Volume 5 • Supplement 1 • Pages S1-S47

Cardiology in the Young

ISSN 1047-9511

Abstracts of the XXX Annual Meeting of The Association of European Paediatric Cardiologists, Bologna, 17-20 May 1995

01

The over-the-wire placement of the buttoned device for the occlusion of atrial septal defects—improved international results

Berger F, Lang P, Rey C, Walsh K, Wang HC, Ballerini L, Leung M, Worms AM, Rao PS, Sideris E

Pediatric Cardiology, German Heart Center, Berlin, Germany

The placement of the fourth generation buttoned device for ASD occlusion was simplified by an over-the-wire introduction (OW). The results for the OW in 37 consecutive patients were compared with those of 104 cases done with direct device placement (DP). OW includes piercing the center of the foam with a wire positioned in the left upper pulmonary vein; all subsequent manipulations are made over the wire.

	n	ASD (mm)	DEV (mm)	Full (%)	Abandoned (%)	Complications
OW	37	12-28	25-55	86	3	0
DP	104	6-26	25-50	78	10	4

The range of ASD repair increased to 28 mm and the abandoned cases decreased significantly. Complications of the DP included embolization (1), mitral insufficiency (1), atrial perforation (1) and unbuttoning (1). No complications have been noticed with OW yet. This change in method offers better stability of the device and it is less operator dependant. Follow-up studies should determine the benefits of the better device placement.

02

Occlusion of large atrial septal defects with a centering buttoned device

Sideris E, Leung M, Yoon JH, Chen CR, Lochan R, Worms AM, Meier B, Rey C

Athenian Institute of Pediatric Cardiology, Athens, Greece

Occlusion of 12 large ostium secundum atrial septal defects was performed by a centering buttoned device. A centering buttoned device is a modification of the regular buttoned device, where the counter occluder is sutured at 40% of the occluder. During centering, the counter occluder is stretched, forming a balloon-shaped structure and pulling the occluder over the center of the defect. The counter occluder forms a double-eight figure on the right side of the atrial septum after buttoning. Occlusion was performed in 12 patients rejected for occlusion by the regular buttoned device, either because a defect larger than 25 mm or an inadequate septal rim. The defect size varied between 23-31 mm and the device size between 45-60 mm. All devices were delivered through 11 Fr sheaths. Nine patients had effective occlusions and three had residual shunts. One patient with unbuttoning without embolization, developed hemolysis and was operated on two weeks after implantation; the other patients are doing well. The early results of the transcatheter occlusion of large atrial septal defects by the centering buttoned device are promising, justifying the continuation of larger clinical trials.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

03

Transesophageal echocardiography for umbrella closure of ventricular septal defects in the cardiac catheterization laboratory

Rigby ML, Horowitz ES, Redington AN
Royal Brompton Hospital, London, UK

Thirty patients have undergone attempted transcatheter occlusion of a ventricular septal defect using a combination of radiographic screening and transesophageal echocardiography. A Hewlett-Packard pediatric biplane or single plane probe was used for patients under 15 kg or an adult single, biplane or omniplane probe was used in larger patients. Retrospective review of the video recordings was undertaken to determine the morphological features of the VSD, the qualitative and quantitative Doppler measurements and to examine the placement of the double umbrella. There were 15 patients with a perimembranous VSD, 7 with muscular defects and 8 with a residual defect following surgery (2 perimembranous and 6 muscular). One patient underwent placement of 4 umbrellas and a second required 3. Incomplete visualization of 3 perimembranous defects and 4 muscular defects was attributable to the use of a single horizontal plane probe alone. Color flow Doppler mapping of the interventricular flow was possible in every patient, but the continuous wave Doppler velocity profile was incomplete in 10. Visualization of the exchange guide wire and Mullins transeptal sheath crossing the VSD was possible in 27 patients. Imaging of the distal and proximal umbrellas was possible in every case, but incomplete imaging of the VSD hindered umbrella placement in 4. In 2 of these, the umbrella was positioned in a tiny and insignificant muscular defect rather than the major VSD. Significant aortic regurgitation caused by the distal umbrella in 2 patients with a perimembranous defect was relieved by rotation of the umbrella. Moderate tricuspid regurgitation observed in 13 patients during the procedure became trivial or disappeared when the umbrella device was released. In conclusion, biplane or omniplane transesophageal echocardiography is essential for transcatheter umbrella closure of ventricular septal defects.

04

Medium-term follow-up after transcatheter umbrella closure of perimembranous ventricular septal defect

Heinsdijk M, Rigby M, Redington AN, Vogel M
Royal Brompton Hospital, London, UK

Since October 1991, 17 infants, children and adolescents have undergone attempted transcatheter umbrella closure of a perimembranous ventricular septal defect (VSD). The procedure was unsuccessful in 3 because the defect was too large. Two patients required surgery—1 because of malposition of the umbrella and one due to late severe aortic regurgitation. One late death was unrelated to the VSD closure. Of the remaining 11 patients, 10 have undergone detailed investigation to assess the outcome of the procedure 1 to 4 years (mean 1.6) later. On the standard surface electrocardiogram, 6 have complete (3) or partial (3) right bundle branch block. On Holter monitoring, all patients remain in sinus rhythm without atrial or ventricular premature beats or tachycardia. With transthoracic and transesophageal echocardiography together with 3-dimensional reconstruction, 4 patients had complete occlusion of the defect. The remaining 6 have small residual defects only. The Doppler derived right ventricular systolic pressure from the VSD or tricuspid regurgitant velocity profiles was 24-36 (mean 30) mm Hg. Although none had evidence of left ventricular outflow tract obstruction or aortic incompetence, apposition of the right coronary aortic valve leaflet and the distal umbrella were evident in 4. At cardiac catheterization, small residual defects were confirmed in 6 patients (mean Qp:Qs 1.9:1 and mean pulmonary arterial systolic pressure 28 mm Hg). In conclusion, following umbrella closure of perimembranous VSD, all patients are symptom-free; right bundle branch block is frequently observed and a small residual ventricular septal defect is present in more than half. Some patients appeared to have the capacity to develop aortic regurgitation or subaortic stenosis because of the position of the distal umbrella in the left ventricular outflow tract. Umbrella closure of a perimembranous VSD cannot be recommended as a routine procedure.

05

Interventional closure of ventricular septal defect with retrievable double cone-shaped coils—an experimental study

Neuss MB, Radtke WAK, Peters A, Redel DA, Kline CH, Swindle M
South Carolina Children's Heart Center, Charleston, USA and the Department of Pediatric Cardiolog, University of Bonn, Bonn, Germany

Retrievable double-cone shaped coils (pfm, Germany) are successfully being used for transvenous closure of patent ductus arteriosus. Adaptive behavior to pulsatile wall thickness, minimal size of the implantation catheter (4 or 5 Fr) and the ability to reposition the coil make it suitable for closure of ventricular septal defect (VSD). Retrievable double-cone shaped coils (0.028, 0.032 inch) with varying diameters up to 9 mm were implanted using venous or arterial approach in 6 Yucatan mini swine (13-17 kg) with congenital perimembraneous VSD very close to the aortic valve. Pressure and oxygen saturation were measured for hemodynamic evaluation of the shunt. Levocardiogram and aortic angiography, as well as color flow Doppler echocardiography were performed before and after coil placement. Complete occlusion or significant shunt reduction could be demonstrated 2-4 weeks after implantation. Successful transcatheter closure of ventricular septal defects with retrievable coils is feasible even in close proximity to cardiac valves.

06

The evolution of long-term follow-up of balloon dilation of native aortic coarctation in children

de Moor M, Fadely F, Oufi S, Galal OM
King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia

Between February 1985 and February 1994, a total of 98 balloon dilations (BAP) for native coarctation were done in 83 children (pts) (median age 16.5 months, weight 8.9 kg). The mean cath peak-to-peak systolic gradient was 49±20 mm Hg before BAP and 18±16 mm Hg immediately afterwards. Recatheterization in 57 patients (69%) after a mean of 21 mos (0-54 mos) demonstrated a gradient of 21±18 mm Hg; 14/57 (25%) pts underwent a second BAP for recoarctation. A total of 19/83 pts (23%) required surgical relief of the coarctation because of recoarctation or unsuccessful BAP. Ten pts (10/83 [12%]) developed complete occlusion of the affected femoral artery, and a further six (7%) partial occlusion. Five pts (6%) developed large aneurysms, of which four had surgery. A further 12 pts (14.5%) developed either small aneurysms or a ductal diverticulum which will require close follow-up. An aneurysm, or opening of a ductal diverticulum developed immediately (n=6) or was only detected at recatheterization (n=11) six to eight mos later. There was no significant difference in age at initial BAP regarding development of aneurysm or need for surgery to relieve the coarctation. There was a significantly greater ratio of balloon-to-descending aorta measurements in the group developing aneurysms (1.2±0.23) as compared to the group which did not (1.02±0.27; $p<0.02$). BAP of native coarcation although safe and successful in the long term in over 70% of pts, and carries a distinct morbidity (femoral artery occlusion, early and late aneurysm formation). Twenty-five percent will need repeat BAP as they grow. Use of a balloon equal to the descending aorta at the level of the diaphragm appears important in avoiding aneurysms. Further follow-up is needed to try to define those who benefit from this procedure.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

07

Endovascular stenting in coarctation

Schneider M, Karnpmann C, Will J, Goeldner B, Hausdorf G
Charité, Pediatric Cardiology and Cardiac Surgery, Berlin, Germany

Stent implantation in coarctation is a new approach. We implanted stents in 9 patients with coarctation. In 5 neonates with native coarctation, stenting was performed because of severe tubular hypoplasia of the aortic arch (diameter 3.5-14.6 mm) when balloon dilation had failed. The Palmaz stents were implanted through a 5-6 Fr sheath and inflated to a diameter of 5.4-8.6 mm. The residual gradient varied from 2-10 mm Hg. Additionally, stents were implanted in 4 pts with recoarctation as palliative or therapeutic approach. In 3 pts, Palmaz stents were implanted through a 5-6 Fr sheath and inflated to 6-12 mm. In the remaining pt, a covered wallstent was implanted to occlude a concomitant postsurgical aneurysm (diameter 20 mm). The pressure gradient was reduced to 0-14 mm Hg. In conclusion, implantation of endovascular stents is an effective palliative or even therapeutic approach in complex coarctation. Implantation of covered stents is of particular interest in post-surgical or post-interventional aortic aneurysm formation.

Notes

08

Balloon dilation (as initial therapy) for neonatal critical aortic stenosis—results up to 8.3 years follow-up

Egito E, Keane J, Moore P, O'Sullivan J, Colan S, Perry S, Lock J
Children's Hospital, Harvard Medical School, Boston, USA

The purpose of this study is to report our results with balloon dilation (BD) of critical aortic stenosis (Crit AS) in neonates. From 1985 to 1991, 46 consecutive babies <33 days of age had BD for Crit AS at median age of 5 days, and mean weight of 3.2 kg. Forty-one (89%) were in congestive heart failure (CHF) or shock, 33 (80%) intubated. The BD was completed in all, retrograde in 43 (umbilical 23, femoral 20), and antegrade in 3. Reduction >50% in the peak-to-peak systolic gradient was achieved in 32 babies with no significant aortic regurgitation (AR) post-BD. The final balloon to aortic annulus ratio was 0.91±0.05. Thirty-two (70%) babies had median hospital stay of 8 days, with no other intervention in 20. The overall mortality was 35%, 11 early and 5 late. Four deaths were considered BD-related, and the remaining 12 related to left-sided lesions. All 20 babies who had BD via femoral artery initially had pulse loss, but in 7 (35%) the pulse was restored with anti-thrombotic therapy. Significant AR post-BD was observed in 5 (11%) babies. Anatomic risk factor score for two-ventricle repair was retrospectively measured; 13 (38%) had a score >2 (Crit AS with 2 or more associated left-sided obstructive lesions, high mortality for two-ventricle repair), and only 1 survived longer than 4 mos. In the 33 babies who had score <2 the survival probability and freedom of reintervention at 8.3 years were 88% and 59% respectively. At follow-up of 4.3±1.8 yrs (1.4 to 8.3 yrs), 27 (90%) of the 30 survivors were in good or excellent clinical condition. Reintervention was undertaken within 1, 4, and 88 months in 22%, 30%, and 41% respectively. Recent Doppler studies showed a maximum gradient >50 mm Hg in 10 (33%), and significant AR in 4 (13%). Our data suggest that BD for Crit AS has satisfactory immediate and mid-term results when no more than 1 associated left-sided obstructive lesion is present.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

09

QT interval prolongation on the fourth day of life and SIDS—ECG findings from a prospective study on 33,034 infants

Stramba-Badiale M, Spagnolo D, Porta N, Bosi G, Perticone F, Pelargonio S, Salice P, Poggio GL, Marni D, Schwartz PJ
Departments of Medicine, Universities of Milan and Pavia, Departments of Pediatrics, Universities of Milan, Ferrara, Reggio Calabria, Rome and Pavia, Italy

We have proposed that some cases of SIDS might result from an episode of ventricular fibrillation triggered by a sudden increase in sympathetic activity affecting a heart with reduced electrical stability. We have recently demonstrated that a left sympathetic dominance, experimentally induced in puppies, prolongs QT interval and increases the susceptibility to ventricular fibrillation in the third week of life. With the objective of analyzing the QT interval in the neonatal period and its relationship with the occurrence of SIDS, we have performed a large prospective study. ECGs were obtained on the fourth day of life and QTc was calculated according to the Bazett's formula in 33,034 newborns. During the one-year follow-up, there have been 33 deaths, 23 for SIDS and 10 for other causes. Eleven of the 23 SIDS victims (48%) had a QTc exceeding the mean of controls (400±20 $msec^{1/2}$) by over 2 standard deviations (>440 $msec^{1/2}$), while all the victims for other causes had a QTc within the normal limits. Since the incidence of QTc greater than 440 $msec^{1/2}$ among the infants who survived was 2.4%, the risk of SIDS in the infants with a prolonged QTc (>440 $msec^{1/2}$) was 37.3 times greater than that of infants who showed a QTc within the normal limits (95% confidence interval 16.4-90.0). These results indicate that a significant portion of the SIDS victims may be identified very early by the observation of a prolonged QT interval on the 4th day of life and that neonatal ECG may become a simple and feasible tool for the assessment of risk for SIDS. The efficacy of a preventive strategy based on interventions that would counter the detrimental action of enhanced sympathetic activity during the first year of life could be assessed.

10

Efficacy and proarrhythmia of oral sotalol in infants and children

Pfammatter JP, Paul T, Kallfelz HC
Pediatric Cardiology, Hannover Medical School, Hannover, Germany

Experience with sotalol, a class III antiarrhythmic agent, is limited in pediatric patients and the incidence of proarrhythmia has not been systematically investigated. All children treated with oral sotalol at our institution were prospectively evaluated with serial Holter-ECG to assess efficacy and proarrhythmic effects during treatment with sotalol. Since July 1987, 71 infants and children (mean age 7.3±5.8 years) have been treated with oral sotalol for symptomatic supraventricular and ventricular tachycardias. Mean follow-up (or duration of therapy if already stopped) was 18±12 months (range 1-40 months). Sotalol was most effective in the group with supraventricular reentrant tachycardias with complete success in 27/41 patients (66%) and partial success in 11/41 (27%). Atrial flutter was completely controlled in 9/19 predominantly postoperative patients (48%) and partial effect was noted in a further 7/19 (37%). Complete success was achieved in 3/11 (28%) of patients with ventricular tachycardias and partial effect in 4/11 (36%). In 10 of 71 patients (14%), proarrhythmic effects were detected by serial Holter-ECG (Torsades-de-pointes in 1 child, frequent premature ventricular beats in 6, sino-atrial block in 1 and 2° AV-block in 2 patients). In 6 of these children (8%), sotalol had to be discontinued. The incidence of proarrhythmia was not dose-dependent. In all but one of the patients with proarrhythmia, it occurred during the early in-hospital phase of treatment begin. Sotalol proved to be an efficient antiarrhythmic drug. Its high rate of proarrhythmic effects warrants in-hospital treatment begin and close supervision by serial Holter.

11

Follow-up study of exercise-related ventricular tachycardia in children with apparently normal heart

Drago F, Vignati G, Calzolari A, Mauri L, Figini A, Ragonese P
Ospedale Pediatrico Bambino Gèsu, Roma, Cardiologia Pediatrica, Ospedale Niguarda, Milano, Italy

Ventricular tachycardia (VT) is the most common diagnosis among children with exercise or emotion-related syncope. The aim of the study was to assess the prognosis and the clinical course of children with exercise-related VT and apparently normal heart. Since January 1984, we studied 12 consecutive children (mean age 9.7±4.5 years) with exercise-related VT and, at the initial detection, without demonstrable heart disease, as assessed by clinical evaluation, electrocardiogram and mono-bidimensional echocardiogram. Five patients had exercise related syncope, 1 exercise intolerance and 6 were asymptomatic. VT, detected by exercise testing and Holter monitoring, was polymorphic and non-sustained (<30 sec) in 2 symptomatic patients, monomorphic with left bundle branch block morphology and inferior axis in the others (sustained in 1 symptomatic and in 2 asymptomatic). The mean VT rate was 256±23 bpm in the symptomatic children and 190±31 bpm in the others ($p<0.005$). Successful chronic treatment was achieved with beta-blockers in 5 cases, propafenone in 5 cases and sotalol in 2 cases. During follow-up (4.6±2.6 years), 1 patient with polimorphic VT on therapy suddenly died during effort, the other symptomatic pts had no variation of their arrhythmia on routine tests, except 1 who, after propafenone was stopped, had ventricular fibrillation during an exercise test. Among the 6 asymptomatic pts, 4 had no evidence of VT on routine tests after 2.5±1 years; the others had no variation of their arrhythmia and treatment. In conclusion, exercise related VT in children seem 1) mostly to arise from the right ventricle, 2) to be successfully treated by antiarrhythmic drugs with beta-blocking activity, and 3) to have more severe prognosis if symptomatic or polimorphic.

