in all parameters. In conclusion, in MAS, even in absence of vessel dilatation, EP of the AOD are significantly different from normal controls. On-line assessment of maximal dA/dt by AQ correlates well with standard measurements of C, D and SI, allows differentiation from normals, and represents a new, safe and less time consuming method for the evaluation of this disease.

23

Postoperative quantification of pulmonary insufficiency in tetralogy of Fallot—correlation with right ventricular function

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We studied the outcome of surgery for TOF in 42 pts (17 M, 25 F, age at repair 3 mo-2 yr; without patch-valvotomy [n=10], infundibular patch + valvectomy [n=17], transannular patch [n=15]) by 2-D-echocardiography (2D-E) and Doppler color flow imaging. Simpson's rule was applied to determine right ventricular volume and ejection fraction (RVEF) echocardiographically, and the results were compared with angiocardiographic data (biplane Simpson's rule). The area of the color Doppler pulmonary insufficiency (PI) jet was calculated and normalized for body surface area (PI-index, PI-i, cm^2/m^2). Furthermore, pulsed Doppler echocardiography was used to detect the distal retrograde flow in the PA and PI was graded 1-3 according to Kobayashi. Right ventricular end-diastolic volume assessed by 2D-E showed good correlation with angiographic data (r=0.91) as did RVEF (r=0.94). In patients with grade 2-3 PI, the PI-i was above 3.5 cm^2/m^2 (mean 6.4 cm^2/m^2 and was less than 1.4 cm^2/m^2 in those with PI grade 1 (mean 0.8 cm^2/m^2 $p<0.01$). PI-i correlated inversely with RVEF (r=0.82). We conclude that echocardiography provides reliable data for the determination of PI and RVEF. Patients after Fallot repair with PI index above 3.5 cm^2/m^2 and impaired RV function may be liable to be candidates for early valve replacement.

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Assessment of right ventricular volumes in children by multisection gradient echo magnetic resonance imaging

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Multisection magnetic resonance imaging (MRI) of the right ventricle (RV) provides adequate volume measurements without geometrical assumptions validated in adults. Few data exist on its application in children. We assessed the accuracy of RV volume measurements by multisection MRI in children. A contiguous set of transverse sections encompassing the ventricles was acquired with cardiac triggered gradient echo MRI in 31 children (age 10±3 years), 21 with normal hearts, 10 with (operated) congenital heart disease (without left-to-right shunting). To increase accuracy, slice thickness was less than standard (8 vs 10 mm). Ventricular cavity area was determined by manual tracing of the endocardial contour and multiplied by section thickness. RV and LV volumes were calculated by summation of the section volumes at end-diastole and end-systole. Internal validation of the technique was used by comparing RV with LV stroke volumes. Eight to 10 sections were adequate to image both ventricles completely. Measured volumes were as follows: RVEDV 82±28 ml, RVESV 25±9 ml, RVSV 55±21 ml, LVEDV 79±28 ml, LVESV 23±10 ml, LVSV 55±19 ml. Linear regression analysis of correlation between RV and LV stroke volume: RVSV=1.03, LVSV=1.6, r=0.96, SEE=6.2. In conclusion, transverse gradient echo MRI with 8 mm section thickness enables accurate measurement of RV volumes in children. This technique provides a basis for noninvasive functional analysis of the RV with a complex shape, as is seen in many forms of congenital heart disease.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

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Magnetic resonance imaging in the preoperative assessment of Fontan patients

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The aim of this study was to assess the value of magnetic resonance imaging in patients being considered for a modified Fontan procedure. We reviewed the findings of echocardiography and magnetic resonance imaging in 45 patients who had both studies during their preoperative evaluation. A 10 point scoring system, assigning one point to each of the following issues relevant to Fontan selections, was employed: systemic venous anatomy, central right pulmonary artery, central left pulmonary artery, distal right pulmonary artery (arborization), distal left pulmonary artery (arborization), right pulmonary venous return, left pulmonary venous return, right AV valve status, left AV valve status, ventricular function. Echocardiographic studies had an average score of 6.5. Magnetic resonance imaging supplied additional information in 29 patients (56%). The average amount of additional information using the above mentioned scoring system was 2.5. The additional information obtained by magnetic resonance imaging was judged to be important to the surgical management in 10 patients (23%). The average amount of useful information in these 10 patients was 1.6. In conclusion, magnetic resonance imaging is a useful technology in the assessment of selected patients being evaluated for a modified Fontan procedure.

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Hypoplastic left heart syndrome—early results following the introduction of a modification of the Norwood protocol

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The results of surgical palliation for hypoplastic left heart syndrome (HLHS) in the UK have been dismal. Since 1993, we have offered a modification of the Norwood protocol to all neonates presenting to our unit with HLHS or variants. Stage 1 surgery consists of a modification of the Damus/Norwood arch repair without use of exogenous material, ductal ligation, coarctectomy, atrial septectomy and insertion of a 3.5 mm Gortex shunt between the innominate artery and the right pulmonary artery. Cardiac catheterization is performed early (three months) and is closely followed by Stage 2 surgery. Arterial reconstruction is performed as necessary and the Gortex shunt is replaced by a superior cavopulmonary (CP) connection, to offload the ventricle and prevent subsequent pulmonary artery distortion. So far, 12 neonates (nine with HLHS and three with univentricular heart and severe systemic outflow obstruction) have been transferred to our unit. Eight were on prostaglandin infusions, five were acidotic, two were ventilator and inotrope dependent. In three infants, congenital heart disease had been detected antenatally. Parents declined surgery for two infants. Ten underwent Stage 1 surgery after stabilization with prostaglandin, inotropes and ventilation. There were three perioperative deaths. Two postoperative deaths occurred, one at six hours, and one, unexpectedly, at 14 days. Thus, five (50%) survived Stage 1 surgery. Three patients underwent elective CP shunt procedures at 3-5 months of age following cardiac catheterization, another had a CP shunt performed on the 19th postoperative day for failure to wean from the ventilator. All survive. The fifth Stage 1 survivor awaits cardiac catheterization and surgery. Successful medium-term palliative surgery for the HLHS can now be offered in the UK.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

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Noninvasive rejection diagnosis using antimyosin scintigraphy after heart transplantation in children

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Noninvasive methods for rejection diagnosis after heart transplantation (HTx) are of importance in pediatric patients (pts), since endomyocardial biopsies should be avoided. The validity of antimyosin scintigraphy (AMS) for diagnosis of acute rejection (AR) was tested after pediatric HTx and compared with echocardiography (Echo), clinical signs of AR and EMB performed in school age children. Fourteen HTx and two heart-lung transplantations (HLTx) were performed in pts with a mean age of 12.2 yrs, ranging from four days to 17 yrs. In these patients, 20 AMS-examinations were carried out from one wk to 3.5 yrs after Tx; 7μg/kg of ^{111}Indium labelled F(ab) fragments of the monoclonal antimyosin antibody were injected intravenously. Forty-eight hrs later, the scanning was performed in SPECT-(single photon emission tomography) or plane technique. For evaluation the ratio of counts per pixel, registered above the region of the heart and the lungs (H/L-ratio) was calculated. H/L-ratios>1.8 (SPECT-technique) or >1.6 (plane technique) were considered as a sign of AR. The survival rates of all children were 93.7% after three month and 87.5% after one year. Mortality due to AR did not occur. In six cases, AR was diagnosed clinically, by Echo or EMB and simultaneous AMS examinations were performed. In five of these cases, AMS revealed an increase of the H/L-ratio (mean 2.04±0.23, sensitivity = 83%); in one case a borderline H/L-ratio of 1.59 was accompanied by mild AR in EMB (IB ISHLT). Specificity of AMS was 100%. The sensitivity/specificity data for Echo were 100/54%. AMS is a reliable method for noninvasive AR-diagnosis in neonates and children. The use of antimyosin scintigraphy (with echo) results in a reduction or even avoidance of invasive EMB.

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Inhaled nitric oxide reduces pulmonary hypertension in infants with congenital heart disease

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Nitric oxide (NO) is an important endothelium-derived relaxing factor (EDRF) and inhalation of NO has been shown to produce selective pulmonary vasodilatation in lambs and in adults with pulmonary hypertension. The aim of the study was to investigate the effect of brief inhalation of NO in infants with congenital heart disease. Twenty-two infants and children aged 3-32 months with congenital heart disease undergoing preoperative cardiac catheterization were exposed to 40 ppm (parts per million) NO. All but one infant had intracardiac shunt lesions and thirteen had increased pulmonary vascular resistance (PVR). Pulmonary and systemic hemodynamics were evaluated prior to and after 10 minutes exposure to NO, using measured oxygen consumption and the Fick principle. No effect of NO was seen on the systemic circulation. Neither was there any effect on the pulmonary circulation in infants with normal PVR, whereas in infants with pulmonary hypertension, PVR was reduced by 34% from 8.6±4.6 to 5.7±3.5 mm Hg min•m^2•l^{-1}, mean and (SD). No statistically significant increase in methemoglobin was seen, although there were large individual differences. No other side effects were seen. Thus, brief inhalation of NO reduced a pathologically increased PVR without affecting systemic circulation and without causing important side effects.

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The Fontan procedure for pulmonary atresia with intact ventricular septum (hypoplastic right heart syndrome)—operative and late results

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From 1973 through August 1993, 855 Fontan procedures had been performed at the Mayo Clinic with 39 (4.5%) having been done on patients with HRHS. Age at surgery ranged from 1.8 to 21 years (median six). Thirty-eight of the 39 patients had a total of 71 prior palliative cardiovascular procedures. Tricuspid valve and right ventricular size ranged from 'minuscule' to a maximum of 50% of expected size. Only four patients had significant right ventricular to coronary artery fistulae and only one of these was 'right ventricular coronary dependent.' At Fontan procedure, the hypoplastic tricuspid valve was left patent to receive systemic venous blood in 19 pts, to receive diverted pulmonary venous blood in nine pts, was patch closed in seven pts, and in four pts the Fontan patch was placed across the tricuspid valve, hypoplastic RV, and opened RVOT to the MPA. There were three operative deaths (7.7%) and there have been two sudden unexpected late deaths (2½ and eight years post-Fontan). One patient had a successful cardiac transplant seven years post-Fontan. Present ages of survivors ranged from three to 29 years (median=11.5 years) with a median post-Fontan follow-up of 5.3 years. At recent follow-up, 33 of 34 survivors were in good or excellent condition and one was fair. Operative and late results of the Fontan procedure for HRHS have been good, and the procedure represents the best 'definitive palliation' available for these patients.

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Combined atrial and arterial switch procedure for congenital corrected transposition with ventricular septal defect

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Four children with congenitally corrected transposition and ventricular septal defect underwent combined atrial and arterial switch procedure to establish the left ventricle as the systemic ventricle after surgery. Two suffered from severe tricuspid regurgitation and one from gross right ventricular failure preoperatively. One child was operated electively following previous pulmonary artery banding. Age at operation ranged from 0.8-3.1 (mean 2.2) years. Postsurgical follow-up ranged from 3-18 (mean nine) months. There were no early or late deaths. Bypass time ranged from 145-194 (mean 167) mins. Progression of conduction abnormalities were encountered in two pts. In-hospital stay ranged from eight to 17 (mean 13) days. Cardiothoracic ratio decreased from a mean of 0.65 (range 0.6 to 0.71) preoperatively to 0.58 (range 0.52 to 0.6) at last follow-up. Latest follow-up examinations excluded significant residual lesions in all patients. Three patients are in functional class I and one child is in functional class II. The combination of an atrial and an arterial switch procedure in children undergoing surgery for congenitally corrected transposition reestablishes the left ventricle as the systemic ventricle. Initial experience suggests excellent immediate and early follow-up results.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

31

The fate of the pulmonary valve after total cavopulmonary anastomosis (TCPA)—potential hazards of systemic arterial embolization?

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The procedure of TCPA includes transsection and closure of the main pulmonary artery, while the systemic venous blood flow is directed to the right pulmonary by an anastomosis of the superior vena cava with the right pulmonary artery. Little is known about the fate of the residual pulmonary trunk and pulmonary valve. From 2/90 until 6/93, 39 consecutive pts underwent a TCPA in our unit. Detailed transthoracic echocardiography studies (TTE) of the pulmonary valve and residual pulmonary trunk were available in 20 pts who form our study group (four pts were excluded who had a congenitally atretic pulmonary valve). The mean age of these pts was 7.4 yrs, with a mean interval of 17 months between the TCPA and the TTE. The pulmonary valve and the pulmonary trunk were visualized in 18 pts (90%). Nine of these 18 pts (50%) showed diastolic regurgitation of the pulmonary valve into the ventricle. In one pt we found several thrombi in the residual pulmonary trunk. One pt with pulmonary regurgitation but without evidence of pulmonary thrombi developed an episode of cerebral infarction. Two pts without pulmonary regurgitation had a small residual communication between the pulmonary trunk and the bifurcation. In our experience, pulmonary regurgitation is frequently present in pts following the TCPA if the pulmonary valve is not closed surgically. The development of thrombi in the residual pulmonary trunk may increase the risk of systemic arterial embolizations. We therefore recommend surgical closure of the pulmonary valve in the procedure of TCPA. Visualization of the residual pulmonary trunk and the pulmonary valve should be attempted routinely in the postoperative echocardiographic assessment of all pts with previous TCPA.