12

Children below six years of age with accessory pathway related tachyarrhythmias—whom to ablate and how?

Hebe J, Weiß C, Siebels J, Petersen V, Kuck KH
Department of Cardiology, University Hospital, Eppendorf and St. Georg Hospital, Hamburg, Germany

By August 1994, a total of 103 children (<15 yrs) underwent radiofrequency current (RFC) ablation for accessory pathway (AP) mediated supraventricular tachycardia (SVT). Of these, 23 were infants aged less than 6 years (11 females, 12 males; 3.9±1.6 yrs). SVT was refractory to an anti-arrhythmic regimen with 3 to 6 different agents. Tachycardia-related symptoms included cardiac arrest in 2 infants with WPW, syncope in 3, reduced physical stress capacity in 11 and SVT-related chest pain in 1. Thirteen pts had an overt AP, with an additional concealed pathway in 2; the remaining 10 pts had a concealed AP, in 6 of whom it sustained the permanent form of junctional reciprocating tachycardia (PJRT). In the latter group, 3 infants had reduced left ventricular function (fractional shortening 0.13-0.21). In 11 infants, a single 5 or 6 Fr steerable catheter was used for therapeutic intervention. In 9 infants a second catheter was required for diagnostic purposes. Within 25 sessions, 22 infants were cured by a median of 7 RFC applications. Procedure duration was 3.4±1.9 hours, with a median radiation exposure time of 29.0 minutes. During a 19 month follow-up (median), 20/22 infants were asymptomatic and required no antiarrhythmic medication. Two pts had recurrences of SVT (1 PJRT), one of which underwent a successful repeat session. Tachycardia-related reduced left ventricular function evolved to normal values in all 3 infants within the follow-up period. We conclude that 1) RFC-ablation of accessory pathway-mediated supraventricular tachycardia is safe and effective in children aged less than 6 years; 2) reduced invasiveness due to simplified catheter techniques is achievable in this particular patient population; 3) in children with an age <6 years, indication for RFC-ablation is restrictive and should be provided in pts with severe symptoms due to drug-refractory SVT.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

13

Radiofrequency catheter ablation for treatment of atrial flutter after surgery for congenital heart disease in young patients

Paul T, Trappe HJ, Pfammatter JP, Luhmer I, Kallfelz HC
Pediatric Cardiology, Hannover Medical School, Hannover, Germany

Radiofrequency catheter ablation has been used successfully in adult patients for treatment of atrial flutter. Three young patients (mean age 14.6 years) with common type atrial flutter (2) and uncommon type atrial flutter (1) underwent electrophysiologic study. Cardiac diagnoses were tricuspid atresia after Fontan operation, status after closure of an atrial septal defect of secundum type, and status after surgical valvotomy for valvar pulmonary stenosis, respectively. Indication for ablation was presyncopal episodes in all 3 patients. Mean flutter cycle length was 310 msec. Right atrial endocardial mapping revealed areas with discrete electrograms with activation times of -20 to -60 msec before P wave onset. Radiofrequency current application (500 kHz) with 30 W for 30 sec at theses sites terminated atrial flutter and prevented reinduction in all 3 patients. No complications were observed. Follow-up (mean 3.7 months) revealed recurrence of uncommon type of atrial flutter in one patient who previously had had common type. The remaining two patients are in stable sinus rhythm. In conclusion, radiofrequency catheter ablation is an effective treatment of atrial flutter after surgery for congenital heart defects in young patients.

14

Recovery time for left ventricular (LV) size and function in children with incessant tachycardia following radiofrequency ablation therapy (RFAT)

De Giovanni JV, Edgar RM, Stümper O, Wright JGC, Silove ED
Birmingham Children's Hospital, Birmingham, UK

Incessant tachycardias in children are uncommon but, when present, can lead to marked LV dilatation with severe reduction in systolic function. We looked at the natural history of the LV in patients with incessant supraventricular tachycardia who underwent RFAT. Six patients aged between 1 month and 12.5 years (mean 6.3 years) underwent RFAT—5 for the permanent form of junctional reciprocating tachycardia and 1 for focal atrial tachycardia. All, apart from the youngest one, showed increased LV dimension with a variable reduction in parameters for LV systolic function—namely ejection fraction (EF) and fractional shortening (FS). The worst figures were found in the eldest patient who had overt heart failure and who had an LV diastolic dimension of 6.7 cm with an EF of 20 percent and an FS of 13 percent. The severity of LV dysfunction probably reflects the long duration of undetected tachycardia. All have shown improvement in LV size and function following successful RFAT with up to 14 months before normalization of function. Gross LV dilatation and dysfunction due to incessant supraventricular tachycardia is reversible, even in extreme cases, although recovery may take many months.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

15

Dynamic 3D reconstruction by multiplane transesophageal echocardiography (MTEE)—superior spatial assessment of cardiac pathomorphology in adolescents and adults with congenital heart disease

Grabitz RG, Franke A, Mühler EG, Krebs W, Klues HG, Flachskampf FA, Hanrath P, von Bernuth G
Pediatric Cardiology, Medicine Clinic I, RWTH, Aachen, Germany

MTEE allows detailed 2D imaging of the heart by rotating the transducer array around a central stable axis using a single echo window, thus providing a unique opportunity for dynamic 3D reconstruction. Utilizing a multiplane transducer (5 MHz, 64 elements, Hewlett-Packard®) steered by a stepper motor and connected to a computer-based image acquisition and 3D-reconstruction system (TomTec®), we obtained 79 tomographic data sets (180° rotation, 2° increments, ECG-triggering, respiratory-gating, mean acquisition time 2.2±1.1 min) in 28 patients (12 male, 16 female, mean age 38) pre- and/or post-surgical repair. The diagnoses included secundum ASD (11), sinus venosus ASD (2), Fontan circulation (2), subaortic stenosis (SAS) (5), functional univentricular heart (3). Dynamic 3D displays from multiple viewpoints were generated to obtain a maximum of spatial and dynamic information. The displays provided unique "unroofed" views from surgeon's point of view, allowing better understanding of complex anatomy and subsequent planning of surgical interventions, and postoperative control. MTEE with dynamic 3D reconstruction provides a superior insight to anatomy and function compared to plain TEE-2D echo, transthoracic 3D approach or TEE-3D reconstruction with parallel slicing probe in adolescents and adults with congenital heart disease. Enhanced qualitative and quantitative analysis is possible.

16

Pediatric three-dimensional echocardiography—comparison of left ventricular end-diastolic volume to magnetic resonance imaging in patients with compressed left ventricular geometry

Vangi V, Apfel HD, Gopal AS, Shen Z, Boxt LM, Reagan K, Katz J, Allan LD, Barst RJ, King DL
Columbia University, New York, USA

In patients with primary pulmonary hypertension (PPH), serial evaluation of left ventricular (LV) end-diastolic volume (EDV) may be helpful since severe PPH is often associated with marked LV compression by the right ventricle. Standard two-dimensional echocardiography is of limited use in determination of LV EDV in these patients since geometric and image plane positioning assumptions are invalid for compressed and distorted LV geometry. Volume determination by 3D echocardiography (3DE) has been validated in normal adults by magnetic resonance imaging (MRI). The purpose of this study was to compare LV EDV by 3DE to short-axis gradient reversal MRI in 20 children with PPH (median age 10.5 years) and severely distorted LV geometry. 3DE diastolic data sets were obtained by an acoustic spatial locator, a line of intersection display using 7-8 non-parallel, non-intersecting cross-sections and a polyhedral surface reconstruction algorithm. The data were analyzed by linear regression and Bland-Altman analysis. Correlation coefficient r =0.92. Regression equation: 3D EDV=0.76 MRI EDV + 4.8, SEE=10.5 ml. Bias (3DE-MRI)=-12.9 ml. Limits of Agreement=25.8 ml. We conclude that 1) in children with PPH, there is good correlation of LV EDV measurements by 3DE with MRI, and 2) Slight underestimation of LV EDV is present by 3DE relative to MRI. This implies that 3DE may be useful for serial determination of LV EDV when evaluating vasodilator therapy for PPH. Furthermore, 3DE also may be useful in evaluation of distorted ventricles in other forms of congenital and acquired heart disease.

17

Interventions and outcome in infants with d-TGA in the Baltimore-Washington Infant Study, 1981-1991

Lurito K, Brenner JI, Loffredo C, and the Baltimore-Washington Infant Study (BWIS) Group
Division of Pediatric Cardiology, University of Maryland School of Medicine, Baltimore, USA

To determine outcome variables and care patterns, records of infants enrolled in the BWIS with the diagnosis of d-transposition of the great arteries (d-TGA) from 1981-91 were reviewed. The impact of gestational age, birth weight, gender, race, associated anomalies (NCA), number of catheterizations/surgeries, year and type of repair, age/weight at repair and neurologic status on outcome were evaluated. Over a 10-year period, 207 infants with d-TGA were enrolled in the BWIS—simple d-TGA in 106 (51%), d-TGA+VSD in 38 (19%), complex d-TGA in 63 (31%). There was an excess of males (66.3%), whites (74.6%), full-term and large babies, compared to a birth cohort of infants with other CHD. NCA were noted in 11%. Follow-up data were gathered in 193 patients through chart review in 5 regional centers—duration of follow-up ranged from 1d-11yr 9 mo (mean 3 yr 8 mo). Four infants died prior to referral and 15 more before surgery. Atrial switch repair (AtS) was performed on 77, arterial switch operation (ASO) on 85, and Rastelli repair on 12. From 1981-85, 96% of repairs were AtS and from 1986-91, 69% were ASO. Average age and weight at repair for AtS was 5.9 mo and 5.7 kg, and for ASO was 19 d, 3.6 kg. AtS patients received more pre-repair caths, post-repair caths and total interventions than ASO patients ($p<0.05$), but did not differ in the number of surgeries. Even simple d-TGA infants undergoing AtS or ASO differed in pre-repair caths 1.44 vs 0.76, total caths 2.16 vs. 1.18, and total interventions 3.41 vs 2.31, but not in post-repair caths 0.82 vs 0.46, or surgeries 1.25 vs 1.13. Survival improved from AtS to ASO groups, 77 to 82%, and for era of repair, from 71 to 82% (1981-85 to 1986-91), p=NS. All-cause mortality was 29.5%. Although cardiac diagnosis was not a risk for death, associated NCA and abnormal neurologic status pre-repair were major risks for poor outcome.

18

Cardiorespiratory response to exercise after atrial correction for transposition of the great arteries (TGA)—role of right ventricular dysfunction

Douard H, Labbe L, Barat JL, Jiménez M, Broustet JP, Choussat A
Hopital Cardiologique, Centre Hospitalier du Haut-Leveque, Pessac, France

We studied 50 patients after atrial correction for TGA (42 Senning, 8 Mustard), mean age 12 years (37 boys, 13 girls) and 50 normal children of comparable sex, age and body surface area. All children have performed an exercise test (Bruce protocol) with gas exchange measurement. Only in the patient group, the right ventricular ejection fraction (RVEF) was calculated by radionuclide ventriculography (Te 99 m) at rest (50 patients) and during exercise (32 patients). RVEF was abnormal (<40%) at rest in 14%. During exercise, right ventricle response was abnormal (RVEF increased less than 5%) in 45% and the end-exercise value of RVEF was abnormal (<50%) in 34%. Exercise duration, maximal oxygen consumption (VO_2max), ventilatory aerobic threshold (VAT), maximal heart rate, systolic blood pressure were significantly lower in patient group ($p<0.001$). There was a relative correlation between end-exercise RVEF and VO_2 max (r=0.61) or VAT (r =0.6). In patients with abnormal exercise RVEF, exercise duration (9.7 vs 11.6 min, $p<0.05$), VO_2 max (27 vs 33 ml/kg/min, 63 vs 74% of maximal theoretic value, $p<0.05$), VAT (3.8 vs 5.2 min p=0.05) were lower. Age at surgery was higher (15 vs 9 months, $p<0.001$) in patients with abnormal resting RVEF. In conclusion, classic abnormal cardiorespiratory response to exercise after atrial correction for TGA seems to be related to right ventricular dysfunction.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

19

Apparent breath-holding spells associated with malignant hypervagotonia treated by permanent pacing

Porter CJ, McGoon MD, Espinosa RE, Osborn MJ, Hayes DL
Pediatric Cardiology, Mayo Clinic and Mayo Foundation, Rochester, USA

Breath-holding is thought to be a benign condition affecting several young children. However, from 1985-94, we evaluated 8 pediatric pts with apparent breath-holding symptoms associated with life-threatening bradycardia spells. All pts were treated with a permanent VVI pacemaker. Pts had onset of symptoms between 2 wks and 12 mos of age (median 7 mos) and presented to our institution at 9 mos to 5 yrs (median 14 mos). There were 7 females and 1 male. Symptoms included clonic/tonic activity (6 pts), cyanosis (4), prolonged apnea (3), status epilepticus (1), and 2 pts required CPR; duration of spells was 15 sec to 2 min. All episodes were provoked by anger or painful stimulus. Medications to prevent recurrent spells were tried in 6 pts—anticonvulsants (7 total), anticholinergics (3 pts), theophylline (2 pts). ECG monitoring during symptoms documented asystolic pauses of 1.7 to 24 sec (mean 12 sec; median 11.8 sec). Permanent VVI pacemakers were implanted at 10 mos to 5 yrs (median 14.5 mos)—6 endocardial, 1 epicardial. Three pts required revision of the pacemaker system. Pts have been followed for 1-112 mos (median 33). After pacemaker implant, 5 pts had immediate and complete resolution of their symptoms while the other 3 had only minor brief spells. All patients have normal development. We conclude that, 1) a small number of young children with apparent breath-holding symptoms have life-threatening bradycardia episodes; 2) Permanent pacemaker therapy for such pts is safe, efficacious and warranted.

20

Spontaneous closure of atrial septal defects

Helgason H, Jonsdottir G
Barnaspítali Hringsins, Landspitalinn, and University of Iceland Medical School, Reykjavik, Iceland

Atrial septal defects (ASD) are found more frequently in the pediatric population and improved diagnostic techniques with echocardiography (2DE) and Doppler facilitate diagnosis so that repair is possible at the optimal time. The purpose of our investigation is to study the size of ASDs at diagnosis and how size changes during follow-up and to explore the relationship between size at diagnosis and need for surgery. We reviewed the medical records of all patients (pts) diagnosed in Iceland with the diagnosis of ASD born in 1984-93. The ASD was confirmed by 2DE in all pts and smaller defects than 4 mm were excluded. The ASD size was measured by 2DE from subxyphoid long- and short-axes views. There were 87 pts, 26 male and 61 female. Four pts died from other causes than the heart defect and had not been operated. Seven pts with ASD primum and sinus venosus defects were excluded from analysis. There were insufficient data on one pt. Thus there were 29 pts with a 4 mm defect, 19 pts with 5-6 mm defects, 6 pts with 7-8 mm defects and 22 pts had defects greater than 8 mm. In the 4 mm group, 23 pts (79%), the ASD closed spontaneously or decreased in size, one pt has been operated. In the 5-6 mm group, 15 of 19 pts (79%) closed spontaneously; 2 pts (9.5%) have been operated. In the 7-8 mm group, 2 of 6 pts (33%) closed spontaneously and 3 have been operated. In the >8 mm group, one of 22 pts has closed spontaneously and 20 pts (91%) have been operated. We conclude that defects smaller than 6 mm in diameter are very likely to close spontaneously, although follow-up is necessary. Defects larger than 7 mm carry high probability of operative repair.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

21

Fifteen-year experience with Kawasaki disease

Fournier A, Marchand T, St-Cyr C, van Doesburg N, Davignon A
Ste-Justine Hospital, Montreal, Canada

Since 1979, 429 children (249 boys, 180 girls; 89% French Canadian origin) were diagnosed as having Kawasaki disease at Ste-Justine Hospital. Age distribution was as follows: 14% less than 1 year, 56% 1 to 5 years; 27% 5 to 10 years and 3% 10 years and older. An average of 24 patients/year were seen during "nonepidemic" years compared to 43 patients/year during 5 "epidemic" years ($p<0.05$). Overall incidence of coronary aneurysms was 16%, varying from 0 to 27% per year, the incidence being significantly higher in "epidemic" years (20%) compared to "non-epidemic" years (12%) ($p=0.01$). Incidence of aneurysms was significantly higher in patients (pts) less than 1 year compared (28%) to those 1 to 4 years old (12%) ($p=0.004$). There were 3 acute deaths, 2 of these in "epidemic" years, all 3 with coronary aneurysms. Two pts presented electrocardiographic signs of myocardial infarction. Patients were followed for a mean of 1.8 years (4 days to 12.5 years) with a longer follow-up when aneurysms were present, mean 2.6 years (17 days to 10.7 years). The majority of aneurysms (77%) regressed completely 1.2 years (mean) after diagnosis. No pts presented long-term complications of sudden death, myocardial infarction or angina pectoris. Overall, the yearly incidence of coronary aneurysms has not changed significantly since 1985—the year of introduction of gammaglobulins. However, only 12/164 pts (7%) developed aneurysms after gammaglobulins compared to 25/100 pts (25%) who did not receive gammaglobulins during the same period ($p=0.001$). This may reflect better echocardiographic imaging of those pts or modification in the virulence of the disease. Continuous analysis of epidemiologic data appears mandatory for a better understanding of Kawasaki disease.