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Evaluation of flow events at the ductus venosus, atrial septum, and aortic isthmus/ductus arteriosus in fetal lambs

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Direct observation of fetal flow events is now possible by ultrasound techniques. We studied nine fetal sheep at 134±6 days of gestation using 2D- and contrast echocardiography, conventional Doppler and color flow mapping by a transuterine approach. The fetuses were instrumented with catheters in the distal umbilical vein (UV) and the inferior and superior vena cava (IVC and SVC); a fetal ECG was also obtained for timing of the events. Fetal blood flow from the UV accelerated through the ductus venosus sphincter from a mean of 23 cm/s with absent phasic change to a biphasic signal with a velocity of up to 80 cm/s. The streams from the UV and the lower body remained separate in the proximal IVC with preferential streaming of UV blood into the left atrium. This was augmented at the atrial level where the Eustachian valve and septum primum formed a tubular conduit that directed UV blood into the left atrium and also prevented a reflux of left atrial blood into the right atrium. This conduit closed during atrial systole, but reopened throughout the rest of the cardiac cycle. Most of the SVC saline contrast was directed by the Eustachian valve into the right ventricle, but some entered the left heart by first refluxing into the IVC. Flow in the descending aorta just distal to the ductus arteriosus/aortic isthmus junction showed definite temporal differences and two distinct streams. Ejection was longer in the descending aorta than from either stream alone. The aortic isthmus stream arrived at a mean of 40 ms earlier than that from the ductus arteriosus and had a peak velocity averaging 15 cm/s lower than the ductal velocity. Thus, phasic flow events affect the circulatory dynamics to the benefit of the fetus.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

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Accessory pathway mediated supraventricular tachycardias in children—curative treatment by radiofrequency current catheter ablation

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Seventy-seven pts underwent radiofrequency current (C) ablation for drug refractory supraventricular tachycardias (SVT). In 70 pts (31 F, 39 M; 9.3±4.1 yrs), SVT was mediated by a total of 74 accessory pathways (APs), 42 of which had manifest (mfst) conduction properties (28 left-, 14 right-sided) and 32 were concealed (conc) (21 left-, 11 right-sided). Nine pts with conc AP had the permanent form of junctional reciprocating tachycardia (PJRT). Ablation was performed via a 5 to 7 French steerable catheter with a 3-4 mm tip electrode. For right-sided APs, the ablation catheter was advanced towards the atrial aspect of the tricuspid annulus, whereas in pts with a left-sided AP, the catheter was advanced retrogradely into the left ventricle and placed below the mitral annulus. In 32 of 40 pts with mfst preexcitation, the single catheter technique mapping and ablation of the AP was attempted and could be successfully carried out in 26. In the remaining six pts, a second catheter was required for eventual abolition of the AP. RFC (22±4 watts, 21±4 sec) was delivered during sinus rhythm in pts with mfst preexcitation and during tachycardia in pts with SVT due to a conc AP, including pts with PJRT. A median of seven RFC applications was required for successful treatment of SVT in 65 (93%) children. In the remaining five pts, only intermittent conduction block of the AP could be achieved which evolved into permanent block after two and four days in two of them, respectively. Procedure duration was 3.4±2.1 hrs, with a median radiation exposure of 35.2 min. During a 13.0±8.9 mths follow-up 63/67 pts were asymptomatic and required no antiarrhythmic medication. Seven pts had recurrences of SVT (5 APs in posteroseptal space), three of which underwent a successful repeat session. RFC ablation is an effective and safe therapeutic modality to cure accessory pathway mediated SVT in children.

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Radiofrequency (RF) ablation therapy for arrhythmias in children—lessons from initial experience

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The introduction of RF treatment for arrhythmias has revolutionized management and prognosis. Apart from achieving a cure or marked clinical improvement, there is the added advantage of avoiding long-term administration of anti-arrhythmic drugs. Since January 1992, we have replaced DC ablation by RF. Fifteen RF procedures have been performed in 12 symptomatic patients, three requiring a second application of RF. There were 10 females and two male patients with an age range from two to 17 years (mean 11.8). Two patients had ventricular tachycardia, three had the permanent form of junctional reciprocating tachycardia and the remaining seven had an accessory pathway, most of them with overt Wolff-Parkinson-White Syndrome. Of the seven accessory pathways, two were in the left free wall, two were in the right posterior wall and three were posteroseptal. All the accessory pathways were approached using a supra-annular approach. Mapping and ablation were performed under general anesthesia during the same sitting. Associated congenital heart defects included Ebstein's anomaly and aortic stenosis. There were no major complications or deaths. The arrhythmias have been abolished in all the patients although one patient with Wolff-Parkinson-White Syndrome still shows a subtle delta wave with a different vector possibly due to a separate pathway but remains symptom-free. None of the patients are on anti-arrhythmic therapy. In summary, RF ablation for arrhythmias in children is safe and successful although some may require a repetition of treatment. Technical considerations related to patient size and cardiac anatomy need to be taken into account.

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Propafenone safety in pediatric dysrhythmias—European retrospective multicenter study

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In order to investigate adverse effects of oral propafenone (P), 772 pts (249 with structural heart disease [SHD]) from 27 European centers treated for the following dysrhythmias were evaluated: reentrant supraventricular tachycardia (SVT) (50.3%), atrial (8.5%) and junctional (5.1%) ectopic tachycardia (JET), atrial flutter (AFL) (2.7%), premature ventricular complexes (PVCs) (18.1%), ventricular tachycardia (VT) (10.1%) and other arrhythmias (5.2%). Benign systemic side effects occurred in 36 pts (4.8%). Significant proarrhythmia was found in 12 pts (1.6%) with P alone or in combination with digoxin (6/12)—sinus node dysfunction in four, complete AV block in one, more SVT in two, acceleration of ventricular rate during AFL in one, aggravation of ventricular arrhythmia in three pts without SHD (dose: 200-290 mg/m²/d) and new development of VT in one pt with JET and SHD (dose 446 mg/m²/d). Cardiac arrest or sudden death was encountered in five pts (0.6%) (digoxin in three) at doses between 187-375 mg/m²/d. 2/5 had SVT due to WPW syndrome and a normal heart. 3/5 had SHD and were treated for PVCs (2) or SVT (1). In conclusion, P is associated with a low, but significant risk of proarrhythmia, cardiac arrest and sudden death in pediatric population, both in the presence and absence of SHD.

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Complete atrioventricular block—prenatal diagnosis, management and long-term follow-up

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Complete AV-block (CAVB) requires specific attention prenatally and postnatally. We studied a group of 40 patients (pts) with fetal CAVB from onset up to 15 yr of age. Diagnosis was established between 14-39 weeks gestational (median 24). Average heart rate at detection was 58 BPM (range 45-72). Isolated CAVB occurred in 28 pts (anti SS-A positive), 12 had associated structural heart disease (ASHD). Fetal hydrops (FH) was present in 13 pts (five isolated CAVB). Four pts died intra-uterine, three in the neonatal period; all had ASHD and FH. Only one fetus with isolated CAVB died intrauterine, having FH. Six pts were diagnosed while still having second degree heart block (four isolated CAVB, two ASHD). Median gestational age at progression to CAVB was 28 weeks (range 20-39). 17/35 surviving pts required pacemaker implantation, thirteen pts with isolated CAVB (48%) and four with ASHD (50% of surviving pts). Median age at implantation was 2.3 yr (range 1 day-11 y). The median age at follow-up is 7.5 yr. All pts with isolated CAVB are in NYHA Class I. In conclusion, perinatal mortality of fetal AVB occurs mainly in the FH-group, with ASHD. Isolated CAVB has an excellent prognosis, though a significant number of pts require pacemakers. Prenatally, a period exists in which second degree develops into complete AV-block, offering a window for possible anti-inflammatory therapy.

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Twenty-four hour ambulatory blood pressure monitoring in patients after coarctation repair

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Despite early correction of coarctation of the aorta (CoA), systolic and/or diastolic hypertension at rest or dynamic exercise may persist or develop years after surgery. Sixty-five asymptomatic patients (39 male, 26 female) were studied 4-24 years after CoA repair by 24-hour ambulatory blood pressure monitoring (ABPM). At rest all patients were normotensive and had a systolic arm-leg gradient <20 mm Hg. Data of patients after neonatal (COA 1: <28 days, n=18) and postneonatal (CoA 2: >28 days, n=47) surgery were compared to 18 normotensive controls (C). Hypertension was defined as mean systolic or diastolic blood pressure values above the 95 percentile for age and gender. Left ventricular dimensions and function were assessed by standard echocardiography. Left ventricular muscle mass index (LMMI) was calculated according to the 'Devereux' formula. There was no significant difference between CoA 1 and CoA 2 in respect to LMMI or to systolic or diastolic blood pressure. 23% of the patients had systolic and/or diastolic hypertension (CoA 1: n=4, CoA 2: n=11). Mean systolic blood pressure was elevated (131 vs 121 mm Hg, $p<0.05$) and LMMI was significantly increased (90 vs 64 g/m^2, $p<0.05$) in CoA when compared to C. There was a weak but significant correlation between mean systolic blood pressure and LMMI ($r=0.33$, $p<0.01$). Despite normal resting blood pressure and a good operative result after CoA repair, ABPM revealed hypertension in 23% of the patients. Thus, ABPM is recommended after CoA repair. If there is accompanying LV hypertrophy, antihypertensive treatment should be considered to reduce the increased risk of cardiovascular morbidity and mortality.

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Anatomical characteristics of arterial ducts with regard to successful transcatheter occlusion

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To investigate possible determinants for successful closure, we reviewed the data of 128 children in whom a Rashkind 'double umbrella' was implanted for an arterial duct and who had an echocardiogram at least three months later. From the lateral aortogram we calculated the ampulla index (diameter of the ampulla at the aortic orifice of the duct/ampullar length) and the ductus index (smallest diameter/length of the narrow segment). In 101 children (80%), total closure was achieved, 26 children had either a residual shunt (in 10 children trivial or small, in 10 children large), or surgery because of embolization (3) or hemolysis (3). Comparison of children with and without successful closure showed no differences in sex, age and weight at time of implantation or in umbrella size. However, the ampulla index was greater (2.9±1.5 (SD) vs 1.7±0.7, $p<0.005$) and the ductus index was smaller (1.5±1.1 vs 2.0±1.6, $p<0.05$) when closure had been successful. When the ampulla index was <1.5 only 45% of the children had successful closure. We conclude that the anatomy of ampulla and narrow segment may predict successful closure. This may have implications for the policy of treatment.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

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Radiofrequency balloon dilation of the arterial duct can maintain medium-term arterial duct patency

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Stent implantation is superior to balloon dilation in maintaining patency of the arterial duct in the long-term. The potential hazards are stent embolization or thrombosis. Laser-balloon dilation whilst effective in coronary arteries in the short-term, when used at temperatures up to 100°C, resulted in excessive neo-intima formation. Radiofrequency balloons of appropriate size are available and by denaturing the muscular layer of the arterial wall may be more effective than simple balloon dilation. We investigated the effects of radiofrequency balloon dilation (RFBD) on the arterial duct in 12 lambs aged 2-9 days. The closed duct was successfully recanalized in 11. Radiofrequency balloons (Boston Scientific Corporation) were advanced over a guidewire across the duct via 7 F sheaths. The balloons were of two diameters (8 mm in seven lambs and 6 mm in four lambs) and were inflated to only two atmospheres during energy delivery to avoid excessive intimal tearing. Radiofrequency energy was delivered to produce constant temperatures in the balloon of either 60 °C or 80 °C for periods of 15 to 60 sec repeated up to six times. Immediately after RFBD the duct was closed in one of four lambs in whom the 6 mm balloon had been used but in none of the seven in whom 8 mm balloons had been used. Subsequently, the duct was critically narrowed within two months in the remaining three lambs in whom the 6 mm balloons had been used. The duct was patent in all those in whom 8 mm balloons were used. In four of these, autopsies were performed immediately after, one d, one month and two months after RFBD. The remaining three had widely patent ducts at repeat angiography 3-4 months later. RFBD can maintain arterial duct patency at least in the medium-term. Larger balloons are more effective. RFBD has a potential role in neonates with duct dependent congenital heart disease. The major limitation is the need for a 7 F introducer sheath.

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Radiofrequency perforation of membranous pulmonary atresia with intact ventricular septum—a new way of treatment

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Newborn infants with pulmonary atresia and intact ventricular septum usually present critically ill and need immediate treatment by surgery with high risk. We tried to avoid surgical management in four neonates by catheter intervention. The age was between two days (two patients) and four weeks (two patients). The oxygen saturation was below 65% in all. The perfusion of the lungs was PDA dependent. Angiograms showed a small RV with good sized infundibulum and presence of a main pulmonary artery in all patients. A 5 F open-end catheter was placed in the RV outflow tract close to the membranous atresia and a small 2 F wire catheter (Fa. Osypka, Grenzach) brought into contact with the membrane. After 4-10 radiofrequency applications (25-30 W) the end of the catheter passed into the pulmonary artery and was advanced into the descending aorta via the PDA. After exchange by a 14" guidewire balloon catheters with increasing balloon size (2-4-6-8 mm) were introduced and the perforated membrane dilated. The procedure was successful in all four patients. One perforation into the pericardium without complication occurred. The oxygen saturation increased to 85%. The RV-pressure was markedly reduced (over systemic to half-systemic level). In two patients, the size of the RV was too small to support normal lung perfusion. In one patient, a modified Blalock-Taussig shunt had to be performed. In the last patient, implantation of a Palmaz-Schatz stent into the PDA was successful. All patients are at home and well. We think radiofrequency perforation is a promising method of treatment for these patients.

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Balloon dilatation of coarctation of the aorta in childhood—medium-term results

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Concerns regarding the efficacy and safety of balloon dilatation (BD) of native coarctation of the aorta (NCoA) in childhood have resulted in its limited acceptance. We present the results of balloon dilatation in 42 sequential cases of NCoA and in 32 subjects ballooned between Jan 1985-Aug 1993 for recoarctation following previous surgical repair (SCoA).

Median (range or percent)	NCoA (n= 42)	SCoA (n = 32)	p
Age at balloon (yrs)	14.5 (1-32)	1.8 (0.3-34)	0.001
CoA gradient (mm Hg)	26 (10-84)	26.5 (14-80)	0.30
Length of follow-up	5.4 (0.2-7.7)	5.8 (0.3-8.8)	0.80
Post-balloon gradient (mm Hg)	4.0 (0-25)	10 (0-36)	0.001
Complications	6 (14%)	6 (19%)	NS
Repeat BD	6 (14%)	6 (19%)	NS
Re-operation	1 (2%)	2 (6%)	NS

Three patients died from associated cardiac lesions; no deaths were attributable to the procedure. There were 12 complications—six aneurysms, two in SCoA and four NCoA that have not progressed on MRI surveillance, four local vascular problems (with sequel in one case), and two subjects with transient neurological signs. One subject from each group had surgical repair after unsuccessful BD, while six NCoA and five of six SCoA had satisfactory results from repeat BD. Balloon dilatation of both NCoA and SCoA in childhood is a safe and acceptable method of treatment providing satisfactory and sustained relief.

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Balloon angioplasty of native coarctation

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From 3/88 to 9/93, 27 patients with native coarctation of the aorta (Coa) underwent angioplasty. Age at intervention was seven to 300 months (mean 98±68). Indications were systemic hypertension and/or a pressure gradient of more than 25 mm Hg. Decrease of the gradient and increase of Coa-diameter were used as parameters of success. Single balloons with diameters of 7-18 mm were used; the ratio of balloon-to-Coa diameter was 2.6±1.0. The pressure gradient decreased from 46±12 to 11±9 mm Hg (decrease of 77±18%, $p<0.0001$). Coa-diameters increased from 5±2 to 10±4 mm (increase of 129±81%, $p< 0.0001$). Four patients needed thrombolysis for femoral artery thrombosis and two developed paradoxical hypertension. Seven patients showed discrete to distinct aortic wall anomalies at the Coa site on post-angioplasty angios. Nuclear magnetic resonance (NMR) investigations were performed in 23 patients after angioplasty. In five of seven patients with angiographic wall anomalies, NMR allowed further differentiation and showed intimal flaps (2x), localized wall dissection (2x) and one small aneurysm. In the remaining 16 patients, NMR only once showed a small area of wall dissection. On repeat NMR after six to nine months, all irregularities had disappeared. Balloon angioplasty of native Coa can be performed with excellent results.Early angiographic irregularities of the aortic wall, however, may indicate localized wall dissection and should be studied by NMR immediately. Though these anomalies seem to heal spontaneously, careful and frequent follow-up by NMR is necessary. NMR is a sensitive method to investigate unclear aortic wall anomalies after angioplasty.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

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Ultrasound guidance of endomyocardial biopsy via the internal jugular vein in infants and children

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Use of right ventricular endomyocardial biopsy (EMB) in children has increased markedly over the last few years. EMB has become essential in the follow-up of children after cardiac transplantation. EMB also is used in the diagnosis and management of myocarditis, doxorubicin cardiomyopathy, AIDS myopathy and complex ventricular arrhythmias. Ultrasound (US) guidance of EMB from the right internal jugular (RIJ) vein has gained wide acceptance in adults, yet the standard pediatric approach remains via the femoral vein under fluoroscopic guidance. We present our experience with EMB from the RIJ using only US visualization. Since December 1990, we have performed 64 EMB on 11 children, ages 1.4-17 yrs (mean 9.4±5.5), weighing 8-70 kg. 15/84 biopsies were performed on children two years or less. Most EMBs were performed for transplant follow-up, but other indications included congestive and restrictive cardiomyopathy, acquired complete A-V block, and symptomatic ventricular tachycardia. Mean sheath insertion time was 3.6±2.0 min. The average time (after sheath insertion) needed to obtain four or five myocardial samples was 15.5±8.5 min. No complications were encountered. We compared these data to nine biopsy procedures performed under fluoroscopy on three patients between 5/90 and 12/90. The average procedure times were comparable, but fluoroscopy time averaged 6.6±2.7 min (range 4-12). We conclude EMB from the RIJ can be performed safely in children. Ultrasound guidance allows precise localization of biopsy site, can be performed at the bedside in critically ill patients and is radiation-free. Since immunosuppressed children are at increased risk of developing lymphoproliferative disease, avoidance of repeated radiation exposure is desirable. Thus, especially for cardiac transplant patients, US should be the imaging modality of choice for EMB guidance.

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Comparison by computer modeling of energy losses in different Fontan connections

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The surgeon disposes of different options when creating a Fontan circulation. The choice of including an atrium into the circuit, or the way of connecting the caval veins into the pulmonary artery is usually empirical and based upon personal experience. The differences between the circuits are small and short-term results are comparable. However, even small differences in the connections in the low pressure systemic venous system may have significant implications during long-term follow-up. The finite element method allows to simulate on a computer the flow dynamics and pressure losses of different connections in a 'single patient' with identical geometry and functional conditions. We compared the atriopulmonary connection with various cavopulmonary connections. The cavopulmonary connections differed by the degree of offset of the implantation of the caval veins into the right pulmonary artery. On the basis of anatomic models and physiologic flow dynamics, three-dimensional geometry's and finite element meshes were created with PATRAN® (USA); flows were calculated with POLYFLOW® (B), and results were visualized with DATA VISUALIZER® (USA). We conclude that the atriopulmonary connection produces higher energy losses than the cavopulmonary connection (±1 mm Hg at rest). The cavopulmonary connection is more efficient when the implantation of the caval veins into the pulmonary artery is completely offset. Energy losses in each circuit are minimized by rounding and streamlining corners as much as possible.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

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Correlation of left ventricular mass index with resting, exercise and 24-hour blood pressure recording in young adults following repair of coarctation

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The aim of the study was to assess the value of single clinic systolic and diastolic blood pressure (CSBP, CDBP), peak exercise blood pressure (XSBP), mean day (0700-2400) and night (2400-0700) systolic and diastolic pressures (SDBP, DDBP, NSBP, NDBP) in predicting left ventricular hypertrophy (LVH) in adolescent and young adult patients following surgical repair of coarctation. Twenty-two pts, median age 19 (13-36), 17 male, four female, 14.5 years post CoA repair and without significant other cardiac pathology, had resting arm and leg BP recording, treadmill exercise test (Bruce protocol), ambulatory 24-hour BP measurement (Spacelabs 90207, recording every 30 minutes) and ultrasound (Toshiba SSH 140A) assessment of indexed left ventricular mass (LVMI). The results were correlated using Pearson's coefficient. Mean LVMI was 96±5 g/m^2 and nine pts had LVH (LVMI >95th percentile for age and gender). The strongest correlation was with XSBP (0.68, p>0.001) then DSBP (0.56, p>0.007). XSBP>225 mm Hg and DSBP >142 mm Hg both identified 7/9 pts with LVH and the false positive rate was 1/13 and 2/13, respectively. All others had insignificant correlation coefficients, -0.19 to 0.39. XSBP and DSBP are the best predictors of LVH in pts after CoA repair and should be measured routinely. In the future, advice regarding activity level and drug management may need to be altered if these values are abnormal.

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Prostaglandin D_2 (PGD_2), eosinophil-cationic protein (ECP) and histamine release in children undergoing cardiopulmonary bypass (CPB)—relation to postoperative dysrhythmias

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To study leucocyte and mastcell degranulation during CPB and the influence of the release of histamine on postoperative dysrhythmias occurring during the first three days, we investigated 30 pts (mean age 60 months) undergoing open-heart surgery for congenital cardiac defects. Histamine was used as marker for both mastcell and basophil degranulation, PGD_2 for mastcell degranulation alone and ECP for eosinophil degranulation. Mediators of the inflammation reaction were studied before, during and after CPB. In all children, PGD_2 release occurred during CPB but ceased immediately after termination of CPB. In contrast, ECP and histamine levels increased during CPB and up to four hours thereafter and normalized slowly within 24 hrs after the operation. In order to discriminate patients with overtly pathologic histamine release during or immediately after CPB, measured values >2 SD of the preoperative mean value calculated for all patients were considered abnormal. Seven pts showed abnormal histamine liberation. Eight pts developed postoperative dysrhythmias (slow nodal rhythm, n=4; junctional ectopic tachycardia, n=3; transitory complete A-V block, n=1). Postoperative dysrhythmias correlated with pathological histamine release (p<0.01). Mast cell degranulation seems to occur only during CPB as seen by PGD_2 release, while eosinophil and basophil degranulation induce a sustained release of ECP and histamine for up to 24 hrs after CPB. Our data suggest a relationship between histamine release during and/or after CPB and postoperative dysrhythmias.

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Lessons learned in the management of double inlet left ventricle with transposition (DILV/TGA) and aortic arch obstruction

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It is stated that pulmonary artery banding (PAB) is contraindicated in the setting of DILV/TGA, especially if coarctation (CoA) is present. The alternative form of primary palliation ('Norwood' approach) is associated with high mortality and morbidity. Therefore short-term PAB remains an attractive option. We have reviewed our results using this approach. Since 1980, 16 infants have presented in CCF with DILV/TGA. Four had no CoA and are all alive and well having achieved Fontan (two pts) or bidirectional Glenn (BDG) with arterial switch (two pts). Of the remaining 12 pts, 11 had CoA and one IAA. 4/12 were considered to have subaortic obstruction (SAS) at presentation. One underwent neonatal aortopulmonary connection and died. The remaining 11 pts underwent arch repair with PAB (median age 1.7 wks, 2 d-22 wks) with one early death. The 10 survivors have been followed for 7.1±3.2 years. Of the eight considered free of SAS at presentation, seven acquired it after PAB. SAS was recognized in these seven pts at 13.7 months (4-46) after PAB. LV-aortic gradient at catheter was 57±21 mm Hg (35-90). Relief of SAS was performed at median age of 16.5 months (5-52). All had proximal aortopulmonary connection (AP window 6, conduit 2, DKS 1), with PAB left *in situ* (4), central shunt (3), BDG (1), or Fontan (1). Only the latter pt died and there was one late death (non-cardiac). Thus, 8/12 pts with arch obstruction are alive and all have persistent relief of SAS. Six have achieved Fontan repair and two have BDG with no contraindication to subsequent Fontan. All have PA pressure <18 mm Hg at follow-up catheterization. Most patients with DILV/TGA and arch obstruction will tolerate temporary PAB with adequate protection of the pulmonary artery bed for future Fontan. Clinical echo assessment may miss early SAS. Thus, cardiac catheterization should be performed soon after PAB. Our current approach is PAB followed by early infant relief of SAS and a BDG shunt.

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Isomerism of the atrial appendages in the fetus

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Since 1982, 95 fetuses with isomerism of the atrial appendages have been seen in our center, 64 with left (LAI) and 31 with right isomerism (RAI). These fetuses represent 9% of the structural cardiac abnormalities in our prenatal series. The diagnosis of atrial isomerism was correctly predicted in 75% by fetal echocardiography. In the remaining fetuses, a complex cardiac abnormality was detected but the association with atrial isomerism was overlooked. The recurrence rate in subsequent pregnancies was 10% and a high rate of consanguinity (11%) was present in our series. One fetus with RAI had a chromosomal abnormality. Conotruncal abnormalities of varying severity were noted in 12% of fetuses, being equally common in RAI and LAI. The most frequent intracardiac abnormality was an atrioventricular septal defect which was present in 76%. This was associated with complete heart block in 52% of fetuses with LAI. In fetuses with RAI, the great arteries were normally related in 26%, transposed in 4% and a double outlet arrangement was present in 66%. The pulmonary outflow tract was obstructed at valvar or subvalvar level in 35% and was atretic in 22%. In LAI, the great arteries were normally related in 44%, transposed in 14% and a double outlet arrangement was present in 27%. Aortic stenosis at valvar or subvalvar level was noted in 13% and pulmonary outflow was stenotic in 19% and atretic in 9%. Of 24 continuing pregnancies (26%), there were three intrauterine deaths, 10 babies died in the neonatal period and three deaths occurred in childhood. There are currently eight survivors with follow-up extending to seven years (mean 14 months). Isomerism of the atrial appendages can be diagnosed in the fetus, has a high recurrence risk and may rarely be associated with a karyotype abnormality. It is still associated with poor long-term outlook.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

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Cross-sectional delineation of coronary arterial origins in complete transposition

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Coronary arterial anatomy in complete transposition of the great arteries was prospectively echocardiographically investigated in 29 consecutive neonates together with the spatial great arteries relationship and the presence or absence of a ventricular septal defect (VSD). In 6/29 pts (21%), less favorable coronary patterns for the arterial switch operation were found (2/29 a single right, 1/29 a single left, 1/29 inverted coronaries, 1/29 inversion between the right and the left circumflex artery, 1/29 'Yacoub type C' anatomy). VSD was present in 8/29 pts, but only 4/8 (50%) had coronary artery anatomy other than the commonest one. In 5/29 patients (17%), a side-by-side great arteries relationship was found, and all of them had the more risky coronary patterns. It is a side-by-side great arteries relationship due to either bilaterally present or absent infundibulum, but not a VSD which is related to less favorable coronary anatomy in complete transposition.