22

Total cavopulmonary connection (TCPC)—intermediate results in patients less than three years of age

Kaulitz R, Ziemer G, Luhmer I, Kallfelz HC
Medical School, Hannover, Germany

From 9/88 to 12/93, 67 pts with functional univentricular heart (65% had abnormal AV-valves, systemic or pulmonary venous drainage) underwent TCPC for definitive palliation. Group I <3 yrs, 123 pts, age range 9-35 months, mean 19 months; group II >3 yrs, 44 pts, age range 3-18.3 yrs, mean 8.3 yrs. Preoperative hemodynamics (PA mean pressure, Rp, ventricular end-diastolic pressure, cardiac index) in group I were not significantly different from those in group II. One or more preoperative risk factors (Rp>3 U·m^2, PA mean pressure >15 mm Hg, ventricular end-diastolic pressure >10 mm Hg, AV-valve anomalies) were found in more than 60% in both groups. Operative variables including cardiopulmonary bypass time, aortic cross-clamp time were similar between both groups. In unfavorable preoperative hemodynamics and complicating intracardiac anatomy, fenestration of the intraatrial tunnel patch was performed in 6 pts of group I. The overall mortality in both groups did not differ significantly (I 2/23 (13%); II 4/44 (9%)). During the mean follow-up period of 28 and 41 months in groups I and II, respectively, one late death occurred in each group. Reoperation rate (15 vs 13%), late arrhythmias (10 vs 7%), postoperative AV-valve insufficiency (50 vs 43%) were observed with equal frequency. Postoperative hemodynamic studies up to 70 months did not reveal significant differences for mean RA (10 vs 11 mm Hg), ventricular end-diastolic pressure (4.8 vs 5 mm Hg) or cardiac index (3.4 vs 2.9 l/min/m^2). Age less than 3 years at TCPC is not a risk factor for successful outcome, in spite of a relatively high frequency of preoperative risk factors. Definitive palliation in early childhood may avoid potential deleterious effects of systemic ventricular volume and pressure overload.

23

Ross procedure in a young rheumatic population

Al-Halees Z, Kumar N, Galal OM, Gometza B, Gallo R, Duran CMG
King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia

Pulmonary autografts have shown promise in the intermediate and long-term results as being an ideal valve substitute for diseased aortic valve. In the period between January 1990 and July 1994, we have performed this procedure on 78 patients (pts); 56 (71.8%) males. Their age ranged from 1-41 years. The etiology was rheumatic in 62 (79.5%) pts and regurgitation was present in 60 (76.9%), stenosis in 5 (6.4%) and both in 13 (16.7%). Associated mitral repair was performed in 22 (28.2%) pts. The mean bypass time was 117.9 minutes and mean ischemic time 79.8 minutes. The right ventricular to pulmonary artery connection was established in all but one case with homografts. There have been no hospital deaths or documented thromboembolisms to date. There have been two (2.2%) late deaths, one unrelated and one due to endocarditis and systemic sepsis and renal shutdown at 12 and 36 months (mos), respectively. Five pts (6.4%) have been reoperated; one for mitral valve repair failure (6 mos) and 4 for autograft failure with progressive regurgitation (20-26 mos). One reported valve showed histology compatible with rheumatic valvulitis. Even though the left ventricular dimensions and function show immediate recovery and are maintained in the short follow-up, there is a small progression of the autograft regurgitation followed beyond two mos. The effect of sizing—surgical distortion, recurrence of rheumatic activity on these transplanted valves and of the repaired mitral valves—remain to be evaluated in the long-term. In conclusion, the Ross procedure, while being a safe and excellent option in the young growing pts with aortic valve disease, should be moderated by the concerns expressed above.

24

Assessment of pulmonary arterial growth after bidirectional superior cavopulmonary anastomosis

Slavik Z, Lamb RK, Horvath P, Webber SA, Keeton BR, Monro JL, Tax P, Reich O, Daubeney PEF, Skovranek J, Salmon AP
Wessex Cardiothoracic Centre, Southampton, UK and Kardiocentrum, Prague, Czech Republic

Pulmonary arterial growth following bidirectional superior cavopulmonary anastomosis (BCPA) may be expected but little data are available to support this assumption. Between 1987 and 1993, 74 children underwent BCPA in 2 centres. Twenty-six of these with right-sided BCPA have undergone postoperative angiography and are the subject of this study. The median age at BCPA was 22 months (range 3 weeks-108 months). The postoperative angiogram followed 1-65 months (median 11 months) after BCPA. Z-scores, which standardize pulmonary arterial diameter to body surface area, were calculated from pre- and postoperative measurements of the systolic diameter prior to the first branching point of the right and left pulmonary arteries. The mean pre- and postoperative body surface areas were 0.46 m^2 (SEM 0.04) and 0.64 m^2 (SEM 0.04; $p=0.0001$) respectively. The mean pre- and postoperative Z-scores of the right pulmonary artery were -1.00 (SEM 0.49) and -0.53 (SEM 0.35) respectively and of the left pulmonary artery 0.06 (SEM 0.41) and -0.14 (SEM 0.24). In the subset of 16 patients with reduced pulmonary blood flow (O_2 saturation <0.75) preoperatively, the small right pulmonary artery grew in excess of somatic growth (mean Z-score -2.48 pre- and -0.84 postoperatively; $p=0.004$). No difference in the growth of the pulmonary arteries was found in the 12 patients with and the 14 patients without competitive pulmonary blood flow. The 9 patients younger than 12 months at BCPA and the 9 patients followed longer than 15 months after BCPA demonstrated satisfactory growth of both pulmonary arteries. Our results show that normal short-term growth of both pulmonary arteries follows the right-sided BCPA and enhanced growth of a small right pulmonary artery can be achieved.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

25

Relation between cardiac anatomy and risk of the arterial switch operation in newborns with complete transposition of the great arteries

von Bernuth G, Däbritz S, Seghaye MC, Nitsch GB, Häusler M, Engelhardt W, Mühler E, Messmer BJ
Departments of Pediatric Cardiology and Thoracic and Cardiovascular Surgery, RWTH, Aachen, Germany

The aim of the study was to assess the influence of coronary anatomy and other features of cardiac anatomy on the risk of the arterial switch operation (ASO) in newborns with complete transposition of the great arteries (TGA). The operative reports of 193 consecutive newborns with TGA (Taussig-Bing complex excluded) presenting for arterial switch operation were analyzed with respect to coronary anatomy and other anatomical features such as ventricular septal defect (40 patients), coarctation of the aorta (11 patients), bicuspid pulmonary valve (4 patients), malalignment of aortic and pulmonary commissures (4 patients), and others. Nine (4.7%) of 191 patients died early after ASO while 2 patients, who survived, were aborted to an early Senning procedure. Three further patients who survived developed signs of myocardial ischemia immediately after ASO and required delayed sternal closure, reimplantation of a single coronary artery, and mammary artery bypass to the right coronary artery, respectively. Statistical analysis of the data showed that of all anatomical features, only a single right coronary artery (11 of 193 patients with 3 deaths and 2 nonlethal complications) increased the risk of ASO.

26

Allograft replacement of pulmonary valve following previous repair of tetralogy of Fallot

Sadiq M, Sreeram N, Stumper O, DeGiovanni J, Wright J, Silove E, Sethia B, Brawn W
Heart Unit, Birmingham Children's Hospital, Birmingham, UK

The aim of this study was to assess the intermediate-term results of pulmonary valve replacement (PVR) using allograft valves, following previous repair of tetralogy of Fallot (R-TOF). Over a 5-year period, 12 patients (8 male, 4 female) have undergone PVR using an aortic homograft (18-27 mm) at a median age of 145 (range 8-332) months. The age at R-TOF was 43.5 (3-75) months and interval from R-TOF to PVR was 96 (1-257) months. Indications for PVR were increasing pulmonary regurgitation assessed by color Doppler echocardiography and angiography (graded from 0 [no regurgitation] to 3 [severe]), right ventricular dysfunction, increasing right ventricular end-diastolic diameter (RVEDD) and decreasing exercise tolerance. Additional procedures at PVR included repair of pulmonary arterial branch stenoses (3) and closure of residual VSD (2). At a median follow-up of 28 (2-57) months, all patients showed improvement of symptoms and of exercise tolerance on formal testing (10). Pulmonary regurgitation decreased from grade 3 (1-3) to 0 (0-1) ($p<0.05$), with 11/12 pts showing a decrease by 2 grades. Concomitantly, RVEDD decreased from 3.55 (1.6-4.7) cm to 2.45 (1.2-3.3) cm ($p<0.05$), and tricuspid valve regurgitation (TR) improved by a median of 1 grade, with 8/12 patients having no TR at follow-up. Despite symptomatic improvement and decrease in pulmonary regurgitation, the 2 youngest patients (age at PVR 8 and 21 months; interval from R-TOF to PVR 1 and 18 months) continue to have persistent severe RV dysfunction, with no significant change in RVEDD or cardiothoracic ratio. Restoration of a competent pulmonary valve using allograft valves produces significant improvement of RV function and of symptoms in the majority of patients. Allograft failure has not yet occurred in any patient at intermediate-term follow-up. PVR is less satisfactory after R-TOF in infancy, if severe regurgitation develops <1 year post-repair.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

27

Inhaled nitric oxide after congenital heart surgery

Lorini FL, Crupi GC, Borghi A, Sonzogni V, Shori A
II° Servizio di Anestesia e Rianimazione, Divisone di Cardiologia e Cardiochirurgia, Ospedali Riuniti, Bergamo, Italy

We assessed whether inhaled nitric oxide (NO), an endothelium-derived relaxing factor, would produce selective pulmonary vasodilatation in pediatric patients with congenital heart disease and postoperative pulmonary hypertension. We determined in 13 pediatric patients during the postoperative period whether inhaling 10-20 ppm of NO would reduce the pulmonary pressure (PAP), pulmonary vascular resistance (PVR) and transpulmonary gradient without any systemic effect. We treated 8 patients with tricuspid atresia who underwent a Fontan procedures; 2 had repair of complete atrioventricular canal, 1 of ventricular septal defect, 1 mitral stenosis and the last patient underwent a correction of tetralogy of Fallot. Inhaled NO was started when PAP was >60% of systemic pressure in patients undergoing a biventricular repair or when transpulmonary gradient was >10 mm Hg in Fontan correction. All the patients were sedated, under neuromuscular relaxation and ventilated in order to maintain a $paCO_2$ <30 and >25 mm Hg. The inspired oxygen concentration (FiO_2) was 0.60. Inhaled NO decreased the pulmonary artery pressure from 46±5 to 29±3 mm Hg ($p<0.05$), the PVR decreased from 692±30 to 267±10 dyne•cm•sec^{-5} and in the Fontan procedures, decreased transpulmonary gradient from 10±2 to 6±1 mm Hg. Central venous pressure, heart rate and systemic pressures did not change after inhaled NO. We conclude that inhaled NO at low doses (10-20 ppm) proved to be an ideal selective pulmonary vasodilator after congenital heart correction. In our study, there were no systemic side effects. The high morbidity and mortality related to the acute pulmonary hypertensive state can be overcome by the use of inhaled nitric oxide.

28

Quantitation of morphologic severity of subaortic stenosis by three-dimensional echocardiography

Vogel M, Richter M, Ho SY, Anderson RH, Rigby M, Bühlmeyer K
Department of Pediatrics, Royal Brompton Hospital, London, UK and German Heart Center, Munich, Germany

Because of the complex contour of subaortic stenosis it has been difficult to assess severity by 2-dimensional (2-D) echocardiography. We assessed the morphologic severity of subaortic stenosis by 3-dimensional (3-D) echocardiography and compared it to measurements of diameters by 2-D echocardiography. Thirty-one patients, all with concordant AV and VA connections, and including 8 with VSD with a median age of 7.2 (0.4-18.2) years, were examined. The tomographic ultrasound probe is placed on the thorax and acquires perpendicular parallel images of the heart, steered by a stepper motor, which moves to plane in 0.5 mm steps. An image is acquired at each step with ECG- and respiratory-gating. Ninety parallel slices of the heart were obtained forming the 3D dataset. We measured the aortic valve orifice and subaortic diameters in peak mid-systole in the 2D cross-sectional long axis. Also in systole, we measured from the 3D-dataset in a short-axis cut perpendicular to the long axis of the area of the aortic valvar orifice and the narrowest subaortic area. Ratios of these valvar and subvalvar measurements were constructed and compared with systolic Doppler gradient across the subaortic stenosis. Average peak pressure gradient was 37 mm Hg and the peak gradient ranged between 10-100 mm Hg. The ratio of subaortic/aortic areas was 0.77 (range 0.37-0.98) and the ratio of subaortic/aortic diameter was 0.78 (range 0.53-0.95). In hearts with normal LV outflow tracts, the ratio of areas was 0.97 (range 0.95-0.98). Comparison between gradient and areas was r=0.90, with a standard error of 13 mm Hg, which is better than the correlation between gradient and diameter (r=0.82, with a standard error of 22 mm Hg). We conclude that 3D has an important role in the evaluation of subaortic stenosis.

29

The long-term prognosis of 81 patients with supravalvar aortic stenosis (1960-1991)

Kitchiner D, Jackson M, Walsh K, Peart I, Arnold R
Royal Liverpool Children's Hospital, Liverpool, UK

The prognosis of supravalvar aortic stenosis (SVAS) is unclear as previous studies have contained small numbers of patients, or included postoperative results only. Eighty-one patients with SVAS were followed for a median of 8.3 years (1-29 years)—40 patients (49.4%) had Williams syndrome, 18 (22.2%) had familial SVAS, 5 (6.2%) had other syndromes and 18 cases (22.2%) were sporadic. Nineteen patients (23.5%) had multilevel obstruction. Forty-seven patients (58%) underwent operation; the re-operation rate was 17%. Three patients underwent balloon dilatation. Twenty percent of patients underwent intervention within a year of presentation. Multivariant analysis predicted that 88% of patients would undergo intervention within 30 years of follow-up. The chance of intervention was increased by more severe aortic stenosis at presentation, the presence of multilevel obstruction (in patients with sporadic supravalvar aortic stenosis) and the presence of Noonan's syndrome. Three deaths occurred before operation and 13 within a month of operation. Predicted survival 30 years after presentation was 66%. Risk factors for survival were the age and severity of aortic stenosis at presentation, and the presence of Noonan's syndrome. Multilevel obstruction did not emerge as a significant risk factor due to the high association with the severity of stenosis at presentation. Seventy-three percent of survivors had mild or insignificant stenosis at follow-up. In conclusion, long-term survival is related to age and severity of aortic stenosis at presentation. Most patients will require intervention, and most survivors will have mild stenosis.

30

Conservative surgery for recurrent aortic and subaortic stenosis can avoid for a long time any type of valve replacement

Petit J, Houyel L, Planché C, Bruniaux J, Lacour-Gayet F, DeSouza-Uva M, Serraf A, Langlois J, Binet JP
Marie-Lannelongue Hospital, Le Plessis-Robinson, France

Recurrence of congenital valvar or subvalvar aortic stenosis (AS, SAS) has always been a challenging surgical problem. The use of a mechanical prosthesis is problematic in children (small aortic annulus, anticoagulant therapy). Bioprosthetic valves have a strong tendency to calcify in children. The Ross operation, now largely advocated, carries an uncertain prognosis, as well for the transplanted pulmonary valve as for the pulmonary homograft. Among 443 consecutive patients (pts) with AS or SAS operated on by the same surgical team between January 1978 and December 1993, 46 needed reoperation—elective conservative surgery in 24 (Group 1), aortic valve replacement in 22 (Group 2). In Group 1, mean age at reoperation was 9 years (2 months-25 years). Mean delay from the first operation was 6.2 years (1 month-13 years). Cardiac anomaly was AS in 11, SAS in 7, combined AS+SAS in 6. Thirteen pts had associated anomalies (ventricular septal defect in 4, coarctation of the aorta in 7, mitral stenosis in 2, pulmonary stenosis in 2) repaired before or during the first operation. Surgery included relief of all types of AS or SAS—aortic commissurotomy in 14, myotomy in 10, membranectomy in 6. Maximal gradient decreased from 92 to 41 mm Hg on average ($p<0.001$). Early mortality was 4.2% in Group 1 (1 of 24), 22.7% in Group 2 (5 of 22), 5% in pts operated only once ($p=0.005$). There was no late death. Heart block occurred in 1 pt in Group 1, 2 pts in Group 2. With a mean follow-up duration of 7.3 years, only 3 pts in Group 1 had a third operation—aortic valve replacement with mechanical prosthesis in 2, conservative surgery in 1. This last patient had 2 previous operations for SAS with residual gradients of 90 and 120 mm Hg. The third operation (myotomy + membranectomy) reduced the gradient from 170 to 30 mm Hg. In conclusion, despite the great interest of Ross operation, conservative surgery for recurrent AS and SAS is often possible with good results.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

31

Ross procedure in children

Bourlon F, Quaegebeur J, Lecompte Y, Dor V
Centre Cardio-Thoracique, Monaco and Columbia Presbyterian Medical Center, New York, USA

To August 1994, 35 children (26 male, 9 female) underwent a Ross operation. Age at operation ranged from 2 weeks to 16 years (mean age 7.5±5.3 years). The preoperative indications are stenotic lesions in 29, incompetence in 5 and mixed lesions in 1. Previous interventions in LVOT were found in 45—surgical valvotomy in 18 cases, percutaneous valvotomy in 10 cases, resection of sub-aortic stenosis in 12, Konno procedure in 2, repair of aortic arch in 2 and repair of VSD and aortic incompetence in 1. In some children, concomitant procedures were performed such as mitral valve repair, COA repair, pulmonary artery banding. Only 4 patients had no previous intervention; 17 patients had 1, 9 patients had 2, 4 patients had 3 and 1 patient had 4 previous operations. In addition to the Ross operation, the LVOT was enlarged with a pericardial patch in 3 and with a septal incision using the RV muscle of the autograft to increase the septum in 7. In the follow-up study using echo Doppler, there was a gradient inferior to 10 mm Hg in 3. One patient was reoperated for allograft replacement. On the LVOT, the post-op gradient was inferior to 10 mm Hg in 28 and between 10 and 30 mm Hg in 3. There was no aortic incompetence in 10 patients, trace in 9 patients, mild in 11 and moderate in 2. Serial echo measurements were obtained in 24 patients. An increase of the neoaortic root was present in all patients except one. In 8 patients, the autograft became larger than expected for their BSA, suggesting an element of dilation. In conclusion, the Ross procedure provides excellent and safe relief of complex left ventricular outflow tract obstructions in children, but the long term of the allograft in the right ventricular outflow tract remains uncertain. Growth of the autograft at the level of the annulus is proportional to somatic growth in most children. There appears to be a degree of dilation in the sinus portion of the autograft.