Notes:

P 01

Arterial duct patency at 18 months after radiofrequency thermal balloon angioplasty in neonatal lambs

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A technique for producing long-term patency of the arterial duct (AD) may replace palliative surgery in some neonates with duct-dependent circulations. We have previously reported that radiofrequency thermal balloon angioplasty (RFTBA) to the AD of 28 neonatal lambs produces short-term patency in over 80% of treated ADs. All ADs that closed did so before three months. Nine lambs were entered into a long-term study. In all cases, RFTBA had been performed within the first five days of life. Following diagnostic catheterization, the AD was crossed with an 0.035" exchange guidewire from the venous side. A 6 mm (n=5) or 5 mm (n=4) radiofrequency balloon was passed across the AD and inflated. Radiofrequency heated the balloons to temperatures of 65 (n=1), 75 (n=1), 85 (n=3), 100 (n=3), 120 °C (n=1) for 15 seconds. Recatheterization with angiography at a mean 17±1.7 months with a mean weight of 43±11 kg showed that all ADs remain patent and their mean narrowest diameter to be 2.96±0.96 mm. Mean systolic pulmonary artery pressure was 26±6 mm Hg and mean aortic-to-pulmonary artery systolic pressure ratio was 4±0.25. Mean step-up in oxygen saturation from the superior vena cava to the pulmonary artery was 7.9±5%. We conclude that once patency is established in the AD with RFTBA, the vessel remains patent for at least 18 months, the normal closure mechanism having been effectively inactivated.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

P 02

The concept of transcatheter application and retrieval of implantable devices

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Transcatheter introduced and retrieved (TIR) devices, should serve their hemodynamic intended purpose for a predetermined period of time and should be removed without residual problems. TIR devices were applied experimentally as follows: (1) Pulmonary artery bands (PABs)—two generations of TIR PABs were applied in 20 piglets and were retrieved in periods ranging from one day to three months. TIR PABs were effective in causing an average gradient of 20 mm Hg and they were retrieved with minimal intimal changes and no residual gradients. Initial thrombogenicity improved with the second generation bands. (2) Arterial stents—they were positioned in the aortic arch and the descending aorta in 10 piglets and were removed one day to two months after implantation without significant intimal changes or thrombogenicity. (3) Valve cages and strands—they could be easily positioned and retrieved in the aortic, pulmonic, tricuspid and mitral positions in three acute experiments. TIR valves were constructed and tested *in vitro* on the above types of cages and strands. TIR devices are feasible and potentially applicable to a variety of conditions where a temporary result is desirable.

P 03

Malformations of the pulmonary artery evaluated by magnetic resonance imaging

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Surgical treatment of patients with pulmonary atresia, central or peripheral pulmonary stenoses requires accurate definition of the pulmonary vascular status. Magnetic resonance images of 71 patients (five PA with IVS, 39 PA with VSD, five PA with TA, six PA with SV, five aplasia of left pulmonary artery, 10 central pulmonary stenosis) were compared with echocardiographic (71) and angiocardiographic (65) findings. Forty patients had undergone palliative surgery with placement of 50 systemic-to-pulmonary shunts. Cardiac ECG-gated MRI studies were performed using a multislice spin echo technique in all and additional 'white blood' imaging techniques in 62. The morphology and size of the pulmonary arteries could be accurately assessed in all patients, in contrast to echocardiography, which failed in 53 patients. MRI discovered unknown hypoplastic pulmonary arteries in 12 patients, which was confirmed either by pulmonary vein wedge angiography or surgery. In contrast to MRI, angiography was not able to demonstrate left or right pulmonary artery in 20 patients with severe central stenoses of the pulmonary artery caused by palliative shunts (11), dislocation of a banding (2), or aneurysm of the main pulmonary artery (7). Better definition of pulmonary blood supply by aortopulmonary collaterals was achieved in five patients. Forty-seven palliative shunts were visualized and could be evaluated for patency in 30 patients, as stenotic in 12 and occluded in four patients. We conclude that MRI is an excellent noninvasive technique for diagnosing pulmonary artery malformations and should be carried out prior to heart catheterization.

P 04

Left ventricular mass determination by echocardiography may yield additional information in patients with aortic stenosis

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Severe LV hypertrophy may lead to subendocardial ischemia. We examined whether assessment of LV mass provides information additional to gradient measurement in deciding when to treat aortic stenosis (AS). LV mass and volume were calculated by two-dimensional echocardiography in 57 aortic stenosis patients with a mean age of 7.8 (0.1-19.8) years, among 13 infants. Newborns with dilated LV and poor function with shortening fraction under 30% and patients with Doppler gradient <30 mm Hg were excluded. Epicardial and endocardial volume was calculated in two perpendicular imaging planes—apical two- and four-chamber views using a Simpson's rule algorithm. Difference between epicardial and endocardial volume (i.e. myocardial volume) was multiplied by 1.05 to obtain LV mass. Mass/volume ratio was calculated at end-diastole. Data were compared to 95 age-matched controls. Mean Doppler gradient across aortic valve was 59 (35-113) mm Hg. Twenty-seven patients had trivial aortic incompetence (AI), 23 grade 1 and seven grade 2 AI. LV mass was 157% (85-337), LV volume 98% (55-175) and LV mass/volume ratio 166% (82-306) of normal. Correlation of Doppler gradient with ECG signs of LV hypertrophy was poor ($r<0.5$). Correlation between LV mass and gradient was $r=0.51$, but correlation between mass/volume ratio and gradient was $r=0.73$. These correlations also reflect the fact that patients had a wide age range. All pts with mass/volume ratio of >2.5 had signs of ischemia on resting ECG, among them two with Doppler gradients <70 mm Hg. We conclude that gradient alone should not guide decision to treat, as occasional pts with moderate Doppler gradients do have a mass/volume ratio in excess of 2.5 indicating resting subendocardial ischemia. LV mass/volume measurements should become part of evaluation of selected patients with AS.

P 05

Permanent junctional reciprocating tachycardia in children—curative approach and diagnosis

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Radiofrequency current catheter (RFC) ablation for accessory pathway (AP) mediated supraventricular tachycardia (SVT) was performed in 70 consecutive pts (15 yrs; 9.2 ±4.1 yrs). Out of these, nine (5 F; 4 M; 6.1±3.4 yrs) presented with the permanent form of junctional reciprocating tachycardia (PJRT). This arrhythmia was noticed at birth in three pts and prenatally in another three pts. All pts had severe symptoms. Depressed LV function (FS 13-24) was evidenced prior to the ablation procedure in three pts. In all pts, a concealed AP with slow and decremental conduction properties was found to be the underlying mechanism of the tachycardia. Localization of the AP was guided by the recording of a presumed AP activation potential at the site of the earliest local atrial interval during SVT. 7/9 APs were located in the posteroseptal space (PSS) (five right-, two left-sided) and 2/9 at the left free wall (both left posterolateral). Eight pts were cured within 11 sessions, requiring 13±10 RFC applications. Procedure duration was 4.8±2.6 hrs; fluoroscopy time was 47.5±38.3 mins. There were no serious complications. Within an 18 month (median) follow-up, five pts were asymptomatic and remained in sinus rhythm. In three pts, PJRT recurred after 0.5 to two months; re-ablation was successfully attempted in two, whereas no intervention was performed in the third pt, due to non-sustained, asymptomatic episodes. In children who presented with depressed LV function, RFC ablation of the AP was associated with return to normal values during follow-up. We conclude: (1) PJRT is a common cause in children referred for RFC ablation of AP-mediated SVT. (2) PJRT may result in depressed left ventricular function even in early childhood. (3) Due to the potential detrimental effects of PJRT on LV-function, RFC ablation should be attempted upon diagnosis in all cases. (4) RFC ablation of PJRT in this subset of pts is safe and efficacious.

P 06

Nonsurgical closure of persistent arterial duct in symptomatic children under two years of age

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The aim was to assess the efficacy and safety of transcatheter occlusion (TO) of persistent arterial duct (PAD) in symptomatic children under two yrs, with a special reference to the new front-loading delivery technique. Between June 1990 and September 1993, 35 symptomatic young children with a PAD underwent elective TO with a Rashkind umbrella. Their ages ranged 1.5-23 months, with the youngest infant weighing 2.9 kg. The diagnosis was established preoperatively in all patients by cross-sectional echocardiography. Symptoms ranged from bronchopulmonary dysplasia to breathlessness, failure to thrive or recurrent chest infections. TO of a hemodynamically important PAD was performed using the Rashkind ductal umbrella. In the past 15 months, the front-loading technique has been used in five cases to deploy the 12 mm umbrella via a 6 F sheath and the 17 mm device via a 8 F or 9 F sheath, so extending the indications for their use. Successful placement was achieved in 30 (85.7%) with symptomatic improvement in all. There were no deaths. Five failures were early in the series, and they were due to kinking of the 11 F sheath in two, embolization into the left pulmonary artery in one, partial protrusion of the device into the descending aorta, and from abandoning the procedure due to a large duct in the fifth case. Four pts were referred for surgery; the pt in whom the umbrella embolized underwent a successful implantation two months later. Two pts developed severe hemolytic anemia which resolved following the deployment of a second device. Only four of the 30 pts had small residual shunts at one yr follow-up (all with 17 mm devices). There was one case of left pulmonary artery stenosis. In conclusion, the transcatheter umbrella closure of the PAD in symptomatic young children is a safe alternative to surgery. Front-loading the device into a smaller sheath extends the age and size limits of the procedure and does not appear to increase the risk of complications.

P 07

Balloon dilation of stenosed systemic venous pathways following the Mustard operation

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The aim was to assess the immediate and long-term effects of percutaneous balloon dilation (BD) of stenoses in the superior (SVP) and inferior (IVP) systemic venous pathways following the Mustard procedure (M). Twenty-five pts (17 M, eight F) underwent 43 BDs (18 SVP, 25 IVP). The median age at M was one (range 0.3-2.9) yrs, and at first BD 6.4 (2-18) yrs. The diameter of the stenosis was averaged from the maximum and minimum diameter of the stenotic segment during one cardiac cycle. Gradients were estimated from the mean pressures in the superior and inferior vena cava and the systemic venous atrium. For 13 initial SVP dilations (median balloon to stenosis (B:S) ratio 3.25), the gradient across the stenosis decreased from 8 (3-21) to 4 (0-7) mm Hg ($p<0.01$). The diameter of the stenosis increased from 4.7 (2.7-8.6) to 11.1 (7.3-17.5) mm ($p<0.01$). For 22 initial IVP dilations (B:S of 2.5), the gradient decreased from 3 (3-10) to 1 (0-5) mm Hg ($p<0.01$). The stenosis diameter increased from 6.1 (4.1-9) to 10.1 (5.7-18) mm ($p<0.01$). Over a median follow-up of 3.5 (0.3-9.1) yrs, patients were evaluated by serial two-dimensional Doppler echocardiography (n=25), and angiography (n=9). Five pts (20%) have undergone eight further BD procedures (five SVP, three IVP). The percentage reduction in gradient (59 vs 69% for SVP and 67 vs 75% for IVP), and increase in stenosis diameter (158 vs 130% for SVP and 58 vs 20% for IVP) were not different between first or subsequent BD procedures ($p>0.05$). Finally, one pt has undergone baffle revision for severe recurrent SVP obstruction, despite an acute decrease in gradient from 15 to 6 mm Hg at initial BD. There were no complications associated with any procedure. Systemic venous pathway stenoses following M respond well to transcatheter BD, with excellent long-term results. Multiple procedures are required in some patients, but the results of subsequent BDs are comparable with those at initial BD.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

P 08

Medium-term results of stent implantation into the arterial duct in cyanotic neonates

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We have attempted stent implantation into the arterial duct in nine neonates with cyanotic heart disease, using a Tower single strand retrievable stent. Pulmonary atresia was present in five, severe subpulmonary stenosis combined with either TGA, DILV or right isomerism and AVSD in three and critical pulmonary stenosis with a hypoplastic right ventricle in one. In two, balloon dilation (BD) of the pulmonary valve had been performed (one after percutaneous radiofrequency valvotomy) but they remained duct-dependent. Under general anesthesia, attempts to place a guidewire across the duct were made using the femoral or axillary arterial approach in seven or the femoral venous approach in the two who had undergone BD of the pulmonary valve. It was not possible to cross the tortuous duct in four pts; a modified Blalock-Taussig shunt was performed in three of them and BD of the pulmonary outflow tract in one. Stent implantation was performed in five using 4-6 mm diameter Tower stents. Two stents were initially malpositioned, were snared, and removed and replaced. Prostaglandins were stopped after the procedure except in one pt who died suddenly at 24 hours in ventricular fibrillation. One pt, in whom the aortic orifice of the duct had not been stented, had further stents at a second procedure. Aspirin and warfarin were continued after the procedure in all. The two pts who had BD of the pulmonary valve are no longer duct-dependent. In one with warfarin non-compliance, the stent occluded uneventfully between five and nine months. In the other, warfarin was stopped at six months and the stent is patent at 15 months. The duct-dependent pts are palliated at four months. Stent implantation into the arterial duct is a useful alternative to neonatal systemic-to-pulmonary artery shunt operations.