32

Congenital mitral valve disease in aortic arch anomalies

Macedo AJ, Oosthoek PW, Wenink ACG
Department of Anatomy, Leiden, The Netherlands

Sixty specimens, 34 (56%) with isolated aortic coarctation (AoCo), 20 (33%) with AoCo and VSD and 6 (10%) with interrupted Ao arch (IAoA), were studied for mitral valve (MV) and left ventricle (LV) pathology, being compared with a control group of 20 normal hearts. All had concordant AV and VA connections and normal LV length for age. Mean age at death was 27 months and 55% were boys. Five groups (G) could be found: G I, 18 cases (30%), with normal morphology and normal MV annulus diameter (MVd) corrected for tricuspid valve annulus d (mean 0.82 vs 0.88 for the controls). All the others (70%) had a small MVd (mean 0.60) or abnormal MV morphology. In G II, 10 cases (17%), MV was small (MV d=0.52) but grossly normal, being compressed by unusually enlarged LV musculature. Malattachment of MV (G III, 17, 28%) occurred more to the interventricular septum (10) than to the lateral wall. Four (G IV, 7%) had dysplastic MV. In G V, 11 cases (18%), asymmetric MV morphology was found, being true parachute in 3. By types of AoA anomalies, MV pathology occurred in 78% of AoCo cases; the malattached MV occurred more (12/17) in isolated AoCo, the parachute type only in AoCo with VSD and 4/6 of AoA cases had normal MV. LV outflow tract obstruction (OTO) occurred in 9 (15%), 4 of muscular type, and in 3 because of dysplasia and malattachment in association. During life, diagnosis of MV stenosis was made in 9 cases. We conclude that MV congenital pathology occurs in 70% of AoA anomalies, mainly in AoCo (78%). Probably, the hemodynamics imposed by the distal LV obstruction and the presence of VSD, the type of MV pathology found, with a small dysplastic group, and that of LVOTO mechanisms, make these specific anomalies particularly difficult to diagnose during life.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

33

Aortic coarctation with hypoplastic arch in neonates—which surgical treatment?

Gargiulo G, Albanese SB, Nicolini F, Pierangeli A, Santorelli MC, Donti A, Prandstraller D, Bonvicini M, Picchio FM
Departments of Cardiac Surgery, Anesthesiology and Pediatric Cardiology, University Hospital, Bologna, Italy

The optimal surgical approach for patients with coarctaction and hypoplastic aortic arch remains still controversial. Between January 1988 and September 1994, 55 patients have been operated on in our institution (<3 months old) using a surgical approach based on the echocardiographic and angiographic findings. According to the location, extension and size of the hypoplasia of the aortic arch, we have treated three groups of patients—group A (32 pts) with resection and a normal or extended end-to-end anastomosis; group B (7 pts) with resection, posterior end-to-end anastomosis and anterior subclavian flap enlargement; group C (16 pts) with direct side-to-end anastomosis between ascending and descending aorta through a median sternotomy. Three (18.7%) patients died during the postoperative course in group C. With a mean follow-up time of 40 months we had 6 cases (16.6%) of residual or recurrent coarctation in group A and 1 (6.5%) cases in group C—successfully repaired at 2 months of age by anterior approach in 2 pts and by percutaneous angioplasty in the others. In conclusion, hypoplastic aortic arch in neonates represents a common difficulty; optimal reconstruction of the entire aortic arch is mandatory to reduce operative mortality and incidence of recoarctation, especially when complex associated intracardiac lesions or left ventricular dysfunction are present.

34

Subvalvular left ventricular outflow tract obstruction—10 years of echocardiographic experience

Milanesi O, Stellin G, Secchieri S, Svaluto Moreolo G, Daliento L, Antonio Azzi W, Rubino M, Zacchello F
Departments of Paediatrics, Cardiac Surgery and Cardiology, University of Padova, Padova, Italy

The aim of this study is to assess 1) anatomical types of subvalvular left ventricular outflow tract obstruction (LVOTO), 2) structures involved in the LVOTO, 3) percentage of associated lesions which were detected by 2-D-echo and Doppler in a pediatric population between September 1984 and September 1994. Among 25,485 echo examinations performed, diagnosis of subvalvular LVOTO recurred 380 times (1.5%) in 126 pts. The obstruction was caused by a fibrous ring in 59 pts (39%, Group (Gr) A) while in 30 pts a fibrous ridge attached to the inferior rim of a conoventricular VSD was present (Gr B). Ten pts (8%) had a fibromuscular tunnel (Gr C). In 3 cases (2%), a posterior deviation of the infundibular septum creating a malalignment VSD was the cause of the obstruction (Gr D). A pure muscular LVOTO was present in 13 pts (10%, Gr E). This was due to IHSS in 6 pts and to biventricular hypertrophic cardiomyopathy in 7 pts. In Gr A and B, the incidence of associated cardiac lesions was 35 and 33%, respectively, with a prevalence of left heart involvement in Gr A (MV dysplasia + Ao Coarct 16%, Ao Coarct alone 7%, others 12%), and of the right heart in Gr B (subpulmonary and/or pulmonary valve stenosis 26%). The associated anomalies rate in Gr C was as high as 70% (VSD + Ao Coarct 40%, MV dysplasia+ Ao Coarct 20%, MV dysplasia + TOF 10%) and was 66% in Gr D (Ao Coarct in all). In comparing echo diagnoses and surgical reports in 45 operated pts (1990-94), we have found 1 false negative and no false positives. We conclude that 1) subvalvular LVOTO is caused by many different anatomical structures whose precise identification is essential for a proper surgical planning; 2) MV valve dysplasia or abnormal MV septal attachment is a frequent associated lesion and contributes to LVOTO in many cases; and 3) 2-D-echo and Doppler is a highly sensitive and specific tool in evaluating LVOTO.

35

Hypertrophic cardiomyopathy—recent genetic advances

McKenna WJ, Goodwin JF
St George's Hospital Medical School, London, UK

Recent years have seen major advances in the understanding of the molecular genetics of hypertrophic cardiomyopathy. Missense mutations in the DNA encoding beta cardiac myosin heavy chain cause disease and to date different mutations originating in the head rod region of the myosin gene have been identified in over 30 families. DNA testing for pre-clinical diagnosis has been applied and the development of a DNA diagnostic test is in progress. Preliminary observations suggest that mutations which result in a change in electron charge of the encoded amino acid will be associated with significantly worse prognosis. Myosin mutations cause disease in between 30-50% of HCM patients. Recently we have identified mutations in cardiac troponin-T and cardiac alpha tropomyosin and these appear to account for approximately 20% of disease. The recognition that HCM is a disease of sarcomeric contractile proteins represents an early stage in the understanding of this condition at a molecular level. Identification of other responsible gene(s) and full assessment of the phenotype/genotype relationship may ultimately lead to a new molecular classification of HCM which will aid in the major problems which clinicians face in diagnosis and management, particularly in relation to sudden death.

36

Clinical course and prognosis of hypertrophic cardiomyopathy diagnosed in pediatric age at a non-referral center

Donti A, Rapezzi C, Negrini E, Prandstraller D, Bonvicini M, Picchio FM, Magnani B
Section of Pediatric Cardiology, Institute of Cardiology, University of Bologna, Bologna, Italy

Hypertrophic cardiomyopathy (HCMP) in pediatric age has been investigated mainly at referral institutions where bias in patient selection is possible. We studied clinical the course and prognosis of an unselected group of pediatric pts with HCMP diagnosed at our (non-referral) institution. Between 1974 and 1993, 211 HCMP pts were examined, 24 (11%) aged ≤14 yrs at first observation (range, 5 mo to 14 yrs). Nine (37%) were obstructive forms; a family history of HCMP was evident in 8 (33%) and only 1 pt had syncope. During follow-up (7.5±4.5 yrs), there were 6 cardiac deaths, sudden and unexpected (SD) in 4 and due to congestive heart failure (CHF) in 2. Two other pts received heart transplantation. The annual cardiac mortality rate was significantly higher among pediatric pts (4.1 vs 2.1% $p<0.05$). In the 4 pts with refractory CHF, the left ventricle (LV) underwent progressive dilatation and thinning. In each of these cases, the LV posterior wall was initially thicker than the interventricular septum (inverted asymmetrical septal hypertrophy. In conclusion, HCMP diagnosed in pediatric age has a high mortality rate (≥4%/yr); also, at non-referral centers, both SD and CHF leading to death or heart transplantation are frequent; inverted asymmetrical septal hypertrophy is an incremental risk factor for subsequent LV dilatation and thinning.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

37

Mitochondrial respiratory chain defects are a frequent cause of isolated hypertrophic myocardiopathy in childhood

Sidi D, Bonnet D, Piéchaud JF, Iserin L, Le Bidois J, Rustin P, Munnich A, Kachaner J
Cardiologie Pediatrique et INSERM U393, Hopital Necker Enfants Malades, Paris, France

Disorders of cardiac energy metabolism can account for any clinical presenting symptom, especially for cardiomyopathy. Owing to the particular genetic origin of mitochondrial enzymes, we hypothesized that disorders of mitochondrial oxidative phosphorylation (MOP) limited to myocardium could be responsible for isolated hypertrophic cardiomyopathy (HCM) as this category of cardiomyopathy is a common clinical feature of mitochondriopathies. We performed endomyocardial biopsies in 32 patients with HCM (2 Noonan syndromes, 4 familial autosomal dominant HCM, 26 "idiopathic" HCM) and investigated respiratory chain activity in myocardium and other tissues (measurement of oxygen consumption by isolated mitochondria and enzymes activities by spectrophotometric studies). In 16/32 (50%), MOP was normal particularly in Noonan syndrome and in familial HCM. MOP defects were identified in the remaining 16, i.e. in 16/26 "idiopathic" HCM. These defects were limited to the myocardium in 8/16, while 5 had altered enzymes activities in skeletal muscle without symptoms of skeletal myopathy, and 3 died without extracardiac evaluation. Conversely, myocardial MOP was normal in 4 patients with mitochondrial disease and multiorgan involvement and no HCM. In conclusion, MOP disorders account for 50% of HCM in childhood. Moreover, endomyocardial biopsy is the only tool available to assess myocardial specific involvement in 25% of HCM. Further investigations on mitochondrial genome in affected individuals will hopefully give evidences for molecular bases in this category of HCM.

38

Myocardial carnitine metabolism in congenital heart defects

Kreuder J, Bauer J, Paul T, Holtkamp F, Bode S, Lampert F
Children's Hospital, University of Giessen and Medical School, Hannover, Germany

Free carnitine (FC) is the essential carrier for transmembranous mitochondrial transport and oxidation of long-chain fatty acids (acyl-CoA). Increased myocardial long-chain acylcarnitine (LCAC) has been reported to be harmful as it induces intracellular Ca^{2+} accumulation. This study examined myocardial carnitine content in 65 children with congenital heart defects undergoing cardiac surgery. FC, LCAC and short-chain acylcarnitine (SCAC) were determined radioenzymatically; ATP was measured by luminometry. Carnitine palmitoyltransferase 2 (CPT) mRNA was determined by a competitive quantitative PCR assay. Right ventricular (RV) myocardium (n=15) showed higher levels of FC (2.13±1.16 vs 3.46±2.12 µMol/gm non-collagen-protein; $p<0.05$), SCAC (2.12±1.59 vs 3.34±1.4 µMol/gm NCP, $p<0.01$) and LCAC (0.21±0.15 vs 0.59±0.3 µMol/gm NCP; $p<0.01$) than right atrial (RA) myocardium (n=50). CPT expression in RV was 3 times higher than in RA. Higher RV end-diastolic pressure was associated with decreased ATP ($r=-0.6$; $p<0.1$) and increased LCAC ($r=0.63$; $p<0.1$) levels. RA pressure >5 mm Hg was related to a higher LCAC/FC ratio ($p<0.05$). Myocardial ischemia due to aortic clamping induced significant reduction of FC (-28%), SCAC (-35%) and ATP (-60%) in RA (n=50). Lower preoperative oxygen saturation significantly correlated with a more pronounced loss of FC ($p<0.01$) and an increase in LCAC/FC ($p<0.005$) in the postischemic RA myocardium. FC reduction ($p<0.05$) and LCAC/FC increase ($p<0.1$) were associated with higher doses of postoperative inotropic support. These findings suggest that the myocardial carnitine status in children with congenital heart defects is related to mechanical load and postoperative myocardial performance.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

39

Diagnosis and follow-up of acute myocarditis in pediatric age—role of magnetic resonance versus endomyocardial biopsy

Gagliardi MG, Polletta B, Bevilacqua M, Di Renzi P, Marino B
Cardiologia Pediatrica, Ospedale Bambino Gesù, Roma, Italy

Criteria for diagnosis of myocarditis (MYO), including the ones of endomyocardial biopsy (EMB), are controversial. Even if EMB is already considered the "gold standard," magnetic resonance imaging (MRI) was suggested to be a valid non-invasive tool for the diagnosis of MYO. We report our experience with MRI in the diagnosis and control of a large group of pts with MYO after immunosuppresive therapy. From January 1990 to June 1994, 60 pts (age range 6 mos-10 yrs) affected by dilated cardiomyopathies were submitted to endomyocardial biopsy (EMB) and MRI. EMB was performed following the "Dallas criteria" for the diagnosis of MYO and MRI parameters applied for tissue characterization were the signal intensity obtained in T1 spin-echo sequences proton density and T2 weighted spin-echo sequences (TE 90-110-130 ms). The EMB demonstrated in 47 pts MYO, and in 13 pts idiopathic dilated cardiomyopathy (IDC). MRI showed the same results (sensitivity 100%, specificity 100%). The 47 patients with MYO treated by immunosuppression repeated EMB and MRI after 6 and 12 months. EMB showed in 34 pts persistence of MYO, in 12 a resolving MYO and in 1 a resolved MYO. MRI presented a signal intensity increase in 28/34 pts with persistent MYO and in 10/12 pts with resolving MYO. In the 8/47 pts (sensitivity 85%) in which the signal intensity did not increase, the EMB demonstrated myocardial fibrosis and 5/6 pts presented normal left ventricular function. We conclude that 1) at the first diagnosis MRI shows the same results of EMB (sensitivity 100%, specificity 100%); 2) MRI repeated after 6 and 12 months presents a lower sensitivity (85%). This latter finding might be due to the misleading of the inflammatory process by EMB. In fact, the diagnosis of MYO by EMB is based just on sample specimens of right ventricle, while MRI estimates the signal intensity on the entire left ventricle.

40

Stenting of pulmonary arteries stenosis—hemodynamic, angiographic and lung perfusion scanning results

Spadoni I, Giusti S, Bertolaccini P, Redaelli S, Borghi A, Carminati M
Department of Cardiology, Ospedale Pediatrico Apuano, Department of Nuclear Medicine, Ospedale Civile, Massa and Department of Cardiology, Ospedali Riuniti, Bergamo, Italy

Twenty patients (pts) underwent endovascular stenting for pulmonary artery (PA) stenosis after lung perfusion scanning and angiographic evaluation. Mean age of pts was 13±6 years (from 5 to 24) and mean weight 38±19 kg (from 12 to 74). Twenty-seven stents were implanted, 12 in right PA and 15 in the left, in 3 pts stenting of both PA was performed. Six pts were recatheterized 5-18 months (mean 10) after treatment. There were no major complications. Hemodynamic and angiographic results: PA diameter increased from 5.5±2 to 11.2±3.3 mm ($p<0.001$). Systolic gradient across stenosis dropped from 41±21 to 16±15 mm Hg ($p<0.001$), systolic PA pressure from 57±21 to 47±20 mm Hg ($p<0.005$) and systolic PA/systemic pressure ratio from 0.56±0.19 to 0.42±0.18 ($p<0.005$). No change in hemodynamic and angiographic results was demonstrated in the 6 pts recatheterized. Lung perfusion scanning results: all pts had reperfusion of defects after treatment. In 10 pts with unilateral stenosis, the flow ratio to the affected and non-affected lung changed from 0.38±0.22 to 0.76±0.22 ($p=0.005$). In 10 pts numerical indices of flow per unit of lung volume were obtained, and improvement in distribution of flow to both lungs was demonstrated. Our results indicate that PA stenting is an effective mode of treatment for peripheral PA stenosis. Lung perfusion scanning is the technique of choice for functional evaluation of PA stenosis and control of results after stent implantation.

41

Pulmonary artery stents in children less than three years of age—early results and intermediate follow-up

O'Sullivan JJ, Lock JE, Perry SB
Department of Cardiology, Children's Hospital and Harvard Medical School, Boston, USA

Endovascular stents have been used with success in the treatment of branch pulmonary artery (PA) stenosis in older children. We investigated the feasibility of placing such stents in children <3 years of age. Twenty-two children had a total of 28 stents placed in the branch pulmonary arteries between April 1991 and October 1993. Age ranged from 1 to 34 months (median 16 months) and weight from 2.7 to 17 kg (median 9.6 kg). The general indication for stent placement was a hemodynamically significant branch PA stenosis which persisted following balloon dilation. A significant increase in the diameter of the stenotic area was achieved in all cases (mean±SD; pre 3±1.1 mm and post 6.3±1.9 mm; $p<0.001$). In 2 patients the proximal portion of an articulated stent detached and embolized to the main PA. Seven lobar vessels were covered by a stent with diminution of flow in 2 cases. In one of these there was no antegrade flow in the covered vessel 10 months later. Angiographic improvement was maintained at follow-up of between 5 and 25 months. The stent was redilated after an interval of between 5 and 25 months in 10 patients achieving an increase in diameter of between 0.6 to 3 mm. Transcatheter placement of stents is technically feasible in small children. Clinical and angiographic improvement is maintained in the intermediate term, but the success of redilation more than 24 months post-implantation remains to be determined.