P 09

Transeptal versus retrograde transaortic approach in radiofrequency ablation of left accessory pathways in children

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In Wolff-Parkinson-White syndrome, radiofrequency ablation of accessory pathways is considered an efficient type of treatment. In this study we compared the retrograde transaortic approach (RTAo) and the transeptal (TS) approach in the ablation of the left accessory pathways in nineteen pts (16 M, 3 F, mean age 17 yrs, range 13-20) with left accessory pathways, 10 left lateral (LL) and two posterolateral (PL). All patients had supraventricular arrhythmias. Thirteen patients underwent a TS approach, six a RTAo approach. During the TS procedure, the K potential of the accessory pathways was registered in all patients. The success of the ablation is demonstrated by the disappearance of the ventricular preexcitation on the surface ECG, the K potential on the endocavitary electrogram and by the abolition of both anterograde and retrograde conduction. The percentage of success (follow-up range 1-18 months) was 100% both for the TS approach and the RTAo approach, even though the TS approach needed two attempts of ablation in two cases (50%). No complications were observed. The mean fluoroscopy time in RTAo was superior to the one by TS 116% (71±12 mins, vs 32±20 min; $p<0.01$). In conclusion, our results demonstrate that the TS procedure can be considered the technique of choice in young patients with left accessory pathways.

P 10

Stent implantation is able to maintain long-term arterial duct patency

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Balloon dilation is disappointing in maintaining arterial duct patency. In neonatal lambs, stent implantation into the arterial duct results in significantly larger ducts with greater pulmonary blood flow than balloon dilation. Little is known, however, about the duration of duct patency after stent implantation. We have evaluated the outcome of stent implantation into the arterial duct in 12 lambs observed for 1-24 months. Stents (Wall stent in nine, Tower stent in three) were implanted after recanalizing the occluded duct at 2-7 days of age. Heparin was given during the procedure but no further anticoagulants or antiplatelet agents were used. Angiographic and postmortem evaluations were made at 1, 1.5, 2, 3, 4, 6, 12, 16 and 21 months in a subgroup of nine lambs who did not undergo reintervention. Neo-endothelium formation developed initially in the center of the stent and then extended towards the orifices. From three months onwards this was extensive, though even at 21 months, parts of the stent not firmly applied to the wall or protruding beyond the duct were not covered by neo-intima. From 4-6 months onwards, stenoses were apparent inside the stent. When the stent did not protrude into the aorta, neointima extended over the duct orifice. The duct was patent in all except one studied at 16 months in whom the aortic orifice was covered. Balloon dilation of the stented duct, performed in two lambs, at three and five months in one and at 15 months in the other, resulted in an improved angiographic appearance and increased pulmonary artery blood flow. In one lamb, the neointimal lining was successfully removed using an atherectomy catheter. Long-term arterial duct patency is possible, but the implanted stents should span the entire duct length. Neo-intima formation may eventually reduce flow which can be improved with balloon dilation. Anticoagulation and/or antiplatelet agents may have a role in maintaining long-term stent patency.

P 11

Immunohistological proven myocarditis—therapy and short-term follow-up

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The purpose of the study was to assess the influence of steroid therapy on immunohistologically proven myocarditis. In our department between 1991 and 1993, endomyocardial biopsies were performed in 21 pts (mean age 4.7 yrs, range 0.1-12) with suspected myocardial disease. Biopsies were examined after hematoxylin-eosin (HE) staining and incubation with various marked antibodies against specific lymphocytic antigens (CD2, CD3, CD4, CD8, HLA-expression). According to the Dallas criteria, HE-staining revealed borderline myocarditis in two pts; immunohistological technique, however, showed severe lymphocytic infiltration in 13 pts (CD2/CD3 positive in all, HLA-expression positive in 12 pts). In these pts, clinical manifestations were cardiomegaly with systolic (5) or diastolic (1) dysfunction (NYHA 2-3), 2°-3° AV-conduction disturbance (4), severe pericardial effusion (1), ventricular tachycardia (1) and changes of repolarization (1). Serological markers detected mycoplasma pneumonia only in one patient. For clinical reasons and immunohistological findings, prednisone (1-2 mg/kg/d) was given to six pts, additional diuretics (4), digoxin and vasodilatators (1). 5/6 treated patients (duration 2-12 months) showed clinical improvement and disappearance or reduction of infiltrations in rebiopsies. In contrast, 6/7 pts without therapy showed no clinical changes and immunohistological proven persistence of the infiltrations. One pt from each group needed permanent pacemaker implantation. Two pts died, one with severe diastolic dysfunction (prednisone therapy 10 days), a second pt (ventilated, NYHA 4) before the histological diagnosis was proven. In conclusion, immunohistological techniques are more sensitive in diagnosis of myocarditis than conventional methods. Steroids seem to have a supportive effect on various severe manifestations of viral myocarditis.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

P 12

Identification of patients at risk for ventricular tachycardia after surgical correction of congenital heart defects—role of late potentials

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This study evaluates the role of late potentials (LP) in identifying pts with sustained ventricular tachycardia (VT) after surgery for congenital heart defects (CHD) using right ventriculotomy. Nine pts with VT (spontaneous 7; induced 2) and 104 pts without VT (complete right bundle branch block in all, mean postoperative follow-up 86±47 (12-89) months) were compared. LP were quantified by two time domain parameters—RMS in the last 40 ms and LAS<20 μV of the terminal filtered (80-250 Hz) QRS complex. RMS was lower in pts with VT (9±6 vs 16±8 μV, $p<0.01$). By multivariate analysis, more complex surgery (extracardiac conduits or Kawashima operation) and lower RMS were the only independent predictors of VT ($p<0.001$ and <0.05, respectively) among several clinical variables (CHD type, age at surgery, aortic cross-clamp time, length of follow-up, postoperative hemodynamics, grade of ventricular ectopy on Holter and LAS). RMS<14 μV (+ presence of more complex surgery) was 89% (67%) sensitive and 60% (95%) specific (positive predictive accuracy 17% (55%), negative predictive accuracy 98% (97%)) for identification of pts with/without VT. In conclusion, LP are independent predictors of VT and may be helpful in identifying pts at risk of VT after surgery for CHD using right ventriculotomy.

Notes:

P 13

Effect of malalignment of left ventricular outflow tract (LVOT) structures in patients with subaortic stenosis (SAS) and a ventricular septal defect (VSD)

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Sixty-five pts with SAS and VSD were studied to determine the incidence and effect of malalignment. Thirty-two had SAS without malalignment and 33 had SAS with malalignment. Seventeen pts with malalignment (51%) had posterior deviation of structures into the LVOT, with SAS above the VSD. Sixteen pts (49%) had anterior deviation of the outlet septum and aortic override (without right ventricular outflow obstruction) with SAS below the VSD. The age at presentation was younger ($p<0.01$) in pts with malalignment than in those without. Aortic arch obstruction was more common ($p<0.002$) in pts with malalignment (76%), than in those without (38%). Median duration of follow-up was 6.6 yrs (range 1-25.7). Sixteen pts (49%) with malalignment and 16 without malalignment (50%) underwent operation for SAS. The age at surgery was younger ($p<0.005$) in patients with malalignment. Patients with posterior malalignment underwent operation for SAS more often ($p<0.05$) than those with anterior malalignment. The VSD required surgical closure more often ($p<0.005$) in patients with malalignment (93.9%) than in those without malalignment (21.9%). There was no difference in the mortality between patients with (27%) and without (12%) malalignment. In conclusion, 51% of patients with SAS and a VSD had malalignment of muscular structures in the LVOT. They underwent surgery at a younger age than those without malalignment. Two types of malalignment occurred, one causing obstruction above and one causing obstruction below the VSD. The presence and type of malalignment should be defined in all patients with a VSD.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

P 14

Right ventricular function—assessment with echocardiography and correlation with magnetic resonance imaging

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Several different geometrical models can be used for 2D echocardiographic (2DE) RV volume measurements. Multisection magnetic resonance imaging (MRI) provides RV volume assessment without geometrical assumptions. We assessed the correlation between 2DE-derived RV volumes and ejection fraction (EF), using different models and MRI derived data. Thirty-three randomly selected children were studied prospectively (15 congenital heart disease, 18 normal heart) (age 10±3 years). Multisection transverse gradient echo MRI with 8 mm section thickness was used. Transthoracic echocardiographic images were obtained from three different views (apical four-chamber (AP4), parasternal short-axis (PSX), subcostal 'RV outflow' (SC)). Calculation of RV, ED and ES volumes and EF was performed with previously reported mono- and biplane area/length methods. Linear regression analysis was used to compare results. Adequate MRI images could be obtained in all patients. Correlation between MRI and 2DE measurements:

		Method (n)		
RV function		AP4(33)	AP4/PSX(28)	AP4/SC(17)
EDV	r	0.71	0.61	0.71
ESV	r	0.80	0.87	0.71
EF	r	0.58	0.08	0.61

In conclusion, 2D echocardiographic mono- or biplane area/length RV volume estimates have a fair to good correlation with MRI derived data in different types of RV morphology. The observed correlation with MRI derived EF suggests that 2DE monoplane (AP4) or biplane (AP4/SC) methods can be used as a screening tool for RV function.

P 15

Cardiac manifestation of acquired immune deficiency syndrome in infants and children

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On 30 June 1993, Romania reported 2,301 children HIV-infected under 13 years, as part of 2,461 total HIV-infected patients. Fifty-four percent of all infected children from Europe live in Romania and 67.9% of them have neither congenital infections or blood-product acquired infections. We have investigated the occurrence and significance of cardiac abnormalities in 223 Romanian children HIV-infected (1990-1993). One hundred-twenty-three consecutive autopsied AIDS children (1990-1993) were reviewed retrospectively to evaluate the possible relation between clinical and hystopathological cardiac finding. In 40% of cases we found myocarditis. A second group of 100 consecutive patients with known HIV-infection, most of them cardiac asymptomatic, were prospectively evaluated (1993), using two-dimensional and M-mode echocardiography (also for study of left ventricular performance), chest X-ray and ECG. Cardiomegaly (chest X-ray) and sinus tachycardia were the most common abnormalities seen in this study. The ECG abnormalities were not related to left ventricular function. Sonographic abnormalities occurred in 70% of cases. Two patterns of left ventricular performance function abnormalities were noted—hyperdynamic left performance with enhanced contractility (28%) or diminished contractility as in dilated cardiomiopathy (32%). In conclusion, routine serial noninvasive assessment of HIV infected children is strongly recommended because occult cardiac abnormalities are common in these children. Possible cardiac involvement are predictable in relation with CDC stage of disease. Cardiac morbidity and mortality are more common with advanced HIV-infection.

P 16

Determination of right and left ventricular volumes in the human fetus

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Ventricular volumes have been determined in the fetal sheep heart using high resolution echocardiographic imaging. We studied 50 human fetuses at 18-40 wks gestational age (GA) who had no cardiac or extracardiac abnormality. Both ventricles were imaged in orthogonal planes that corresponded to views obtained postnatally—subcostal coronal and sagittal views for the right ventricle (RV) and apical four- and two-chamber views for the left ventricle (LV). Studies were recorded on videotape, and off-line analysis was performed from digitally acquired loops of consecutive frames using a computer assisted system. Volumes were calculated at end-diastole (EDV) and end-systole (ESV) using a biplane Simpson's rule algorithm. Ejection fraction (EF) was calculated as (EDV-ESV)/EDV. LV volume calculation was possible in all fetuses; in five fetuses RV volume calculation could not be done due to inadequate imaging. The RVEDV ranged from 0.65 ml at 18 wks GA to 7.03 ml at term, the LVEDV ranged from 0.45 to 6.55 ml, respectively. The mean RVEDV normalized for estimated fetal weight (±SD) was 1.84±0.32 ml, the mean normalized LVEDV was 1.47±0.26 ml. Mean RVEF was 0.66±0.04, LVEF was 0.67±0.04. The RV was larger than the LV throughout the second half of gestation. The ratio of RVEDV/LVEDV ranged from 1.44 at 18 wks GA to 1.07 at term. There was no significant difference between the mean ratio of RVEDV/LVEDV at 18-22 and 23-28 wks GA (1.31±0.08 vs 1.32±0.07), but there were significant differences between mean ratios later in GA (1.24±0.07 at 29-33 wks, and 1.13±0.05 at 34-40 wks, respectively; $p<0.05$). Measurement of RV and LV volumes are feasible in the human fetus. This may be useful for prenatal assessment of ventricular size and function. Our results confirm the dominance of the RV at mid-gestation and show that the faster growth of the LV occurs mainly in the last quarter of GA.

P 17

Secondary fenestration late after Fontan repair improves a poor hemodynamic condition

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Fenestrating a Fontan has become an established procedure in selected high risk patients. It has been shown to reduce early mortality and morbidity. A residual connection between the left and right atrium reduces right atrial pressure and improves cardiac output at the expense of some desaturation. In three patients (4, 12 and 28 yrs) with progressive exercise intolerance, a secondary fenestration was performed late after Fontan repair (4, 44 and 45 months). The first two patients had a low cardiac output with high systemic venous pressure and hepatomegaly (5-6 cm). The third patient (28 yrs) had low cardiac output without congestion but severe protein loosing enteropathy, abdominal distension and complaints of thoracic oppression. The atrial septum was punctured with a Brockenbrough needle and dilated with an 8 F sheath. The septum was then further dilated with increasing sizes of balloons until a resting arterial saturation of 85% was obtained (final balloon size 8-10 mm). After the procedure, the cardiac output had significantly increased by 25±3%; all patients reported an increased exercise tolerance. The mild degree of cyanosis was very well tolerated. The hepatomegaly and central venous pressure had decreased. The complaints of thoracic oppression in the third patient had disappeared; the protein losing enteropathy improved with no further need of intravenous albumin or steroids. Partial reclosure of the fenestration occurred in one patient by one month after the procedure; the atrial shunt remained stable in the other patients. We conclude that in patients with a poor result of the Fontan operation, late percutaneous fenestration can be performed safely with a good result. This procedure should be considered before proceeding to more incisive treatments.

P 18

Assessment of great vessels abnormalities in conotruncal malformations by magnetic resonance imaging

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The purpose of this study was to define the role of MRI in the assessment of great vessels abnormalities in conotruncal malformations (CTM). Forty-five pts ranging in age from 2 d to 16 yrs (mean six yrs) with CTM were studied on a 1.5 T system (Picker) with standard gated spin-echo (SE) sequences in multiple planes in conjunction with cine Gradient-echo (GE) sequences (TR=20-40 ms, TE=12 ms, θ=30°) in all cases. In some cases, flow velocity map (FVM) sequences were used as well in order to evaluate associated flow alterations. Anomalies were tetralogy of Fallot (19), pulmonary atresia (4), double outlet right ventricle (5), truncus (3), transposition of great vessels (5), postoperative studies of CTM (9). Conjunction of the three types of sequences (SE, GE and FVM) provided added diagnostic information compared to echocardiography and angiography for a better determination of: (1) Segmental description and connections in complex anomalies; (2) site and degree of pulmonary stenosis; (3) size and connections of pulmonary arteries; (4) evaluation of collaterals; (5) presence or absence of pulmonary trunk in pulmonary atresia; (6) shunt patency; (7) postoperative complications after surgery such as tracheal compression, aneurysmal dilatation of pulmonary outflow tract, pulmonary regurgitation and stenosis; (8) associated anomalous venous vessel course in the thorax; and (9) associated aortic arch anomalies such as double aortic arch and other vascular rings. Combined utilization of these three types of sequences added valuable morphological, functional and flow related diagnostic informations in assessment of congenital great vessels anomalies in CTM. The use of GE and FVM sequences in adjunction to SE sequences brought about additional informations.

P 19

Efficacy of routine fetal ultrasound screening for congenital heart disease

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In order to assess the test characteristics of routine fetal echocardiography, e.g., the fetal four-chamber view, a follow-up study on fetuses screened routinely by means of ultrasound has been performed. Major arguments in favor of screening are the strong association of congenital heart disease with other congenital malformations and/or chromosomal aberrations, optimal postnatal care for the newborn with congenital heart disease and the possibility of termination of pregnancy after detecting a fatal malformation. The efficacy of this strategy for prenatal detection of congenital heart disease, however, has not yet been evaluated. A prospective cohort study of 7,000 normal pregnancies has been conducted in the Rotterdam metropolitan area to establish the value of routine fetal echocardiography. Women referred for routine obstetric ultrasound, between 16 and 24 wks gestational age, were invited to participate. The results of the fetal four-chamber view evaluation were reported. With a suspected abnormality, further assessment in a level three referral institution followed. Postnatal follow-up was ensured by way of Dutch community health services. If a cardiac malformation was suspected, further evaluation by a pediatric cardiologist was requested. Follow-up was completed at six months postnatally. Comparison of prenatal with postnatal data provided information concerning the efficacy of this screening procedure. For the first 4,657 participants, the sensitivity reached 4.3% (0.53-15.0), the specificity 99.9% (99.8-100), the predictive value positive 28.6% (3.7-71.0) and the predictive value negative 99.0% (98.7-99.3). We conclude that in routine fetal ultrasound the fetal four-chamber view is of limited value as a screening test for congenital heart disease.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

P 20

What to prefer? Cold or warm cardioplegia

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Both cold and warm cardioplegia are used to limit left ventricular (LV) ischemic injury during prolonged cardiac arrest, but it is not clear which form of cardioplegia is superior. Three groups of pigs (body weight 80-90 kg) were studied—a control group C (sham operated, no cardioplegic arrest, n=5), group 1 (n=5) with 3½ hours intermittent cold and group 2 (n=5) with continuous warm cardioplegic arrest. LV pressure was measured by a Millar micromanometer and LV short-axis and wall thickness by pairs of ultrasonic crystals. Measurements were performed at rest (B), after one hour (R1) and after two hours of reperfusion (R2). LV systolic function was assessed from fractional LV short-axis shortening (Sh) and LV diastolic function was assessed from the time constant of relaxation (T, ms) and the constant of myocardial stiffness (stress-strain-relationship, b). Sh was reduced at R1 and R2 in group 1 and 2 when compared to C. T was significantly prolonged at R1 and R2 in group 1 (111 and 149 ms, respectively) but remained unchanged in group 2 (61 and 62 ms, respectively) when compared to C (45 and 47 ms, respectively; $p<0.05$ vs group 1); b was increased with cold but was unchanged with warm cardioplegia. In conclusion, both cold and warm cardioplegic arrest are associated with systolic dysfunction. However, warm cardioplegia keeps the ventricle elastic and leaves the inactivation process unaffected, whereas cold cardioplegia makes the ventricle stiff and poorly relaxing. Our data indicate that warm cardioplegia is superior to cold.

P 21

Quality of life after surgery for vascular airway compression—long-term follow-up

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Children after surgical relief of vascular airway compression (VAC) are considered cured and therefore rarely receive long-term follow-up by a pediatric cardiologist. To test this approach, we studied noninvasively our patients who underwent surgery for VAC at least five years ago. Thirty-two pts operated within 1979-1988 were studied 9.1±3.0 years after surgery for vascular ring (n=20), pulmonary artery sling (n=3) and anomalous innominate artery (n=9). Clinical evaluation, echocardiography, pulmonary function testing, pulmonary perfusion scintigraphy and magnetic resonance were performed. All pts were clinically well, although one-third suffers from frequent respiratory tract infections and one-quarter manifests occasional stridor. Magnetic resonance did not show any significant airway or vascular deformation. Scintigraphy revealed in 19% asymmetric pulmonary blood flow. Lung function abnormality was encountered in 80% of patients, predominantly (72%) features of central airway obstruction. Although quality of life of children following surgery for VAC is excellent, high percentage of pathologic lung function tests is of concern and substantiates the need for close long-term follow-up.

P 22

Effectiveness of oral sotalol for treatment of pediatric cardiac arrhythmias

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The results of oral sotalol therapy in 12 children aged 7 wk-15 yrs (mean 6.8 yrs), weight 6-57 kg (mean 27.2) are presented. Two groups of pts were analyzed—Group I, seven pts, narrow QRS tachycardia and/or preexcitation (four pts normal heart, three pts LV dysfunction in course of arrhythmia) and Group II, five pts, ventricular complex arrhythmias (two pts normal heart, three pts prolapsed MV). Prior to sotalol, in all pts, treatment with 1-8 antiarrhythmic agents had been unsuccessful. Oral dose of sotalol ranged 1- 8 mg/kg/day, mean dose 2 mg/kg/day bid or tid. The efficacy of sotalol was estimated on standard ECG, 24-hr Holter and symptoms. In Group I (seven pts, four with WPW, two PJRT, one EAT), sotalol was effective in three pts, in three was partially effective together with digoxin (one pt) or digoxin and verapamil (two pts). In one child it was ineffective. In Group II (five pts, four with VeEx Lown group IV B, one with Lown group III), sotalol was only partially effective in three pts with VT. In two pts, sotalol was ineffective. Overall treatment was successful or partially successful in nine pts (75%) during 1-36 months of follow-up (mean 8.6 months), but in three cases (25%) only as combined therapy. No adverse effects and proarrhythmia during sotalol therapy were observed. QTc interval did not change significantly. In three pts (25%), sotalol was completely ineffective.

P 23

Prevalence, its seasonal and regional variations, risk of dying, and survival in aortic stenosis

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The data are based on a prospective study of children born with a cardiac malformation in Bohemia (population 6.3 million). All infants were examined four times in the first year of life and at the age of three and seven years. Those suspected of having a heart disease were referred solely to our center. Necropsy was compulsory in all children who died and those confirmed as having a cardiac malformation were included in this survey. Prevalence of aortic stenosis (AS) at birth was calculated in all infants born from 1977 to 1984. Of 664,218 infants, 4,409 had a congenital heart defect and in 274 of them (41.2/100,000 live birth, 6.2% of all heart defects), this proved to be AS. The ratio of boys to girls was 1.95:1. The prevalence differed significantly among regions. Variations of the prevalence in different months of the year were not significant. AS was found in 28 (2.5%) of 1,122 infants with a malformation of the heart who died or were admitted with a high risk of dying. A prospective population based-study on survival has shown that of all 409 infants born with AS from 1980 to 1990, 96.3% survived six months, 96.1% the first year, 92.9% five years and in total, 91.2% reached the age of 10.

P 24

Cardiac pacing in children—18 years experience

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The aim of the study is to analyze the results of permanent pacing in children. From 1975 to August 1993, 53 children at mean age 8.4±4.3 yrs (0.6-15.3) had pacemakers (PM) implanted. Forty-four had AV block and eight sinus node diseases. In 22 pts, PM implantation followed open heart surgery. Primarily, 18 epimyocardial leads were implanted (group I) and 34 endocardial (group II). The mode of pacing was VVI-48, VDT-2, VVI-R-1, DDD-R-1. Fifty-one pts were followed for a mean period of 7±4.6 yrs (0.3-17) and all of them had normal physical and psychological development. Six pts died (all from group II), but only in one of them was failure of PM system suspected. PM were explanted because of sepsis in two pts (one from each group). In 25 of the remaining 43 pts, 52 reimplantations were carried out (45 changes of generators and 29 of leads), and in seven pts this happened more than twice. Mode of pacing at the end of the follow-up was VVI-31, VVI-R10, DDD-R-2. Group I (17 pts followed-up)—No fracture or displacement of leads were observed. Exit block was the cause of reimplantation of 14 leads in 10 pts. The mean time of an effective stimulation was 5.4±4 yrs. At the end of follow-up, seven pts were with EMC and 10 with EC leads. Group II (26 pts followed-up)—Fifteen heads were implanted in 10 pts (one displacement, nine fractures in six pts and five exit blocks). The mean time of an effective stimulation was 5.2±2.3 yrs. Explantation of two endocardial and placement of an epimyocardial leads using cardiopulmonary bypass was done in two pts. The results of the study do not demonstrate any significant difference in the duration of an effective stimulation between the two groups. We now prefer the placement of one ventricular endocardial lead in childhood because of low frequency of exit block, lack of troubles in lead placement and avoidance of thoracotomy.

P 25

Long-term follow-up after surgical repair of tetralogy of Fallot in infancy and childhood

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In order to gain insight into the effects of sequels after surgical repair of tetralogy of Fallot on the health related quality of life in the long-term, we studied 77 patients 14.7±2.9 years after surgical repair of tetralogy of Fallot in infancy or childhood. All patients underwent a complete cardiac examination, including medical history, physical examination, standard 12-lead ECG, echocardiography, exercise test and 24 hour ambulatory ECG. Personal health assessment was 'good' or 'excellent' in 82% of the patients, which is not significantly less than in the normal population. Fifty-five patients (71%) had been free from medical or surgical intervention, 12 (16%) underwent repeat cardiac surgery because of anatomic/hemodynamic sequels and 10 needed treatment of arrhythmia. Of these, seven patients had sinus node dysfunction. Thirty-nine patients (50%) had a markedly dilated right ventricle with substantial to severe pulmonary regurgitation, six (8%) had signs of elevated right ventricular pressure, and 32 (42%) had a normal sized right ventricle. The maximal exercise capacity of the patients with a normal sized right ventricle was 98±14%, of patients with a markedly dilated right ventricle 83±19% ($p<0.05$), and of patients who had elevated right ventricular pressure 86±19% ($p=0.08$). The 24-hour ECG showed that 72% of the patients had arrhythmias. There was no association between right ventricular dilatation or elevated pressure and arrhythmias. There was no difference in personal health assessment between patients who had needed repeat cardiac surgery and patients who had been operated only once. Differences in exercise capacity and whether or not antiarrhythmic medication was used did not account for differences in personal health assessment. In conclusion, the health related quality of life long-term after surgical repair of tetralogy of Fallot is good, and comparable to that of the normal population. Neither sequels nor re-intervention seem to affect the subjective well-being.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

P 26

Examination after aortic coarctation repair—comparison of serial Doppler ultrasound and blood pressure measurements during follow-up

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The aim of this study was to assess the usefulness of serial Doppler ultrasound compared with blood pressure (BP) measurements and with clinical findings during follow-up of patients (pts) after coarctation repair. The study included 84 pts (52 M, 32 F, mean age 5.9±4.5 years at the first investigation) with two to five consecutive examinations (n=251) over a mean time of 2.5 years. In seven cases reoperation or balloon dilatation of recoarctation was performed during the study period. Peak systolic velocity and diastolic flow in the descending aorta (DA) were assessed by continuous wave Doppler and compared with oscillometric BP measurements on the upper and lower limbs as well as with clinical findings (pulse quality). Mean value of peak systolic velocity showed no significant difference between first and last investigation (2.41±0.48 vs 2.37±0.49 m/s) whereas blood pressure differences were significant (3.5±15.0 vs 0.1±14.7 mm Hg). Over all, 38 pts presented with increasing velocity in the DA and 34 with increasing BP gradients during follow-up. The flow profile remained limited to systole in 62 pts; three pts developed an abnormal diastolic flow, combined with increasing peak velocity in one of them. Twelve pts showed normalization of diastolic flow, nine of them spontaneously and three after intervention. There was a low correlation between intraindividual velocity changes and blood pressure gradient changes over time—only 96 of 167 pairs of consecutive intraindividual measurements showed concordant increase or decrease of values. In conclusion, Doppler ultrasound measurements of peak systolic velocities, proven to correlate better than BP-measurements with the degree of isthmic narrowing, showed only slight intraindividual differences during short-term follow-up compared with noninvasive BP gradients.