42

Laser valvotomy followed by balloon valvoplasty for pulmonary atresia with intact ventricular septum—four-year experience

Gibbs JL, Parsons JM, Dickinson DF, Wren C, Ladusans E, Chatrath RR, Blackburn ME
Killingbeck Hospital, Leeds, UK

Since 1991 laser valvotomy followed by balloon dilatation has been the primary treatment for pulmonary atresia with intact ventricular septum and tripartite right ventricle in our centre. The procedure was attempted in 7 infants aged 1-70 (median 6.5) days with weight of 2.1-4.7 (mean 3.5) kg. Patient selection was based upon the presence of a tripartite right ventricle with no more than minor hypoplasia of the tricuspid valve. A 4-5 Fr Cobra catheter was positioned immediately below the imperforate pulmonary valve. The orientation of the catheter tip was adjusted with the aid of hand injections of contrast and a frozen lateral angiogram showing the pulmonary trunk opacified via the duct. The valve was perforated using a Trimedyne 0.018" laser guide wire with 13 continuous wave laser firings of 3-5 W of 3-5 sec duration. The laser wire or an 0.018" FlexT wire was passed into the right pulmonary artery or the descending aorta via the duct. The valve was dilated with a 3-3.5 Fr coronary angioplasty balloon of 2-4 mm diameter and then by a 5-8 mm balloon. Valvotomy failed in only one patient due to laser breakdown. Transient SVT was the only complication. Prostaglandin E was stopped immediately in all but one patient who remained duct-dependent for 49 days. Right ventricular outflow (RVOT) velocity after duct closure was 2.0-3.8 (mean 2.9) m/sec and O_2 saturation was 70-90% (mean 77). Two patients had a repeat balloon dilatation at 22 and 116 days post laser. At latest follow-up 7-1720 (median 112) days after valvotomy, RVOT velocities were 2.0-4.5 (median 3.2) m/sec and O_2 saturation ranged from 70 to 97%. One child of 4 years is awaiting surgery for relief of infundibular stenosis and closure of ASD and one aged 4 months is awaiting repeat balloon dilatation. Laser valvotomy and balloon dilatation is safe and is an effective alternative to surgical pulmonary valvotomy in these patients.

43

Role of interlocking detachable coils in embolization of coronary artery fistulas

Qureshi SA, Reidy J, Alwi M, Lim MK, Wong J, Tay J, Baker E, Tynan M Guy's Hospital, London, UK, Instut Jantung Negara, Kuala Lumpur, Malaysia, National University of Singapore, Singapore

Interlocking detachable coils (IDC) are new controlled-release unfibered coils, which are available in lengths of 1-30 cm and diameters 2-8 mm. Thirteen patients underwent transcatheter embolization of congenital coronary artery fistulas (CAVF) using IDC and conventional fibered coils. Their ages at embolization ranged between 1.8 and 17 yrs (mean 6.8 yrs) and their weights between 9 and 54 kg (mean 23.5 kg). One pt (1.8 yrs) was on diuretic treatment for heart failure. Five of the fistulas originated from right coronary, 5 from left anterior descending and 3 from circumflex coronary arteries. The drainage was to right atrium (RA) in 7 and right ventricle in 6 pts. Four pts had two arterial feeders. Retrograde arterial approach was used in 11 pts. Through 5 or 6 Fr guiding catheters, Tracker-18 catheter was passed over 0.014" guide wire to the site of embolization. In 8 pts, successful embolization was achieved with IDCs of 2-8 mm diameters and lengths of 1-20 cm. In the other 5, because of large fistulas, fiberd coils of 8-15 mm diameter were combined with IDCs to achieve occlusion. Complications occurred in 3 pts. In 1, an IDC coil embolized to RA and was retrieved with a snare. In 2 other pts, 1 fibered coil was retrieved from aortic bifurcation and 2 fibered coils from left pulmonary artery. The fistulas were completely occluded in 12 pts, with small residual flow in 1. IDC coils are invaluable in transcatheter occlusion of CAVF. If misplaced, they can be easily retrieved. In patients with high-flow large fistulas, fibered coils may need to be placed before IDC coils to achieve satisfactory occlusion.

44

Balloon angioplasty of the pulmonary artery after arterial switch operation

Hutter PA, Beekman RP, Hoorntje TM, Harinck E, Meijboom EJ Divisions of Pediatric Cardiology, Wilhelmina Children's Hospital, University of Utrecht and University of Maastricht, Utrecht and Maastricht, The Netherlands

Seventy-six patients were followed after arterial switch operation for transposition of the great arteries. Mean follow-up was 8.0 yrs (2.3-16.2 yrs). Twenty-one patients required reintervention for pulmonary stenosis (PS). Mean reintervention-free interval was 11.2 yrs (95% CI 9.6-12.9 yrs). Patients were selected for cardiac catheterization based on hypertrophy on ECG, Doppler gradient >50 mm Hg, and/or >60% reduction in pulmonary artery diameter on MRI. Nine pts had balloon angioplasty a total of 13 times—9 as first reintervention and 4 as second reintervention. Seven of 9 successful as first reintervention, 2 of 4 successful as second reintervention. Twelve pts had surgical angioplasty a total of 18 times—12 as first reintervention and 6 as second reintervention. Seven of 12 successful as first reintervention, 3 of 6 successful as second reintervention and the other three with partial reduction in gradient only. PS located distal in the pulmonary trunk or at the bifurcation was successfully treated with balloon dilatation in 5 out of 7 attempts. Hypoplasia of pulmonary artery branches in two patients necessitated a staged approach dilatation with a view to later stent placement. A third patient has complete occlusion of the right pulmonary artery. Surgery, most often transanular patching, was necessary whenever PS was located proximally, with distortion of the pulmonary valve. In PS after ASO three groups exist. In distal suture-line stenosis balloon dilation is the re-intervention of first choice. In hypoplasia of pulmonary branches it is the only option. In the group with proximal stenosis with valve distortion balloon dilatation is seldom effective. In the selection of these patients MRI plays a decisive role, as echo visualization of localization and severity of stenosis is increasingly difficult in older patients.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

45

Transcatheter closure of the moderate sized patent ductus arteriosus—comparison between Rashkind occluder and single or multiple Gianturco coils

Galal OM, de Moor M, Al-Fadley F, Hijazi ZM
King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia and Department of Pediatrics, New England Medical Center, Tufts University School of Medicine, Boston, USA

Ten consecutive patients (pts) median age 27 months (12-96 months), who weighed 11.9±5 kg (6.3-23 kg) underwent anterograde transcatheter occlusion of their patent ductus arteriosus (PDA) using one or multiple Gianturco coils (coil group). The results in the coil group were compared with an age and weight matched group of ten pts who underwent PDA occlusion previously at KFSH using the double umbrella device. The narrowest diameter of the PDA measured 3.4±1.6 mm (1-6 mm) in the coil group and 2.72±0.8 mm (1.7-4.3) [p=n.s.] in the device group. The fluoroscopy time in the coil group was significantly higher (35±35 min, range 13-131 min) than in the device group (15±11, range 7-43 min; $p<0.01$). Nine out of 10 (90%) pts in the coil group were discharged with total occlusion as demonstrated by echocardiography, whereas only 5/10 (50%) in the device group were completely occluded. These apparent differences did not reach statistical significance in the these small group of pts. In the coil group, there was one embolization of a coil which was retrieved with a snare catheter, and one patient needed streptokinase therapy overnight because an occluded left femoral artery. In the device group, there was one embolization which was retrieved with a snare catheter, after which another device was deployed successfully. A different device patient had to be treated overnight with heparin for femoral artery occlusion. Thus, the complication rate was similar. Anterograde deployment of singular or multiple Gianturco coils to occlude small or large PDA (up to 6 mm) is safe and effective. Moreover, it is less expensive than the umbrella technique.

46

Alternative uses of the Rashkind umbrella device in congenital and post surgical cardiovascular lesions

Tabatabaei H, Nykanen DG, Williams WG, Freedom RM, Benson LN
The Hospital for Sick Children, Toronto, Canada

Implantation of the Rashkind umbrella device for closure of patent ductus arteriosus (PDA) is now a routine procedure in many units. Alternative application to non-PDA positions has been attempted less frequently, often employing a modification to the delivery system. A retrospective review of all such transcatheter occlusions was undertaken to determine feasibility and efficacy. From 9/88 through 9/94, implantation of the occluder was attempted in 22 patients (8 female, 14 male) with congenital or postsurgical cardiac lesions other than a PDA. Median age at implantation was 2.5 years (17 months-12 years) and weight 12 (4-45) kg. The lesions included a ventricular septal defect (4), aortopulmonary window (1), aortopulmonary collateral (1), atrial septal defect (1), Gortex shunt (4), and residual left persistent superior vena cava (SVC) to right atrium (RA) following Glenn operation (1). Ten patients post Fontan operation underwent occlusion of an interatrial shunt (5), SVC to right or left atrial shunt (3), LV to RA (1) or ventricle to pulmonary artery (1) communications. Transcatheter implantation was accomplished in all without morbidity utilizing a 12 mm (12) or 17 mm (10) diameter device. In 19 patients (86%), loading and delivery of the device was modified to allow the use of a smaller delivery system (6 and 8 Fr for 12 mm and 17 mm devices respectively). Surgical removal of the occluder was required in 1 patient 4 weeks post-implantation. At latest follow-up (1-66 months, median 4) color Doppler study revealed complete occlusion in 17 patients (77%). Modification of the delivery system allows application of the umbrella device for safe and effective transcatheter occlusion of many unusual intracardiac and vascular communications, supplementing or avoiding the need for additional surgery.

47

Intimal thickening in the coronary arteries of children—its relation to atherosclerosis

Pesonen E, Kaprio J, Martımo P, Ruismäki A
The Children's Hospital, Department of Pediatrics, University of Helsinki, Finland

Newborn children have significant intimal thickenings in their coronary arteries. Their magnitude increases with age; they are bigger in males than in females, and they are directly correlated to the coronary artery disease mortality of the grandparents. In the present study of semiserial cross sections of the coronary arteries of 71 autopsied children, we have found that intimal thickenings are related to local thinning of media in the same fashion as media is thinner under the atherosclerotic plaques. They appear in the beginning of the coronary arteries and are larger in the left as compared to the right coronary artery. A three-dimensional model of serial cross sections of the coronary arteries shows the inner surface three-dimensionally through the outer surface. In this model, a narrowing of the lumen can be demonstrated. In a sagittal longitudinal cross section along the course of the coronary artery, a hypothetical coronary "angiocardiogram" demonstrates a narrowing in the beginning of the coronary artery. These narrowings have similar distribution as later atherosclerosis and are related to risk factors for coronary artery disease.

48

Anatomic repair of corrected transposition or atrioventricular discordance—report of five cases

Metras D, Kreitmann B, Wernert F, Voisin M, Riberi A, Pannetier A, Silicani MA
La Timone Children's Hospital, Marseille, France

It is now well recognized that a right ventricle may eventually fail if working as systemic. This has led to the switch operation for TGA but more recently to anatomic correction of corrected transposition or AV discordance combining an atrial switch and a ventricular outflow repair (arterial switch or Rastelli type). We report here 5 cases of such anatomic correction of AV discordance in patients from 3 months to 7 years. Three had situs solitus, 2 situs inversus. All patients had a VSD. Three had an increased pulmonary flow; two had a decreased pulmonary flow with pulmonary stenosis and atresia. Three had discordant, one had concordant ventriculoarterial relations, and one had DORV. Two had an associated coarctation. All had undergone one or more previous operations. Correction was done between 3 months and 7 years of age. All had a Senning procedure (twice left-sided). Ventriculoarterial outflows were repaired—3 VSD closures, 2 LV to aorta rerouting, 2 arterial switches, 1 RVOT repair and 1 Rastelli. A tricuspid annuloplasty was done once. All patients survived the operation (2 had a successful reoperation—late obstruction of pulmonary venous channel in a left-sided Senning and a residual VSD closure in one). Mean follow-up is 20 months (4-100). Three patients are in sinus rhythm; two have intermittent junctional rhythm. All have a normal LV function as evidenced by echocardiography. Despite a more demanding procedure, the anatomic repair of hearts with AV discordance is possible, even in infancy, with good early results. Despite the potential late rhythmic problems of the Senning operation, with increasing experience, it may become the procedure of choice.

49

Surgical results after bidirectional Glenn procedure in the presence of forward flow from the ventricles

Uemura H, Yagihara T, Kawashima Y, Anderson RH
National Cardiovascular Center, Osaka, Japan and the National Heart & Lung Institute, London, UK

Persistent pulsatile pulmonary arterial flow along with the maintenance of an appropriate amount of total pulmonary perfusion may assist growth of the pulmonary arteries. Twenty-seven patients considered unsuitable for either biventricular repair or a Fontan type procedure have had a bidirectional Glenn procedure in the presence of forward flow from the ventricles to the pulmonary arteries. Forward flow from the ventricles was maintained through either the pulmonary trunk in 22 or a systemic-to-pulmonary shunt in 5. Banding of the pulmonary trunk and/or pulmonary valvotomy were performed as necessary to regulate pulmonary blood flow. Surgery was performed at the age of 0.8-22.3 (6.2±5.2) years, with one operative mortality due to arrhythmia. Nine patients have successfully undergone total cavopulmonary connection 2.7±1.9 years after this procedure. Preoperative and postoperative cardiac catheterization revealed changes in arterial oxygen saturation (75±11 vs 83±7%, $p<0.001$) and end-diastolic volume of the systemic ventricles (238±92 vs 188±97% of the expected normal volume, $p<0.01$), while no difference was detected in the mean cross-sectional area of the right and left pulmonary arteries compared with the expected normal value for the right pulmonary artery (76±21 vs 81±20%), nor in ventricular ejection fraction (53±8 to 50±14%). The maintenance of forward flow from the ventricle is, therefore, the option of choice when performing a bidirectional Glenn procedure so as to protect against regression of pulmonary arterial size, as well as off-loading the ventricles and improving arterial oxygen saturation.

50

Thoracoscopic interruption of patent ductus arteriosus in premature infants

Kececioglu D, Forster R, Gehrmann J, Vogt J, Kotthoff S, Scheld HH
Westfälische Wilhelms Universität, Münster, Germany

Patent ductus arteriosus (PDA) is a problem frequently encountered in the treatment of premature infants. These infants are at significantly high risk. Therefore a minimally invasive surgical procedure is needed to reduce the risks associated with open thoracotomy and surgical closure. The aim of this study was to assess the feasibility and safety of the thoracoscopic closure of the PDA using a clip applier. In 7 premature infants (weight 750-1200 gm; 25-30 weeks of gestation), closure of the hemodynamically significant PDA was performed in the pediatric intensive care unit by a thoracoscopic technique using an endoclip applier. In 3/7 pts the attempted thoracoscopic closure was not feasible because of massive pleural effusion and an open procedure was used. In 4/7 pts, the PDA was closed by the thoracoscopic technique, the procedure required 30 min in the mean. Echocardiograms, performed 6 and 12 months postoperative confirmed the complete closure of the PDA. In conclusion, the thoracoscopic closure of the PDA is feasible even in premature infants with very low birth weight. As a minimally invasive procedure, it offers several benefits—the overall length of the incision is reduced, the ribs are not spread and the musculature of the chest is not interrupted, thus preventing an injury to the shoulder mechanism.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

51

Pediatric heart transplantation—experience with FK506 and CyA

Welz A, Netz H, Böhm D, Schmitz C, Meiser B, Reichart B
Department of Pediatric Cardiology and Cardiac Surgery, University of Munich, Munich, Germany

The aim of this study was to analyze the immunosuppressive efficacy of FK506 as compared to CyA. Twenty-three patients (0.1-18 years) underwent cardiac transplantation. Eight patients were treated with FK506 (group I), 15 patients (group II) received CyA in addition to azathioprin and prednisone. The primary pathology was congenital heart disease (3 gr I, 5 gr II) and dilating cardiomyopathy (3 gr I, 10 gr II). Two patients in group II received a cardiac retransplantation; one of them was on a Berlin Heart Assist Device preoperatively. Two children in group II were on Novacor Left Heart Assist. All patients of group I survived the first 4 weeks, whereas 3 patients in group II died (primary graft failure 2, cerebellar infarction 1). One neonate in group I died on day 59 during a diagnostic intervention in an external hospital; one patient of group II died from cytomegaly virus pneumonitis (day 67). No acute rejection episodes were observed with therapeutic serum levels (10-15 ng/ml) of FK506. One adolescent revealed two rejections (IB and IIA) while FK506 levels were below 5 ng/ml. In contrast, acute rejections were more frequent ($p<0.5$) in group II (1.5/pt 3 month). One patient died after one year due to rejection. The rate of infections was not different in both groups. Actuarial survival for 2 years is 83% (gr I) and 68% (gr II). Regarding acute rejection episodes, FK506 was clearly superior to CyA. This is a major advantage especially in the pediatric patients with the well known difficulties in the diagnosis of acute rejections.