P 27

Early- and mid-term results of total cavopulmonary connection in 19 consecutive patients

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From 2/90 to 4/93, 19 consecutive pts (age 1.5 to 16.3 yrs, mean 5.7) with tricuspid atresia (11), single ventricle variants (7) and hypoplastic right ventricle (1) received total cavopulmonary connection (TCPC). Previous surgery in 18 pts included arterial shunts (19), pulmonary artery (PA) banding (4), Glenn shunts as first stage of TCPC (2), surgical enlargement of left PA (1). Mean PA pressure was eight to 20 mm Hg (mean 15). There was neither stenosis nor hypoplasia of PA branches except for two pts with moderate left PA stenosis. TCPC was performed with exclusion of total or most of right atrium. Pediculated pericardium extracardiac tube was used in nine cases. Two pts with left PA stenosis died postoperatively. One critically ill pt was reoperated early for baffle fenestration; we could close the defect percutaneously with buttoned device 11 months later. Residual shunt was closed by surgery in another pt. Other complications included severe congestive heart failure in eight pts, pleural and/or pericardial effusion in 10, pneumothorax in two, inferior vena cava thrombosis in one. In the follow-up (FU) (2.5 to 40 months, mean 11), the 17 survivors are well, in NYHA I or II class. Arterial oxygen saturations range from 83-96% in 16/17 pts. Chest x-ray shows normal heart size in 14/17 pts. All pts are in sinus rhythm. In 12 recatheterized pts, there is no obstruction of the TCPC, no ventricular dysfunction, but a small residual shunt in three. Superior and inferior vena cava mean pressures are respectively 12.6±2.7 and 14.3±2.4. From this series of 19 pts, we conclude that early and mid-term results of TCPC are very encouraging in the preoperative well selected pts, but longer FU period is needed.

P 28

Alterations of the beta-adrenergic system in congenital heart disease—experimental data and implications for therapy

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Heart failure in infancy and childhood is usually related to congenital heart disease (CHD). Several radioligand studies were performed to investigate beta-adrenoceptor (β-AR) number and coupling to the adenylate cyclase (AC). We found in right atrial myocardium of children undergoing cardiac surgery the β-AR density significantly reduced depending on severity of CHD. The β-AR down-regulation was subtype selective; however, in critically ill newborns there was also a significant decrease of β_2-AR. The β-AR reduction in CHD correlated to an increased sympathetic tone reflected by increased plasma noradrenaline levels. Signal transduction into the myocytes was measured by AC stimulation tests. While in moderate CHD the activity of the AC was reduced according to the reduced β_1-AR density, in severe acyanotic and cyanotic CHD, an additional partial decoupling of the β_2-AR to the AC occurred. In further investigations we found increased activities of the regulating Gi proteins in severe CHD probably responsible for these findings. Activated Gi proteins inhibit the intracellular signal transduction of the β-AR to the adenylate cyclase and consecutively damp myocardial contractility. These alterations of the transreceptor response to inotropes may explain why postoperative catecholamine therapy in infants and children with CHD is sometimes only effective in high dosages. Hypothermia and cardioplegia may additionally affect the β-AR system. Phosphodiesterase-inhibitors, acting beyond the β-AR and AC level, are probably useful to amplify the efficacy of catecholamines and seems to be protective against myocardial β-AR down-regulation. Further controlled studies are needed to validate the actions of these therapeutics. For treatment of chronic heart failure in CHD, drugs which are able to correct the neurohumoral imbalance should be used beside the standard regimens.

P 29

Cardiovascular exercise performance after venous switch operation for transposition of the great arteries

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Treadmill exercise testing was performed in 13 pts (age 9.9±3.4 yr) 9.1±2.5 yr after repair for simple transposition of the great arteries (TGA). All pathways had been proven to be unobstructive. Respiratory gas exchange was measured on a breath-by-breath basis. Aerobic capacity was assessed by determination of the ventilatory anerobic threshold (VAT) during exercise. VAT reflects the highest aerobic exercise level prior to a disproportionate increase of carbon dioxide output (VCO_2) and ventilation, relative to oxygen uptake (VO_2). Aerobic function was also assessed by analysis of the steepness of the upper segment of the V_{slope} (plot of VCO_2 vs VO_2, above VAT until HR=170). Ventilatory threshold in the patients was significantly ($p<0.05$) lower than normal and averaged 78±15% of the predicted normal value for children of comparable age respectively. Above VAT, VCO_2 rises significantly ($p<0.05$) faster compared to VO_2, with a slope of 1.3±0.2 in the patients vs 1.10±0.2 in the controls. This was associated with a significantly lower value for VO_2 during exercise in the patients compared to the normal controls (-10%; F =5.7; $p<0.05$). We conclude that aerobic capacity in patients after good repair for TGA is significantly lower than normals. The lower VO_2 and more excessive CO_2 elimination during exercise above VAT suggests impaired O_2 delivery to the tissues and development of lactic acidosis already at low exercise intensities.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

P 30

Is there a future in pediatric cardiology?

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This study investigates the effect of antenatal diagnosis of congenital heart disease (CHD) or Down's syndrome. All cases of CHD in infancy in one health region in 1985-1991 were classified as either 'complex' (C), 'significant' (S) or 'minor' (M). CHD was also classified as detectable (D) or not detectable (N) on a routine four-chamber scan. There were 269,913 births, 2,206 infant deaths and 300 cases of Down's syndrome. The outcome of 1,340 cases of CHD was:

Group	Total cases	Mortality (%) no surgery		Mortality (%) with surgery		Alive (%) at one yr	
C/D	100	46	(46)	17	(17)	37	(37)
C/N	76	24	(32)	18	(24)	34	(45)
S/D	96	21	(22)	11	(11)	64	(67)
S/N	651	53	(08)	59	(09)	539	(83)
M/N	417	9	(02)	0		408	(98)
Total	1340	153	(11)	105	(08)	1082	(81)

Assuming 20% detection on ultrasound and 60% on maternal blood testing, increased antenatal diagnosis of CHD or Down's syndrome would reduce activity as follows:

	Fetal ultrasound	Maternal Down's test
Birth prevalence of CHD	2.0%	3.7%
Total infant mortality	0.6%	0.3%
Pediatric cardiac surgery	2.9%	2.7%
Birth prevalence of Down's	2.7%	45.0%

These data suggest increased fetal diagnosis of CHD or Down's syndrome will have only a small impact on the practice of pediatric cardiology and surgery.

P 31

Patterns of ductus arteriosus constriction in utero by fetal Doppler echocardiography

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The features characterizing fetal ductus arteriosus constriction (FDC) diagnosed by echocardiography and Doppler examination (DE) are increased peak ductal systolic (S) and mid-diastolic velocities (D). Our purpose is to report other FDC patterns depending on the degree of constriction and fetal hemodynamics. Ductus arteriosus velocities (DAV) were measured in 100 normal (NL) fetuses from 17 to 38.5 weeks (wks); S ranged 0.45-1.5 m/s and D 0.06-0.35 m/s. FDC was found in 10 cases (32-36 wks). It was drug-induced by indomethacin (n=6), acetyl salycicylic acid (n=1), betamethazone (n=1). In two cases, no drug intake was found. The usual FDC pattern was found in five cases (S>2 m/s and D>0.5 m/s), associated to right ventricular hypertrophy (RVH) and tricuspid regurgitation (TR). DAV with C=0.90 m/s and D=0.55 m/s, reflecting low cardiac output, was observed in a case of supraventricular tachycardia with hydrops. NL DAV were obtained in all after drug withdrawal and arrhythmia control in one. Ductal occlusion was found in four cases with DAV and pulmonary velocities barely or not detectable, TR and pulmonary regurgitation, RVH and increased right-to-left atrial shunt (>0.6 m/s). Drug withdrawal in two showed first, increased DAV as observed in constriction, followed by progressive normal DAV. In the two other cases, not induced by drugs, transient right ventricular ischemia was found with a non-patent ductus at birth. In conclusion, FDC is not characterized by a unique pattern but depends on the degree of constriction and fetal hemodynamic. Ductal constriction and occlusion can alternate *in utero* and illustrates the dynamic ductal behavior.

Notes:

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

P 32

Natural history of congenital valvular aortic stenosis—an echo-Doppler study

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With echo-Doppler techniques it is possible to follow valvular aortic stenosis noninvasively. The aim of this study is to examine retrospectively how pressure difference (PG) developed over the years in a group of 129 children, age 0-18 yrs, followed by echo-Doppler exam from 1986 to July 1993; 77% of them were boys and 23% girls. Among these pts, 46 had only one examination, the rest had 2-7 measurements, mostly at an interval of one yr. If the PG between the left ventricle and the aorta increased above 60 mm Hg, balloon valvuloplasty or valvulotomy was done and the measurement before operation was taken as the last measurement of the child. Also, the septal and left ventricular posterior wall thickness were noted at each examination to see if there was left ventricular hypertrophy. Results: (1) Children under the age of two yrs showed a significantly faster increase in PG than older children (median 0.92 vs 0.43 mm Hg/month; Wilcoxon test $p<0.05$); (2) of 83 pt measured two or more times, 40 pts had an increase in PG. Though many patients showed a sudden increase, it is not possible to set a rule at which age this occurs; (3) left ventricular hypertrophy is a late and uncertain manifestation in aortic valvular stenosis. In conclusion, PG measurements are useful for the follow-up of aortic valve stenosis. A yearly control seems reasonable for children >2 yrs. For children 2 yrs, a control once per six months is advisable. Left ventricular hypertrophy is a late sign of the severity of the disease.

P 33

Fetal and perinatal diagnosis of complete AV-block and its outcome—a retrospective national multicenter study

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The time and mode of the diagnosis of complete AV-blocks (CAV-B), the risk of intrauterine and perinatal death as well as the occurrence rate of CAV-B are the objects of this retrospective study over an eight year time period (1985-1992) concerning 27 pregnancies. CAV-B was diagnosed by fetal echocardiography (F-echo) between the 18th and the 36th gestational week in 20 fetuses, while in seven a CAV-B was found only at birth. The mean annual occurrence rate was 0.04% (range 0.024-0.065) for a total population of 6.8 million inhabitants and a mean annual birth-rate of 81,000 between 1985 and 1992. In 13/27 CAV-B was the only finding; antinuclear antibodies were studied in eight of these 13 and were positive in five. The other 14/27 fetuses or newborns had important cardiac (CM) malformations. Death occurred in eight fetuses with CAV-B and CM and in one without CM. These nine fetuses had cardiac failure (CF); their ventricular heart rate (<50/min, range 36-70) was however not significantly lower than in 18 survivors; their atrial rate varied between 98 and 160/min. A pacemaker implantation became necessary in survivors without CM immediately after birth in 7/13 and at age 3-4 years in two. These nine had signs of CF or syncope but are still alive and well after a mean follow-up of six years. Of the six infants with CAV-B and CM, three died within the first year of life; in one case sinus rhythm occurred with tumor regression at nine months of age. Fetal echo should nowadays allow diagnosis of CAV-B and CM early in gestation (18th to 20th). CAV-B without CM has a good survival rate. These fetuses need to be followed closely till term. Monitored maternal steroid treatment may be used when anti-SS A/RO and -SS B/La increase. Termination of pregnancy should be considered in the presence of CAV-B with severe CM and/or chromosomal abnormalities before the 24th gestational week.

P 34

Left ventricular function after pulmonary autograft (Ross) procedure in children with severe aortic stenosis and/or insufficiency

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The pulmonary autograft (Ross) procedure is a more physiological approach to manage severe aortic stenosis and/or regurgitation in childhood than insertion of a mechanical or bioprosthetic valve. The newly developed on-line acoustic quantification (AQ) echocardiography utilizes quantitative differences in acoustic properties to differentiate between endocardium and blood. A continuous display of left ventricular (LV) cavity size is provided and systolic and diastolic function are assessed. The purpose of this study was to evaluate LV function after the Ross procedure by M-mode and AQ echocardiography. Thirteen patients were studied 0.6 to four yrs (mean 2±1.2) after surgery. Age ranged between 1.5 and 16 yrs (mean 12.7±4.2 SD). AQ measurements included end-systolic area (ESA), end-diastolic area (EDA), fractional area change (FAC), peak emptying (PER) and peak rapid filling rate (PFR), both normalized for EDA. M-mode measurements included LV internal dimensions and wall thickness (WT), expressed in percentiles for weight. The AQ measurements were compared to those of an age- and body weight-matched group of normal children. The preoperative M-mode measurements were compared to those after surgery. LV systolic and diastolic function returned to normal in all children shortly after pulmonary autograft operation.