53

Chronotropic response after the Senning operation for transposition of the great arteries (TGA)

Labbe L, Douard H, Clementy J, Srour S, Broustet J-P, Choussat A
Hopital Cardiologique, Centre Hospitalier du Haut-Leveque, Pessac, France

We studied 42 patients (31 boys, 11 girls), 6.5 to 15 years (mean 11 years) after the Senning operation and 42 normal children of comparable sex, age, size, weight and body surface area. All children underwent graded exercise testing on a treadmill with respiratory gas exchange measure: maximal oxygen consumption (VO_2 max); ventilatory aerobic threshold (VAT), exercise duration, maximal heart rate (MHR) were determined. All children were in stable sinus rhythm at the end of the exercise and the metabolic chronotropic relation slope (S) as defined by WILKOFF was analyzed but with real oxygen consumption (METS). We confirmed reduced exercise tolerance (10.5±2 vs 13.2±2 min), VAT (4.5±1 vs 5.3±1 min), VO_2 max (33±5 vs 44±6 ml/kg/min) and MHR (168±18 vs 189±17 bpm) in the patient group ($p<0.001$). There was also an abnormal chronotropic response to exercise: S value 0.62±0.19 in patient group vs 0.77±0.14 in control group ($p<0.001$). We found no correlation between exercise duration, VAT, VO_2 max and S value ($r=0.16$, NS) or MHR ($r=0.14$, NS). In conclusion, abnormal cardiorespiratory and chronotropic response is observed after the Senning operation for TGA but the chronotropic insufficiency does not influence exercise performance in these patients.

52

Outcome of congenital complete heart block diagnosed *in utero*

Rosenthal E, Groves A, Allan LD, Qureshi SA, Baker EJ, Tynan M, Sharland GK
Fetal and Paediatric Cardiology, Guy's Hospital, London, UK

Complete heart block is now reliably diagnosed *in utero*. The prognosis is better in the absence of structural heart disease. Little is known, however, about the prognostic value of the presenting fetal heart rate. We have observed the outcome of a consecutive series of 36 fetuses with complete heart block and structurally normal hearts diagnosed *in utero* between 1980-1993. Presenting fetal heart rate, change in fetal heart rate and development of fetal hydrops were documented. The maternal anti-Ro antibody status was determined in 34/36 pregnancies and was positive in 32. Significant and progressive hydrops developed in 12 fetuses. Pregnancy was terminated in 2 of these and a further 6 died *in utero*. Two fetuses were delivered prematurely because of worsening hydrops but both died in the neonatal period from cardiac and renal failure despite effective pacing in 1. Two fetuses with hydrops recovered, 1 after a course of maternal sympathomimetic treatment. One non-hydropic fetus died suddenly *in utero* (mother anti-Ro negative). Heart rate at presentation ranged between 45-80 beats per minute (bpm). A fall in heart rate with advancing gestation was detected in 9/25 fetuses who had more than one examination; of these 5 died *in utero*. The presenting heart rate was related to the outcome as follows: 7 pts with ≤50 bpm: 4 died, 2 alive with pacemaker, 1 alive with no pacemaker. Nineteen pts with 51-60 bpm: 7 died, 6 alive with pacemaker, 6 alive with no pacemaker. Ten pts with >60 bpm: 1 died, 3 alive with pacemaker, 6 alive with no pacemaker. One non-paced infant died from pertussis. Of the 24 survivors, 11 patients (46%) have required pacing with follow-up ranging from 1-13 years. One patient whose mother was anti-Ro negative developed a cardiomyopathy despite pacing and underwent transplantation. Isolated complete heart block does not always have a good prognosis. Hydrops carries a poor prognosis. A heart rate <50 bpm at presentation, or a falling heart rate, dictate the need for careful evaluation of cardiac function to guide possible intervention and a guarded prognosis.

P 01

Linkage analysis as an additional test to diagnose long QT syndrome in persons with borderline QTc

Pulles-Heintzberger CFM, Gorgels APM, Wellens HJJ, van den Berg MH, Meijer H, Schaap C
Departments of Pediatric Cardiology, Cardiology, Clinical Genetics and Biochemistry, Academic Hospital, Maastricht, The Netherlands

The long QT or Romano-Ward syndrome (LQTS) is an autosomal dominant inherited disorder. Linkage analysis has demonstrated genetic heterogeneity, with a major locus on chromosome 11p15.5 (H-Ras oncogene). In a family with 2 cases of sudden death because of the LQTS, 4 generations were examined. Clinical diagnosis was based on QTc >440 msec on ECG at rest. A value of <420 msec was considered to be normal. LQTS was diagnosed in 12 of 25 relatives (60%). The two cases of sudden death occurred at the age of 32 and 24 years, and 4 others had clinical symptoms. Five individuals had a normal QTc, 5 were between 420 and 440 msec, and in 2, no ECG was available. To get better insight in the presence of LQTS in the borderline QTc group, linkage analysis was performed in 28 of the 32 family members using the markers pUC EJ6.6, H19, TH-PCR (Southern Blotting). The results yielded a maximum combined LOD score of 10 at a recombination fraction of 0.044. This confirms a positive linkage in this family with chromosome 11p15.5. The persons with normal or borderline QTc did have the low risk allele configuration. However, 1 girl with borderline QTc showed a crossing over. Because clinical diagnosis for this girl is yet uncertain, no further determination of the risk locus for LQTS is possible at this moment. In conclusion, linkage analysis is helpful to diagnose LQTS in individuals with uncertain clinical diagnosis from families with the LQTS.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 02

Comparison of inhaled nitric oxide, oxygen and prostacyclin as vasodilators in pulmonary hypertension

Turanlahti M, Laitinen P, Pesonen E
Children's Hospital, University of Helsinki, Helsinki, Finland

The aim of our study was to measure the vasodilating effect of inhaled nitric oxide, oxygen with and without NO and the combination of oxygen with intravenous prostacyclin in pediatric patients with pulmonary hypertension. One of the theoretical advantages of inhaled NO compared with intravenous PGI_2 is its selectivity in pulmonary circulation. During cardiac catheterization, we investigated the hemodynamic changes caused by 1) inhaled NO (20, 40 and 80 ppm); 2) pure oxygen; 3) O_2 and NO (20-80 ppm) and 4) O_2 and iv PGI_2(10 and 20 ng/kg/min). In 10 intubated anesthetized patients (age 3 mos-13 yrs) with primary (n=1) or secondary pulmonary hypertension (n=9), we measured the individual oxygen consumption with metabolic monitor (Deltatrac, Datex, Finland). The pressure in the pulmonary and systemic arteries was measured and the vascular resistance was calculated using the Fick principle.

	Percent change from baseline				Percent change from FiO_2 1 0				
	NO			O_2	NO + O_2			PGI_2 + O_2	
	20	40	80	100	20	40	80	10	20
PAP (mean)	-6 4	-12 8	-5 0	3.6	-5 4	-9.9	-7.0	-8 0	-12 0
PVRI	-17 9	-13 6	14 3	-38 2	32 9	-3 3	-20.3	1 0	-14 3
SVRI	-2 2	-0 2	-4.7	-1 4	2.9	-1 3	-6 7	-25 3	-35 6

We conclude that inhaled nitric oxide in low concentration (40 ppm) is a selective, effective and safe vasodilator in the testing of reactivity of pulmonary circulation in children with congenital heart defect and pulmonary hypertension. The maximal effect in the PVRI was seen by combination of oxygen and inhaled nitric oxide of 80 ppm ($p<0.05$) both of which can be used in postoperative pulmonary hypertension.

P 03

Transthoracic three-dimensional echocardiography for the diagnosis of site, extent and dynamic morphology of subvalvar left ventricular outflow tract obstruction in complete transposition

Vogel M, Ho SY, Basso C, Rigby M, Anderson RH
Department of Paediatrics, Royal Brompton Hospital, London, UK

We have assessed, whether three-dimensional echocardiography (3D-echo) yields additional information over 2-dimensional echo or angiography when evaluating the subvalvar left ventricular outflow tract in complete transposition studying 9 patients aged 1 day to 1.2 years with transposition and subpulmonary stenosis. The tomographic transducer was positioned on the chest. It is steered by a stepper motor, which moves the probe in 0.5 mm steps on the thorax perpendicular to the heart and acquires an image of the heart at each step with EKG and respiration-gating. The slices thus acquired form a 3-dimensional data cube, which then can be "sectioned" in any plane. Conventional imaging planes like long-axis and four-chamber views were reconstructed in three dimensions as well as new views, such as looking downwards through the pulmonary valve or upwards from the LV apex towards the LV outflow, permitting display of the obstruction as seen from above or below. The subpulmonary stenosis was shown to be due to posterior deviation of the outlet septum (4), fibrous tissue shelves (2), accessory mitral valve tissue (2) or a combination of the latter two (1). New information was provided in respect to information concerning the insertions of the mitral valve tension apparatus on the septum, in the extent and location of deviation of the outlet septum, and with regards to the recognition of accessory mitral valvar tissue. 3D-echo findings have been confirmed in 5/9 patients who have thus far undergone open heart surgery. We conclude that transthoracic 3D-echo yields important new information on the anatomy of subvalvar LV outflow obstruction in complete transposition, and is invaluable in planning therapeutic interventions.

Notes

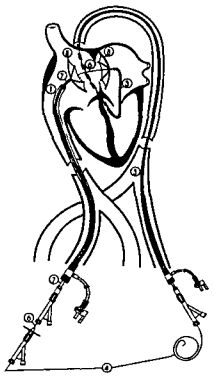
https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 04

A new system for closure of atrial septal defects and large arterial ducts

Babic U, Sievert H, Franke A, Grabitz R, von Bernuth G
Bethanien Hospital, Frankfurt and the Departments of Cardiology and Pediatric Cardiology, University of Aachen, Aachen, Germany

Transcatheter closure of cardiac defects with currently available systems has many limitations. To improve the procedural safety, implant centering and fixation and to avoid residual shunting, a new system was developed, manufactured by Dr. Osypka (GMBH, Germany).The delivery system consists of 10 or 11 Fr venous introducer delivery sheath (1), a long Nitinol wire with a conus (4), torquer catheter (7), and a transseptal metal cannula (6). The prosthesis consists of two umbrellas (8) made of Nitinol wire skeleton and microporous polyurethane. After creation of a venoarterial long wire track, the umbrellas were individually placed over the wire into the atria (for ASD) or pulmonary artery/aorta (for PDA) and, after positioning with metal cannula, were screwed together at the defect/duct level. The procedure was guided by selective right- and left-sided angiography and/or TEE. Patient selection for ASD closure can substantially be improved by 3-D reconstructed echocardiography. Animal experiments showed an excellent incorporation of the umbrellas into the atrial septum 3 months post-implantation. Experience with initial clinical application for ASD and PDA closure with this system will be presented.



P 05

Techniques for transcatheter closure of ventricular septal defects

Rigby ML, Redington AN
Royal Brompton Hospital, London, UK

When patients undergo transcatheter umbrella closure of a ventricular septal defect (VSD), the most important aspect of the procedure is to pass an end-hole catheter across the defect; this is then replaced with an exchange wire over which is passed a long transeptal sheath through which the double umbrella is maneuvered into position. There are 4 potential routes for crossing a VSD: when the catheter is passed from the right to the left ventricle, it can be via a femoral vein or right internal jugular vein; when the catheter is passed from the left to the right ventricle, it can be via a femoral artery or via a femoral vein, with the catheter crossing the atrial septum to the left atrium and then into the left ventricle. When the femoral artery is used, it enables an exchange guide wire to form a loop from the femoral artery to the femoral or internal jugular vein. A transeptal sheath can then be advanced from a vein across the ventricular septal defect. Thirty-four patients have undergone attempted transcatheter closure of a VSD with a modified Rashkind ductal umbrella—17 with a perimembranous defect (group I), 8 with a muscular defect (group II) and 9 with a residual defect following cardiac surgery (group III). We have performed a retrospective review of the technique used successfully to pass the transeptal sheath across the VSD. In group I, the defect was crossed directly from the femoral vein in 16 and by an arteriovenous loop in one. In group II, the defect was crossed directly from the femoral vein in 1, from the right internal jugular vein in 6 and from the femoral vein via the left atrium in 1. In group III, 1 residual perimembranous defect was crossed from the femoral vein; 6 residual muscular defects were entered via the right internal jugular vein (1 patient required an arteriovenous loop). One defect could not be crossed. We conclude that umbrella closure of a VSD can usually be performed from a direct venous approach. For perimembranous defects and defects close to the central fibrous body, the femoral vein should be used. For most muscular defects, the right internal jugular vein is more appropriate, although passing the sheath from the femoral vein via the left atrium is an alternative approach.

P 06

Occlusion of ventricular septal defects by the buttoned device—early international experience

Sideris E, Walsh K, Chen C-R, Losay J
Athenian Institute of Pediatric Cardiology, Athens, Greece

Ventricular septal defect (VSD) occlusion was performed in eight patients using the 4th generation adjustable buttoned device. Six of the defects were congenital and two post-infarction. The size of the congenital defects varied between four and seven mm; the post-infarction VSDs had diameters of 22 and 25 mm respectively. The age of the congenital VSD patients varied between 7 and 22 years; the post-infarction patients were over 70 years old. Four of the congenital defects were perimembranous and two were muscular. All congenital defects were crossed from the left side but repaired from the right side using 7-8 Fr long sheaths. Devices of 20-25 mm were delivered directly or over a wire connecting the femoral artery with the femoral vein. All congenital VSD procedures ended in full occlusions, in contrast with the post-infarction VSDs, which had partial occlusions with symptomatic improvement. No complications were noticed. The buttoned device appears promising in the occlusion of perimembranous as well as muscular VSDs. Perimembranous defects can be effectively and safely repaired in older children or adults. Further trials are justified.

P 07

Successful management of postoperative junctional ectopic tachycardia in infants by early moderate by hypothermia

Pfammatter JP, Paul T, Kallfelz HC
Pediatric Cardiology, Hannover Medical School, Hannover, Germany

Junctional ectopic tachycardia (JET) is a severe complication in the early postoperative course after open heart surgery for congenital heart disease. Because of the high heart rate, the atrioventricular dissociation and the insufficient response to conventional antiarrhythmic drugs, it is associated with considerable morbidity and mortality. Between June 1991 and June 1994, 6 consecutive infants who developed JET after corrective surgery for heart disease were treated with body surface hypothermia. By placing ice bags onto the child's surface, a rectal temperature between 32 and 34 °C was achieved in all patients. Cooling of the children resulted in a significant decrease in the ventricular rate of the tachycardia from 219±27 to 165±25 bpm (mean±SD) within 4 hours after initiation of hypothermia. In 3 patients with signs of low cardiac output, decrease of the tachycardia rate due to cooling resulted in restoration of stable hemodynamics. Hypothermia was maintained without complications for a period of between 24-88 hours (mean 59). Patients were rewarmed once they were in a stable hemodynamic condition for at least 24 hours. Sinus rhythm reoccurred spontaneously after a mean of 5.5 days. All patients survived the tachycardia, and after a follow-up period of 16 months (mean), are in sinus rhythm and do not show late sequelae of hypothermia. Body surface hypothermia is a safe and efficient treatment of postoperative JET.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 08

Balloon valvuloplasty in newborns with critical aortic valve stenosis

Tax P, Reich O, Skovránek, Tuma S, Marek J, Samánek M
Kardiocentrum, University Hospital Motol, Prague, Czech Republic

From March 1987 to October 1994, percutaneous balloon valvuloplasty (VPL) was performed in 140 children with aortic stenosis (AS), including 37 newborns (NB) with critical AS. In 3 NB (8%), the stenotic valve could not be crossed and they are not included. In 34 NB, the VPL was performed at a median age of 4 (range 10-36) days. The body weight ranged from 2 to 4.3 (3.2±0.6) kg. Aortic annulus diameter was 3.8 to 8 (6.1±1.1) mm and balloon-to-annulus ratio ranged from 0.79 to 1.25 (1.03±0.10). Hospital mortality rate was 21%, including a NB who died after subsequent surgical intervention. In survivors, the left ventricular (LV) shortening fraction increased from 0.26±0.12 to 0.43±0.09 ($p<0.001$) and LV systolic pressure decreased from 131±42 to 97±20 mm Hg ($p<0.001$). AS peak gradient decreased from 56.7±31.5 to 39.4±15.9 mm Hg ($p<0.005$). Mean follow-up period was 2.5 years, ranging up to 7 years. Late mortality was 17%. Over-all mortality was 38% and actuarial survival at 7 years was 55%. None of the NB with annulus diameter less then 5.2 mm or body weight less than 2.8 kg survived. At least one of those risk factors was present in 9 (69%) of 13 non-survivors. Five patients required repeated VPL. Thirty-seven percent of patients are expected to reach age of 6.5 years free of repeated VPL or surgery. In conclusion, VPL can be performed in more than 90% of NB with critical AS. The procedure carries the highest risk in NB with body weight <2.8 kg and/or aortic valve diameter <5.2 mm. VPL improves myocardial function.

P 09

Aortic balloon valvuloplasty is as safe and effective as surgical aortic valvotomy in children with aortic valve stenosis

Kitchiner D, Bundred P, Walsh K, Peart I, Arnold R
Royal Liverpool Children's Hospital and Department of Primary Care, University of Liverpool, Liverpool, UK

The aim of the study was to compare the re-intervention rate and survival in patients who have undergone aortic balloon valvuloplasty (BV) or surgical aortic valvotomy (AV) for aortic valve stenosis (AVS). One hundred-sixty-one patients underwent intervention for AVS. Patients with critical aortic stenosis were excluded. Initial intervention was BV in 46 patients and surgery in 115 patients, at a median age of 7.7 years (yrs) and 9.7 yrs respectively. The median duration of follow-up in survivors was 5 yrs (range 0-20). Ten patients (22%) who initially had BV and 31 patients (30%) who initially had AV, underwent re-intervention after a median interval of 8 months (mos) and 64 mos respectively; 51% of re-interventions were BV. There was no difference in the median age at first procedure of those who did and did not undergo re-intervention. No patient who initially had BV died or had aortic valve replacement (AVR), but one patient required heart transplant for myocardial failure. Seventeen patients who initially had AV subsequently had AVR (after a median duration of 5.1 yrs). Ten patients died after AV (early=4; late=6). Statistical analysis (Wilcoxon & log-rank tests) showed that survival and re-intervention rates were not significantly different between patients who initially underwent BV or AV ($p>0.1$). In conclusion, aortic balloon valvuloplasty offers palliation which is as safe and effective as aortic valvotomy in the treatment of aortic valve stenosis in children. Balloon valvuloplasty can also be more easily repeated for residual or recurrent stenosis.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 10

Stent implantation in aortic coarctation

Qureshi SA, Tynan M, Rosenthal E, Brzezinska-Rajszys G
Guy's Hospital, London, UK and Child Health Centre, Warsaw, Poland

Three patients had Palmaz-Schatz stents implanted for aortic coarctation (2 postoperative). Their ages ranged between 14-29 years. A 14-yr-old girl had had a subclavian flap repair at 2 yrs of age, poor result from balloon dilation of recoarctation at 9 yrs and patch aortoplasty at 10 yrs of age. She had an 8 mm conduit inserted between left carotid artery and descending aorta for residual gradient 3 weeks later. She needed captopril, nifedipine and atenolol for hypertension until a stent was implanted at site of recoarctation 4 yrs later. A 17-yr-old boy had a Waterston shunt for tetralogy of Fallot at 9 months, correction at 4 yrs of age, and 8 yrs later, hypertension due to coarctation with a right arch and an atretic left arch was noted. A 12 mm conduit was inserted between the ascending and descending aorta. He needed captopril, nifedipine and atenolol until stent implantation at 17 yrs of age. The 29-yr-old man had had coarctation repair at 6 months and aortic valvotomy at 3 yrs of age. Patch aortoplasty was performed for recoarctation 7 yrs later and aortic valve replacement with Starr-Edwards valve at 20 yrs of age. He had two balloon dilations for recoarctation at age 23 and 27 years with moderate result. Because of hypertension, he was maintained on nifedipine until stent implantation. In all 3 pts, stents of 3-4 cm length were implanted via femoral artery. Stents were dilated to 12 mm in 2 pts and 20 mm proximally and 14 mm distally in the third. The gradient was abolished in all 3. Hemorrhage at the puncture site was the only complication in 1. Follow-up was between 1-8 months. Anti-hypertensive medication has been reduced in 2 patients and maintained in 1. In older patients with aortic coarctation resistant to conventional treatment, stent implantation is an effective alternative.

P 11

Surgical preconditioning and transcatheter completion of univentricular repair—a new approach for high-risk patients

Schneider M, Konertz W, Hausdorf G
Charité, Pediatric Cardiology and Cardiac Surgery, Berlin, Germany

A new two-stage approach for univentricular repair of high risk patients is reported, using a combined surgical-transcatheter approach. At the first stage, a hemifontan procedure is performed surgically, but the communication between the superior vena cava and the right atrium is not ligated, but narrow banded. Additionally, a multiperforated baffle is inserted into the right atrium. Two months after surgical preconditioning, interventional completion is performed by balloon dilating the banding of the superior vena cava and either inserting a covered wallstent as intracardiac conduit between the superior and inferior vena cava or inserting a stent into the banding and occluding the multiperforated baffle using umbrellas. This new technique was successfully applied with no mortality in 5 patients and a mean pulmonary pressure above 18 mm Hg. The right atrial pressure dropped to 3-9 mm Hg. No complications occurred within the follow-up period of 3 to 6 months. This new approach allows a staged univentricular repair without the need for a surgical completion, thus being a promising approach for high-risk patients.

P 12

Transcatheter coil occlusion of the arterial duct—early results

Rosenthal E, Qureshi SA, Reidy J, Baker EJ, Tynan M
Department of Paediatric Cardiology, Guy's Hospital, London, UK

Transcatheter occlusion of the arterial duct with the Rashkind double umbrella is widely used and is the method of choice in many centres. The sizes of the catheter delivery system and of the device limit its use. Implantation into small ducts (<2 mm) can be difficult. A persistent leak is present in 10-20% of patients after umbrella closure. We attempted coil implantation into the arterial duct in 10 patients aged 1.5 to 14.5 (median 3) years and weighing 9.1-43.7 (median 13) kg. In 8 patients, the narrowest duct diameter was <2 mm. In 1, a leak persisted after previous umbrella closure. Standard radiological coils of 4 different types were chosen according to the duct morphology: Gianturco (Cook) 0.038" coils, platinum 0.018" coils (Cook or Target), Jackson controlled release 0.038" coils (Cook) or interlocking detachable 0.018" coils (Target). Coils were positioned inside the duct in the long tubular ducts or straddling the duct (with some loops in the pulmonary artery and some in the aortic ampulla). All coils were implanted using 5 Fr catheters via an arterial approach. In those with a duct <2 mm, the duct was occluded at the end of the procedure in 2 patients, by the following day in a further 4 patients, and by 3 months in 1. In 1 patient, it was not possible to position a coil successfully. The coil embolized and was retrieved. The duct was ligated at the time of surgical repair for recoarctation. In the patient with a leak following umbrella implantation, flow was reduced but still present on color flow Doppler the following day (follow-up only 2 weeks). In the patient with a 3 mm duct, a 5 mm diameter coil was implanted initially. This embolized to the pulmonary artery, was retrieved and replaced with an 8 mm coil. Persistent flow across this coil caused hemolysis. The coil was removed 2 days later and a Rashkind umbrella implanted with resolution of the hemolysis. Occlusion of small ducts is readily accomplished using coils selected according to the duct anatomy. This has both cost and practical benefit. It may have a role in leaks following implantation of a Rashkind umbrella. Standard coils, however, may not be suitable for larger ducts.

P 13

Dobutamine stress echocardiography in children and young adults

de Wolf D, Verhaaren H, de Craene T, Matthijs D
AZ.VUB, Brussels and UZ.RUG, Ghent, Belgium

Dobutamine stress echocardiography is widely accepted in the evaluation of regional wall motion disturbances in adult patients with coronary artery disease. We assessed the feasibility of dobutamine stress echocardiography in 30 children and young adults aged 6 to 26 years. The subjects or their parents agreed to participate as a control group in order to be compared with a group of patients who completed anthracycline chemotherapy. Dobutamine was given in 3 periods with a perfusion rate of 0.5 μg/kg/min, 2.5 μg/kg/min and 5 μg/kg/min respectively, each with a duration of at least 10 minutes. Attention was focused on both systolic and diastolic functions, rather than wall motion. A paired-t test showed significant differences between rest values and values at 5 μg/kg/min for VmaxAo (1.27 to 1.74 cm/sec), mean velocity of Ao (0.83 to 1.1 cm/sec), Ao acceleration time (AT 0.09 to 0.07 sec), AT/ET (0.32 to 0.28), blood pressure (107 to 128 mm Hg), E/A ratio (2.0 to 2.7), E wave (1 to 1.3 cm/sec), EF (63 to 80%), FS (34 to 50%), systolic wall stress (64 to 37 gm/cm^2), VcFc (velocity of circumferential fiber shortening corrected for heart rate 1.1 to 1.8 c/sec), wall thickness in systole (1.1 to 1.5 cm) and percentage of wall thickening (82-159%). No differences were found for heart rate, deceleration time of the E wave and A wave. Analysis of these results showed that dobutamine at a rate of 5 μg/kg/min had a positive inotropic effect, decreased afterload and influenced ventricular relaxation. There was no chronotropic effect and ventricular compliance was not altered. No side effects were noted. In conclusion, dobutamine stress echocardiography is rather simple to perform in children without serious adverse effects. The dobutamine stress echocardiography takes into account the inotropic state of the myocardium. Interpretation of results is straightforward as dobutamine at a rate of 5 μg/kg/min has limited hemodynamic effects, a positive inotropism, a decrease in afterload and alterations in relaxation. The test results are not influenced by individual variables as training, motivation and ventilatory efficiency.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 14

Myocardial hypoxemia detection by PET in children with valvular aortic stenosis

Strengers JLM, Pruim J, Blanksma PK, deJong R, Vaalburg W, Kuipers JRG
Beatrix Children's and University Hospital, Groningen, The Netherlands

Myocardial hypoxemia in children with valvular aortic stenosis is the indication for intervention. Assessment of myocardial perfusion (MP) and perfusion reserve (MPR) by PET might be a better indicator for myocardial hypoxemia than currently applied diagnostic modalities. Fifteen children (14 boys, 1 girl, 8-14 yrs) were studied by ECG, Holter, bicycle-ergometry and echo. By (N-13) ammonia PET, MPrest, MP after dipyridamole stress test (dst) and MPR were assessed. MPR was calculated as MPdst-MPrest/MPdst•100. These values were compared with those of normal adults. According to standard echo and electrocardiographic criteria, patients were divided into three groups: mild stenosis (echo-assessed pressure drop <50 mm Hg), moderate stenosis (50-75 mm Hg pressure drop, without signs of hypoxemia on ECG), severe stenosis (>75 mm Hg or with hypoxemia). PET data were normal in all cases with mild or moderate stenosis. In the patients with severe stenosis, at least MPrest, MPdst or MPR was abnormal. In 2 patients, an increased MPrest and MPdst with a normal MPR indicated an adequate compensation of myocardial perfusion to the increased pressure load. In 2 patients, an increased MPrest with a decreased MPR might indicate a failing compensation during dst and myocardial hypoxemia, as maximum oxygen delivery to the heart is mainly flow related. We conclude that assessment of MP and MPR in children with valvular aortic stenosis might be a more sensitive tool in the recognition of myocardial hypoxemia than currently applied non-invasive diagnostic modalities.

P 15

Air travel in grown-ups with cyanotic congenital heart disease

Harinck E, Hutter PA, Hoorntje TM, Simons M, Benatar AA, de Bruijn D, Fischer JC, Meijboom EJ
Wilheimina Children's Hospital, Utrecht, University Hospital Maastricht, Netherlands Aerospace Medical Centre, Soesterberg, The Netherlands

In-flight atmospheric conditions in commercial jet aircraft approach altitude equivalents of 6000 to 8000 ft (1829 to 2438 m). In healthy people, this causes a marked decrease in arterial pO_2, but only a mild reduction of the arterial oxygen saturation (SaO_2). Concern has been expressed that such cabin altitudes (CA's) may induce dangerous hypoxemia in pts with cyanotic congenital heart disease (CCHD). To check the validity, the SaO_2 was measured transcutaneously in 12 grown-ups with CCHD and 27 young air pilots for control during simulated commercial flights of 2.5 and 7 hours in a hypobaric chamber at the Netherlands Aerospace Medical Centre. This study was extended with 10 of those pts and 6 controls during a return flight Amsterdam-Malaga with a DC10 on the outbound and a A310 on the return flight. Duration of the flights was approximately 3 hours with maximal CA's of 6000 and 5800 ft respectively. In addition to the SaO_2 measurements the capillary pH, blood gases and lactic acid levels were also measured. During the simulated flights, the SaO_2 was at all times lower in the pts than the controls. However, the mean actual percentage drop, even after 6 hours at 8000 ft CA, was <10% in both pts and controls. The mean SaO_2 in the pts decreased from 86±5% to 79±5%. The duration of the "flight" had no influence on the SaO_2 reduction. During the actual air travel, the maximal mean SaO_2 reduction was <6%. A marked drop of the pO_2 was measured in the controls but only a mild and not significant reduction in the pts. This may explain the relatively small drop of SaO_2 in the pts despite the fact that their arterial pO_2 values were all near the steep slope of the hemoglobin/oxygen dissociation curve. The flights had no influence on the capillary blood pH, pCO_2, bicarbonate and lactic acid levels. We conclude that the changes of atmospheric pressure during commercial air travel do not seem to be detrimental to pts with CCHD.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 16

Significant pure aortic regurgitation in children and adolescents with bicuspid aortic valve—prevalence and the possible role of the aortic root dilatation

Daskalopoulos DA
Athens, Greece

Significant chronic aortic regurgitation (CAR) in patients (pts) with congenitally bicuspid aortic valve (BAV) without coexisting aortic stenosis (AS), or of evidence of inflammation, is an uncommon clinical problem. The objectives of this retrospective study were: 1) to study the prevalence of significant (sign), pure CAR among our pediatric pts with BAV, the prevalence of such pts among our pts with sign CAR, and their clinical profile; and 2) to attempt to define the cause of the possibly dilated aortic root (Ao) in pts with BAV and sign CAR. Over the past 6 years, 84 pts studied by 181 echocardiograms (echos), were found to have BAV. Ten of the 84 pts (11.9%), 7 male and 3 female, had sign CAR and constitute the study group. During the same period, 29 pts studied by 52 echos were found to have sign CAR.The CAR was moderate in 7 and severe in 3 pts; the latter have already undergone aortic valve replacement at age 8, 11, and 17 years. The Ao was dilated in 9 of the 10 pts (90%). By comparison, dilated Ao was uncommon in the other pts with BAV—57 pts had a functionally normal BAV or mild AS, and were studied by echo at age 1 day-23 years (median 5 years). Only 3 pts (5.2%) had a dilated Ao. The remaining 17 pts had significant AS, with or without AR, and were studied at age 16 days-20 years (median 2 years). We also studied the frequency of dilated Ao among pts with other causes (other than BAV) of sign CAR. Six such patients were studied by 9 echos at age 1-14 (median 12) years. Only 1 pt (16.7%) had dilated Ao. In our experience, sign CAR is not uncommon among children and adolescents with BAV. BAV seems to be a common cause of sign AR in pediatric patients, and there is a clear and specific relationship with dilated Ao. In some of the pts with sign CAR and BAV, dilation of the Ao appears at least to contribute to the pathogenesis of AR (possibly as a form of annulo-aortic ectasia), and does not appear to be the effect of the forces related to the disturbed flow and exerted on the aortic wall.

P 17

The ventilatory responses to exercise late after repair of tetralogy of Fallot

Gatzoulis MA, Clark AA, Redington AN
The Royal Brompton Hospital, London, UK

The purpose of this study was to assess ventilatory responses to exercise late after tetralogy of Fallot repair (TOFr) and their relationship to exercise capacity and right ventricular (RV) diastolic function. Thirty TOFr patients (7 female) aged 27.8±6 years and 30 age and gender-matched controls underwent exercise testing (modified Bruce protocol) with metabolic gas exchange to determine peak oxygen consumption (peak VO_2), the slope of the relationship between both respiratory rate (RR) and ventilation (VE) against carbon dioxide production (VCO_2). Patients and controls underwent spirometry. Restrictive RV physiology was defined by the presence of late diastolic antegrade flow in the pulmonary artery, shortening the duration of pulmonary regurgitation(PR). Respiratory function tests in the TOFr group were normal, but significantly lower than the controls when expressed as percent predicted, $p<0.03$. Patients' average peak VO_2 was 35.3±7.5 ml/kg/min (93.6±15.3% of expected for age, weight, height and gender). The RR/VCO_2 slope was steeper in the Fallot group (6.8±2.6 vs 9.6±4.7, $p<0.02$). Within the group, those with RV restriction achieved a higher peak VO_2 than those without (82.5±10.1 vs 100.9±13.8%, $p<0.001$). In the Fallot group alone, there was an inverse relationship between ventilatory response and peak VO_2 (RR/VCO_2 vs peak VO_2, $r=0.63$, $p=0.003$ and VE/VCO_2 vs peak VO_2, $r=0.62$, $p<0.001$). TOFr patients as a group had near normal exercise capacity, but as exercise capacity decreased, the ventilatory response to exercise was increased. Within the tetralogy group, those with restrictive RV Doppler physiology as a whole performed better than those without, perhaps by limiting PR. We propose that in TOFr patients the increased ventilatory rate at a given level of carbon dioxide production acts as a respiratory pump, aiding right ventricular function by making an important contribution to effective pulmonary blood flow.

P 18

Assessment of perioperative changes of ventricular functions and dimensions in totally corrected tetralogy of Fallot

Jaeggi E, Verine V, Oberhaensli I, Friedli B
Pediatric Cardiology Unit, Hopital Cantonal Universitaire, Geneva, Switzerland

Perioperative changes of ventricular systolic/diastolic function and dimensions were studied in 11 patients (5.8±3.4 yrs) with TOF. Measures were obtained sequentially by transthoracic (TTE) and transesophageal (TEE) echoes: preoperatively (1), before sternotomy (2), after sternal closure (3), 4-6 hours (4),1-2 days (5), and 1 week (6) after total repair. Diameter, area and volume of LV and area of RV were measured at end-systole (ES) and end-diastole (ED) and corrected for body surface area. Ventricular function was assessed by ejection phase indices and load independent stress/velocity relations (rate corrected mean VCF/peak systolic wall stress=mVCFc/PsWS). Diastolic LV function was evaluated by early and late peak velocities (E;A) and corresponding velocity time integrals (Ei; Ai; Ei/MVTI) and rate corrected isovolumetric relaxation time (IVRTc). (See poster for table of results.) Total repair of TOF resulted in an immediate increase in LV diastolic and systolic diameter and in LV/RV-area-ratio. LV systolic function was depressed by anesthesia and bypass but recovered to near normal values by one week. MVCFc/PsWS suggests depressed function under anesthesia but preserved systolic function with excellent response to vasoactive drugs after coming off bypass. Diastolic LV function parameters normalized within 2 days after operation, showing an increase in early and decrease in late diastolic filling, and a shortening of the IVRTc.