	ESA cm^2/m^2	EDA cm^2/m^2	FAC %	PER EDA/s	PFR EDA/s
Pts	3.3 (1.3)	7.1 (2.6)	58 (7)	4.8 (1.1)	5.3 (1.3)
Nls	3.5 (1.3)	7.3 (2.4)	52 (9)	4.2 (1.1)	5.6 (1.5)

M-mode	LV systole	LV diastole	WT diastole
Before	96 (9)	92 (22)	72 (22)
After	56 (25)	50 (26)	66 (20)

P 35

Perioperative changes of cardiac areas after repair of congenital lesions—quantitation by intraoperative transesophageal echocardiography (TEE)

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The aim of this study was to assess the ability of TEE to demonstrate intracardiac volume changes after repair of lesions with intracardiac shunting by changing cardiac areas on echocardiography. Twenty-five pts with atrial septal defect (ASD), 16 pts with ventricular septal defect (VSD) and 12 pts with tetralogy of Fallot (TOF) were studied perioperatively before sternotomy (pr), at sternal closure (sc) and at the intensive care unit (in) by TEE. Manually traced TEE images of the cardiac areas were used to determine perioperative volume changes. Repair of ASD resulted in a significant decrease of the right atrial area (pr 11.9 cm^2, sc 6.6 cm^2, in 6.9 cm^2) and right ventricular area (pr 10.7 cm^2, sc 6.0 cm^2, in 6.5 cm^2). Repair of VSD resulted in decreasing areas of the left atrium (pr 4.7 cm^2, sc 3.7 cm^2, in 3.0 cm^2), of the right ventricle in diastole (pr 6.4 cm^2, sc 4.9 cm^2, in 4.4 cm^2) and of the left ventricle in systole (pr 4.7 cm^2, sc 4.4 cm^2, in 3.5 cm^2). After repair of TOF, TEE showed a moderate reduction of the right ventricle (pr 5.8 cm^2, sc 4.9 cm^2) and left ventricle (pr 5.7 cm^2, sc 5.0 cm^2) We conclude that perioperative TEE may detect early changes of cardiac filling during repair of congenital lesions. These volume changes after repair of shunt lesions should be considered, when the postoperative cardiac filling status is monitored by TEE.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

P 36

Complications after arterial switch operation

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The aim of our study was to determine the influence of surgical management and associated heart defects on the incidence, nature, and severity of complications after arterial switch operation (ASO). Since 1977, 76 pts were followed at the Wilhelmina Children's Hospital after ASO. Mean age at follow-up was 7.3 yr (range 0.9-16.7). Pulmonary stenosis (PS) was the most frequent complication (12 moderate, 21 severe). PS was diagnosed 2.1 yr (0-7.2) after ASO. Predictors of severe PS were use of conduit in pulmonary artery reconstruction, coarctation and year of operation. Re-intervention for PS was performed in 18 pts. As first reintervention, 4/6 balloon dilatations and 8/11 patch angioplasties relieved stenosis. After previous patch angioplasty, balloon dilatation relieved PS in 1/3. Other complications include pulmonary hypertension in one pt (late death at age 9 yr), LV-dysfunction in seven, aortic stenosis in two. Abnormal 24-hr ECG's were seen in five pts—four had multiple ventricular extrasystoles with bigeminy, one had wandering pacemaker. Predictors of both LV dysfunction and dysrhythmias were age at ASO and previous banding. In conclusion, pulmonary stenosis is the most frequent complication of ASO and a cause for concern. Postponing the arterial switch operation after the neonatal age, is associated with dysrhythmias and LV dysfunction in later life.

P 37

Follow-up after aortic balloon valvuloplasty

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Balloon valvuloplasty (BVP) for management of valvular aortic stenosis (VAS) has been routinely performed in our hospital since 1987. Between April 1987 and December 1992, 41 children (age range 3 d-15.6 yrs, mean 5.9 yrs) underwent BVP for VAS. Ten of these had had previous surgical valvulotomy. After a mean follow-up of 3.9 yrs (range 1-6.5), 31 patients (pts) were free of reintervention or death. One pt died of mitral valve endocarditis, and one of persistent left ventricular dysfunction. BVP was inadequate in three pts, of whom two underwent surgical valvulotomy at short-term and one repeat BVP at longer-term. The continuous wave (CW) Doppler gradient had decreased to 54±27% (mean±SD, $p<0.0001$) and after one yr to 50±21% of the pre-BVP value. At latest follow-up, the mean gradient had not changed, although this feature varied in the individual patient. There was no significant difference in residual gradient at latest follow-up between patients treated before and after the age of one year. Aortic regurgitation (AR) on echo shortly after BVP had occurred or increased in 41% of the pts, and was severe in 7%. At later follow-up in another 12%, AR progressed to severe. This necessitated valve replacement by a pulmonary autograft in five pts after 0.3-4 yrs. Progression to severe AR at follow-up was only observed in pts presenting with already moderate AR shortly after BVP. In conclusion, after BVP for VAS, 76% of pts remained free from reintervention for either AR, residual VAS, or death. Only in pts who presented with moderate AR after BVP, progression to severe AR during follow-up was observed.

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https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

P 38

Comparative responses of systemic and pulmonary veins to the presence of an intravascular stent in a swine model

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Intravascular stenting of pulmonary vein (PV) stenosis has a very high rate of reocclusion. Using a Yorkshire pig model, we studied the response of the normal PV to the presence of an intravascular stent (Johnson & Johnson Palmaz), using the inferior vena cava (IVC) as its control. Eight Yorkshire pigs (weight 19-24 kg) were placed on cardiopulmonary bypass (duration 17-36 minutes) and stents placed in the right lower pulmonary vein and IVC above the bifurcation. A total of 10 stents [PV (1 cm medium, n=5), IVC (1.5 cm iliac, n=5)] were placed in 5/8 animals with sacrifice at 4.9-6.1 months. Hemodynamics and angiography of PV and IVC were performed at one and three months. At sacrifice all stents were patent with no clot formation, and smooth endothelium seen covering the prosthesis. No difference was seen between PV and IVC. The pulmonary veins proximal to the stent appeared normal and no side vein obstruction detected. In this swine model, no difference was seen in PV to IVC reactivity to the presence of a stent. Swine pulmonary vein anatomy is such that the diameter of the inflated stent is much larger than that in the human clinical trials. The stent diameter and flow may have a major impact on the re-occlusion rate seen in pulmonary vein stenosis.

P 39

Lipoproteins 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 three years of age was carried out in a private pediatric practice over a two-year period. 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 with a myocardial infarction prior to 55 years of age. Data from the study supports universal cholesterol screening after three 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.

P 40

Comparison of endocardial and epicardial dual chamber pacing in children

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Dual chamber pacing combines atrioventricular synchrony and rate responsiveness but as yet is not widely used in children. To determine the safety and efficacy of endocardial (ENDO) dual chamber DDD pacing, we compared 34 ENDO systems with eight epicardial (EPI) systems in 41 children, implanted between May 1986 and June 1993. Indications for DDD pacing were congenital heart block (n=20), post-op heart block (n=16) and sick sinus syndrome (n=5). ENDO implants were significantly older (median age 11.1 vs 5.4 yr) and heavier (mean weight 34.6 vs 16.5 kg) than EPI implants ($p<0.005$). Voltage thresholds were similar in both groups at implant; however, ventricular ENDO leads had significantly lower thresholds at follow-up (1.2±0.7 vs 2.3±2.4 volts) than EPI leads ($p<0.01$). Lead impedance was significantly lower in atrial EPI (395±182 vs 299±55 ohms) compared to ENDO leads ($p<0.01$). Lead impedance was also significantly lower in ventricular EPI (350±66 vs 525±117 ohms) compared to ENDO leads ($p<0.01$). Three EPI generators were electively replaced; one of these was replaced with an ENDO system because of lack of atrial sensing. Two EPI atrial leads had poor sensing. One EPI ventricular lead had very high thresholds at generator replacement and the other previously placed ventricular lead was used. One ENDO ventricular lead was repositioned later on the day of implant. All ENDO systems have functioning DDD modes whereas one EPI system has lost atrial sensing and pacing and is programmed to VVI mode. There were two deaths from heart failure but no pacing related deaths. ENDO DDD pacing systems in children have lower thresholds and higher impedances with significantly less current drain than EPI systems. There are less atrial lead problems than EPI systems. Smaller generators and lower profile leads should allow more infants to be paced transvenously.

P 41

Immediate occlusion of vessels—percutaneous, intravascular radiofrequency electrocoagulation (RF-E) using retrievable coils—an experimental study

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Newly developed retrievable coils were tested as detachable electrodes for immediate occlusion of high velocity vessels using RF-E. Newborn piglets (8-18 days old, weighing 4.5-5.5 kg) were anesthetized and acutely instrumented via cutdowns into both carotid and one brachial arteries. Two identical cylindrically shaped coils (length 3 mm, outer diameter 2.4 mm, inner diameter 1.4 mm; pfm, Cologne, Germany) were mounted on titanium-nickel core wire and placed via 3 F Nylon catheters into both femoral arteries and kept connected to the delivery wire which is isolated against surrounding tissue by the catheter. The system on one side served as control, the system on the other side was connected to a RF-generator (HAT 50, Dr. Osypka, Grenzach, Germany), the electrical circuit being closed via an external indifferent electrode. Angiograms (A) via the brachial artery demonstrated the adequate placement of the coils and the patency of the femoral arteries immediately thereafter. Application of 25 watt RF-current over 10 sec to the one side was carried out in five minute intervals until the occlusion of the artery was demonstrated by A. Thereafter the coils were detached from the wire and additional A were performed after 5, 15, 40 and 60 minutes to follow-up the patency of the control side. Complete occlusion was achieved in all cases after a maximum of three consecutive applications of RF-current. The control remained patent for a minimum of 45 minutes. On the side RF-E was performed, there was no evidence of contrast medium extravasation, and the arteries macroscopically remained intact. In contrast, continuous application of RF-current over 30 sec ruptured the arteries. In conclusion, intravascular RF-E may be a method for coil fixation and immediate occlusion of vessels with high blood flow velocities.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press

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Safety and efficacy of transvenous PDA closure with the Rashkind devise

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Ninety-four patients were enrolled in a prospective protocol for transvenous PDA closure. Eighty-eight patients, aged seven months to 43 years (median 2.9 yrs) weighing 6.3-67.0 kg (median 14) underwent implantation of the Rashkind PDA occluder. In five patients, occluder implantation was not attempted for duct diameters of less than 0.2 mm or more than 5 mm. In one patient with previous coarctation surgery, the occluder could not be placed. Duct diameters ranged from 0.5-3.6 mm (mean 2.1 mm); mean Qp/Qs was 1.4. One patient had previous surgical duct ligation. One procedure was performed through the internal jugular vein. Two occluders were implanted in three patients. No embolizations occurred. The complication rate was 6% with hemolysis in one (surgical device explantation and PDA closure), iliac vein thrombosis in one, femoral artery stenosis in two, mild left pulmonary artery stenosis in one. Closure was complete in 59% after 15 min, in 81% after 24 hrs and 89% after six months. Recanalization occurred in three patients. Clot formation on the umbrella was occasionally found within 15 min despite heparinization. Incorporation of umbrella legs into the vessel wall was seen on follow-up angiography. Incomplete closure was associated with large duct diameters.

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The Ross operation in critical aortic stenosis—a life-saving procedure?

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Critical aortic stenosis requires urgent intervention for patient survival. In most cases primary palliation as open commissurotomy or dilation procedures effectively reduce the left ventricular outflow tract obstruction (LVOTO). However some patients continue to be in severe left heart failure due to persistent LVOTO and/or aortic regurgitation (AR). In these infants we have employed the autotransplantation of the pulmonary valve into aortic position (the Ross operation) with favorable results. Eight patients aged five wk to nine mo underwent the Ross operation. All but one had previous palliations as commissurotomy, balloon dilation, transventricular Hegar dilation, singly (1), or in combinations (6). The severely malformed aortic valve and the proximal aorta was excised and replaced by the patient's own pulmonary valve and proximal artery into which the coronary arteries were reimplanted. A homograft conduit was inserted into pulmonary position. There were two operative deaths, both having severe endocardial fibroelastosis (EFE) at autopsy. One late death occurred due to progressive hypertrophic cardiomyopathy (in absence of any stenosis) and pulmonary hypertension one yr post-op. Four are thriving (two have trivial AR). One having small LV and localized EFE is in moderate failure but improving. In all survivors, the pulmonary autotransplant is growing with the patient. In conclusion, the Ross operation may be successfully employed in patients where other palliative measures fail. This technique should be considered as primary mode of correction in high risk patients, i.e. those with hypoplastic aortic valve rings, small LV, mitral regurgitation and localized EFE.

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Ventricular arrhythmias (VA) in patients with tetralogy of Fallot (TOF) and with follow-up longer than 20 years

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After surgical correction of TOF, a positive correlation was found between the incidence of VA and length of follow-up. In this study we evaluated the incidence and complexity of VA in a group of 48 pts with a postoperative follow-up longer than 20 yrs. The age of 48 pts at last evaluation (May 1993) was 34±9 yrs. All pts were evaluated with 24-hour Holter monitoring, treadmill exercise test, M, 2D and color Doppler echo. Holter monitoring showed VA in all 48 pts. According to the Lown class, 21 pts (44%) (Group A) were in class 4 (20 pts with couplets and nine pts with non-sustained ventricular tachycardia with mean heart rate of 145/min); 27 pts (55%) (Group B) were in class 3 or less (polimorphic ectopies were detected in 14 pts). In three pts (2A, 1B), there was concomitant 2° degree AV block, and in two pts (1A, 1B) both with PM, complete AV block. Ventricular couplets were induced by exercise test more frequently in Group A than in B (71 vs 14%, $p<0.05$); nevertheless, exercise tolerance was similarly reduced in both group (exercise endurance A 9±3 min, B 10±3 min). All pts had tricuspid incompetence, the right ventricle systolic pressure was higher in Group A than in B (47±4 vs 37±8 mm Hg, $p<0.05$). There was no difference between A and B for transpulmonary gradients (A 18±6, B 13±8 mm Hg) or right ventricle diastolic diameter (A 34±9, B 30±5 mm). In conclusion, the incidence of VA is very high in pts with long follow-up, and a Lown class 4 is found in a large number of pts (44%). Finally, there is good correlation between values of systolic pressure in right ventricle and Lown class. According to our results, it is not possible to draw firm conclusions about the correlation between VA and sudden cardiac death in this particular subset of pts.

https://doi.org/10.1017/S1047951100011318 Published online by Cambridge University Press