P 19

Early peripheral vascular manifestations of heterozygous familial hypercholesterolemia

Johnson D, Fournier A, Therrien J, Marchand T, Perrault H, Montigny M, Levy E, Davignon A, Lambert M
Ste-Justine Hospital, Montreal, Canada

Thirty-four children aged 13.0±3.6 years with heterozygous familial hypercholesterolemia (FH), all carrying the frequent French Canadian mutation (>10kb deletion) underwent cardiac and peripheral vascular evaluation. Twenty-nine patients (pts) (85%) had a family history of premature atherosclerosis. All pts were asymptomatic, except 4 who complained of nonspecific chest pain. Physical examination was normal in all including blood pressure, body mass index (BMI) and absence of tendon xanthoma except 2 pts with arcus cornealis. Resting and exercise electrocardiograms as well as echocardiogram were normal. Duplex ultrasound provided transcutaneous measurements of carotid, brachial and femoral arteries flow kinetics at rest, immediately after cuff deflation, 20 seconds and 5 minutes later (brachial and femoral arteries: cuff inflation at 200 mm Hg for 3 minutes). The following data were obtained: peak, diastolic and mean velocities (cm/sec) and Pourcelot resistance index (RI). These results were compared to those of 33 control subjects (ctrl) of same age, blood pressure and BMI. Cuff deflation resulted in an increase in velocities and a decrease in resistance index in all subjects. Brachial peak and mean velocities of pts were similar to those of ctrl under all conditions. Changes in velocities were also similar in both groups. However, femoral mean velocity at rest and 20 sec after cuff deflation was significantly lower in pts than ctrl. Femoral RI at rest was significantly higher in pts than ctrl (1.32±0.11 vs 1.25±0.09, $p<0.01$). Both brachial and femoral RI at 20 sec and 5 min after cuff deflation were significantly higher in pts than ctrl. In conclusion, although having normal cardiac evaluation and having responded in the same fashion to reactive hyperemia, children with FH show a higher peripheral resistance index following cuff deflation. This may reflect already at this early stage of the disease the presence of an endothelial dysfunction and/or alterations in vasomotor control.

P 20

Complications of cardiac catheterization in a pediatric patient population

Vitiello R, Nykanen D, McCrindle B, Freedom RM, Benson LN
The Hospital for Sick Children, Toronto, Canada

A study of adverse events related to diagnostic (DIAG) or interventional (INTERV) pediatric cardiac catheterization was undertaken between 1/1/87 and 31/12/93 from 4,953 consecutive procedures. Patient ages ranged from 1 day to 20 years (median 2.9 yrs). There were 1,447 (29%) INTERV, 3,117 (63%) DIAG, and 357 (7%) electrophysiologic studies (EPS), with 32 (1%) patients having both INTERV and EPS. One or more complications were reported in 494 (10%) patients. Independent risk factors from multiple logistic regression analysis for any complication included age (<1 mo, odds ratio 3.2, p<0.0001; age 1-6 mo, odds ratio 2.9, p<0.0001 and age 6 mo-2 yr, odds ratio 2.2, p<0.0001) relative to age 5-10 yr, and INTERV (odds ratio 1.5, p<0.0001) relative to DIAGN. After controlling for patient age and type procedure, no additional factor independently predicted any complication. Major complications (death, 35; cardiac arrest, 7; cardiac perforation, 8; complete heart block, 14; ventricular tachycardia/fibrillation, 5) were reported in 70 (1.4%) patients, with 63% of deaths occurring in patients age <1 yr. Independent risk factors for major complications included only age <1 mo (odds ratio 3.8, p<0.003) and age 1-6 mo (odds ratio 2.4, p<0.05) relative to age 5-10 yr. Minor complications occurred in 476 patients and included 186 (3.8%) arterial complications with residual pulse weakness after therapy in only 14 patients. Independent risk factors for arterial complications included age <1 mo (odds ratio 4.1, p<0.0002), age 1-6 mo (odds ratio 8.0, p<0.0001), and age 6 mo-2 yr (odds ratio 5.3, p<0.0001) relative to age 5-10 yr, and INTERV (odds ratio 1.8, p<.0002) relative to DIAGN. In conclusion, pediatric cardiac catheterization is not risk-free, although the incidence of major complications is low. With trends toward more intervention in younger patients the incidence of complications may increase.

P 21

Gamma globulin retreatment in Kawasaki disease—clinical, laboratory and echocardiographic preliminary data

Pedroni E, Auriacombe L, Squarcia U, Perrone S, Baldioli C, Burgio GR
Pediatric Cardiology Department, University of Pavia, Pavia, Italy and Pediatric Cardiology Department, Hopital Necker Enfants Malades, Paris, France

In children with Kawasaki disease (KD) and persistent fever (>38 °C, more than 4 days) and/or coronary artery abnormalities (CAA), we applied a retreatment with standard intravenous gamma-globulin (IVGG) and we report the preliminary results. From 1/2/88 to 31/10/94, 29 patients (pts), 18 males and 5 females, aged 3 to 132 months (median 26), were retreated with IVGG, 2 gm/kg. Eight pts were retreated because persistent fever ± other clinical or inflammation signs, 15 pts because of CAA—echocardiography identified 3 pts with giant aneurysms (diameter≥ 8 mm), 7 with aneurysms (>1 mm of normal value, <8 mm) and 5 with dilatations (≤1 mm of normal value). Pts with CAA were separated into two groups: 7 pts group A—1 giant aneurysms, 2 aneurysms, 4 dilatation, in whom CAA detection occurred early (not after first IVGG), normalized their ultrasound coronary aspect (median 142 day from disease onset); 8 pts group B—2 giant aneurysms, 5 aneurysms, 1 dilatation, in whom CAA detection occurred late (after first IVGG), didn't normalize their ultrasound coronary aspect at medium follow-up of 249 days. Both groups had first (median 11 day) and second (median 32 day) course of IVGG in the same medium day of illness. Group A differed significantly from B in median age (A: 39 months, B: 8 months, p<0.01), but there was no difference in gender distribution. Laboratory findings (including platelet and leukocyte count, fibrinogen, ESR) and magnitude of fever were more elevated in group B. In all pts, retreatment with IVGG produced defervescence within 72 hours with quick improvement in clinical signs and prevented CAA diffusion from becoming worse. We conclude that elevated laboratory findings, greater magnitude or persistence of fever, younger age, seem to be greater risk factors of unfavorable clinical and cardiac evolution.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 22

Echocardiographic diagnosis and management of cardiac lesions in Kawasaki disease—a 174-patient study

Pedroni E, Squarcia U, Caramia G, Vertua G, Sellitto F, Martini A, Musumeci S, Ceccarelli M, Chitti D, Perrone S, Baldioli C, Burgio GR
Pediatric Cardiology Department, University of Pavia, Pavia, Italy

Echocardiography (ECHO) imaging technique has become an essential tool for the diagnosis of the coronary lesions and management of children with Kawasaki disease (KD). From 1/2/88 to 31/10/94, 174 patients (pts) with KD (120 male, 54 female) were submitted to the Coordinator Center in Pavia from participating groups to KD Aneurysms Prevention Protocol and Italian Multicenter Trial suggested by the Pediatric Cardiology Research Group of the Italian Paediatric Society. Pts mean age was 31 months (range 2-156). Diagnosis of KD was made when 5 of the 6 diagnostic criteria were present. Two-dimensional ECHO was performed routinely to evaluate coronary arteries at a mean interval from disease onset of 12th day (range 1- 75). A coronary artery with a diameter ≤1 mm of normal value (NV) was judged as non-significant dilatation (11 pts); an artery was judged as abnormal when its diameter was >1 mm of NV <8 mm—aneurysm (16 pts) and ≥8 mm—giant aneurysm (5 pts). Multiple aneurysms were present in 9 pts and fatal myocardial infarction occurred in one. In 34% of the cases, the lesions concerned the common branch, in 30% the right coronary artery, in 19% the circumflex branch and in 17% the anterior interventricular artery. On ECHO M-mode, we found that 21 pts presented pericardial effusion associated with coronary abnormalities in 11; 3 pts presented left ventricular dilation and 5 pts decreased fractional shortening. All pts with aneurysms received IVGG (2 gm/kg) on the mean interval from disease onset of 13th day. Echocardiographic follow-up on the 30th day of illness showed persistence of aneurysms. We emphasize that ECHO is widely recognized as a sensitive screening tool for coronary aneurysms in KD, with very high sensitivity and specificity both in the acute and chronic stages. When coronary artery abnormalities are detected, ECHO can serially evaluate long-term results of treatment and monitor the evaluation of coronary lesions.

P 23

Neurally mediated syncope in children—treatment versus non-treatment

Pongiglione G, Rimini A, Ribaldone D
Giannina Gaslini Children's Hospital, Department of Cardiology, Genoa, Italy

The treatment of children with neurally mediated syncope (NMS) at a follow-up of 6 months has been shown to be equally effective either using atenolol (A) or fluorocortisone (F). The purpose of this study was to investigate the efficacy of therapy for NMS during the same follow-up period. Patients having at least 2 episodes of syncope in the preceding 6 months and with a positive tilt table testing were randomized to receive R, F or no treatment (O) and were followed for 6 months. Outcomes were classified as cured (C), improved (I) or not improved (N). A total of 36 patients with a mean age of 12.8 ± 2.5 years were studied. There were 16 males and 20 females. During tilt, 15 children required isoproterenol infusion to induce syncope, while 21 fainted in the basal state. Cardio-inhibitory responses were seen in 8 patients and were more frequent in the basal state ($p<0.05$). Vasodepressor and mixed responses occurred in 5 and 23 patients respectively. There were 7 C, 2 I and 1 N among the 10 children treated with R; 6 C, 5 I and no N in the group of 11 patients treated with F and 10 C, 4 I and 1 N among those patients who did not receive any treatment (O). There was no difference in outcome between the groups nor between the patients treated (R or F) and not treated (O). Assessment of treatment for neurally mediated syncope is difficult because of the high rate of spontaneous improvement of the disease and of the high variability of occurrence of the symptoms.

P 24

In vitro assessment of subaortic obstruction caused by restrictive VSD in double inlet left ventricle by intravascular and 3D ultrasound

Vogel M, Webber SA, Ho SY, Gnanapragasam J, Anderson RH
Department of Paediatrics, National Heart and Lung Institute, London, Wessex Cardiothoracic Center, Southhampton, UK

The study was undertaken to compare utility of intravascular and 3D-echo in measuring severity of subaortic stenosis caused by restrictive VSD in specimens with double inlet left ventricle and transposition. Eight hearts were studied by 3D and intravascular ultrasound (IVUS) and compared with anatomy (ANAT). The longest axis of the VSD and its smallest diameter, which in 7/8 was perpendicular to the longest axis, were measured along with aortic root size. IVUS, 3D and anatomic measurements were done in a blinded fashion. In 7/8 specimens the longest VSD diameter was smaller than the aortic root.

Diameter of VSD by method

Pt	IVUS	3D	ANAT	Aortic diameter
1	2 ± 1	2 ± 1	2 ± 1	9
2	12 ± 10	14 ± 9	11 ± 9	13
3	15 ± 10	16 ± 8	13 ± 8	17
4	9 ± 7	11 ± 7	12 ± 6	13
5	5 ± 5	5 ± 4	5 ± 4	7
6	15 ± 8	7 ± 3	8 ± 4	12
7	6 ± 4	12 ± 11	12 ± 10	16
8	9 ± 3	7 ± 5	9 ± 5	9

Correlation between anatomic measurements and IVUS for longest and smallest VSD diameter was poor at r=0.61 and 0.68 respectively, whereas it was good for 3D echo at 0.93 and 0.98 respectively. This may be due to the IVUS catheter not being completely inside the internal circumference of VSD. This technical problem of IVUS may be compounded in the beating heart, whereas 3D-echo provided excellent VSD size measurements *in vitro* and in addition is clinically easy to use, if applied transthoracically.

P 25

The asymmetric mitral valve as a spectrum of parachute deformity

Wenink ACG, Oosthoek PW, Macedo AJ
Department of Anatomy, University of Leiden, Leiden, The Netherlands

Twenty-two of 100 specimens with congenital mitral valve (MV) pathology were selected because of asymmetry of the valve (21) or single papillary muscle (1). Three groups were made, with slight (I, 4/21), moderate (II, 13/21) and severe (III, 4/21) asymmetry. All specimens had other congenital heart disease, aortic arch obstructions occurring in 12. Eight of G II and all of G III also had a VSD. The following measurements were taken: MV annulus diameter (corrected for tricuspid valve annulus and left ventricle (LV) length); the length of MV leaflets, chords, papillary muscles (PM) and the distance from the PM basis to the LV apex—all being compared with a control group of 20 normal hearts. Twenty of 21 MV had pathology of the anterior tensor apparatus (TA), 1 had pathology of the posterior TA. In all of these, the involved PM continued as a muscle bundle reaching the aortic valve. The MV annulus was smaller than controls in G II and G III. For the other values, a grading was found from G I not different from controls, though G II in which a smaller anterior TA and a higher APM position within the LV was noted. In G III, unifocalization of MV chords (parachute anomaly) was found in all, 3 having chords to the PPM, 1 to the APM (asymmetric unifocalization). In the sole case of single PM, the PM had a central position (symmetric unifocalization). The parachute MV may be considered as consisting of incomplete forms with a small annulus and smaller TA to the anterior commissure, as a complement of the complete form, with unifocalization of the chords most frequently to the PPM. This variety of the parachute malformation has to be distinguished from that with a single PM which is centrally positioned and which is symmetrical. The association with aortic arch anomalies and also with VSD, and the presence of an additional muscle bundle at the site of the APM, may have clinical and etiological implications.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 26

Location of the chambers in the ventricular segment in atrioventricular and ventriculoarterial discordance and double inlet left ventricle with situs solitus

Quero-Jiménez M, Maître-Azcárate MJ, López-Zea M, Quero-Jiménez C, Fernandez-Piñeda L, Cazzaniga M
Servicio de Cardiologia Pediatrica, Hospital Ramon y Cajal, Madrid, Spain

The aim of this study is to communicate the practical generalizations on the location of the chambers in the ventricular segment which have been obtained after studying 31 patients (pts) with atrioventricular and ventriculoarterial discordance (AV and VA DC) and 90 pts with double inlet left ventricle (DILV), all of them with situs solitus visceroatrialis (SS). In every pt, a careful segmental analysis has been performed with particular attention having been paid to the relative special locations of the ventricles (V) and/or accessory chambers (AC), the direction pointed at by the apex (A) and the type (D or L) of bulboventricular (BV) loop. All pts with AV and VA DC had a right-sided ventricle of left ventricular morphology, which was anterior in 17 cases with A pointing at the left, posterior in 4 with A pointing to the right and side-by-side in 10 with mesocardia and mesoapex. Among the 90 pts with DILV, 49 had an L-loop (A pointing at the left in 32 and at the right 17) and 38 a D-loop with A pointing at the left, the BV-loop being undetermined in 3. The combination of an L-loop with A pointing to the left (in a "discordant" way) showed the chamber of left ventricular morphology to be anterior in each pt. The association of a D-loop with an A pointing at the left (in a "concordant" way) revealed always that the morphologically left ventricle was posterior. In conclusion, 1) the anterior or posterior location of a V and/or an AC may be predicted indirectly in the great majority of pts through the location of the aorta and the direction of the A; 2) this is definitely useful both in echo and angio examinations and in surgical inspection of complex congenital heart defects.

P 27

Transverse arch hypoplasia predisposes to aneurysm formation at the repair site after Dacron patch angioplasty for coarctation

Gewillig M, Bogaert J, Mertens L, Vanhove M, Dumoulin M, Raedemakers F, Verschaekelen J, Daenen W, Baert A
Gasthuisberg University Hospital, Leuven, Belgium

Aneurysm formation is a well-known complication after Dacron patch angioplasty in patients with coarctation of the aorta. However, the underlying mechanisms (flow pattern, wall abnormality) remain unresolved. This makes the postoperative history unpredictable and therefore adequate follow-up difficult. We evaluated 28 patients (20 males, 8 females; mean age 17.8±3.8 years, range 13-29 yrs) after Dacron repair for coarctation with MRI (mean time between surgery and MRI, 11.5±1.8 years, range 8-15 yrs). An aneurysm was defined by a ratio of the aortic size at the repair site to the aortic size at the diaphragm greater than 1.5. A transverse arch hypoplasia was defined as a ratio of transverse arch size to diaphragmatic aorta size less than 0.9. Twenty-one patients presented with an aneurysm at the repair site (ratio, 2.00±0.46, range 1.50-3.20; diameter, 40±15 mm, range 22-74 mm). All those patients showed a concomitant hypoplastic transverse arch (ratio, 0.74±0.08, range 0.52-0.85). Cine MRI was performed in 18 cases. In 14, signal loss suggesting flow acceleration or turbulence, was seen from the hypoplastic transverse arch to the aneurysm. In two, signal loss was seen only in the aneurysm. Two cine MRI studies were excluded due to inferior image quality. Seven patients showed a ratio at repair site less than 1.5 (mean 1.13±0.29, range 0.71-1.41). All showed a normal transverse arch ratio (mean 1.0±0.1, range 0.9-1.19). On cine MRI performed in 5 patients, no signal loss was detected. A hypoplastic aortic segment predisposes to aneurysm formation after Dacron patch aortoplasty for coarctation (Chi square $p<0.0001$). We speculate that the jet originating in the hypoplastic segment and the poststenotic turbulence cause wall dilation with late aneurysm formation.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 28

Protein-losing enteropathy after Fontan operation

Malcic I, Sauer U, Brodherr S, Eicken A, Genz T, Greil G, Kirchner K, Sebening W, Soppa C, Vogt M, Bühlmeyer K
Deutsches Herzzentrum, München, Germany and University Hospital Rebro, Zagreb, Croatia

Protein-losing enteropathy (PLE) is a well known late complication after Fontan operation (FO). This study examines the possible causes of this condition. Between July 1974 and July 1994, 181 pts (102 male, 79 female) underwent FO at DHM (mean age 8.6 yrs; range 4 mo-27 yrs). In 23 pts (12.7%), PLE developed within 1-127 (median 10) mo after FO (G1). Besides a low serum protein 4.4 gm/dl, relevant findings were: edema (95%), pleural effusions (93%), ascites (88%), chronic heart failure (62%), ECHO-EF <55% (86%), AV-valve insufficiency (40%), hypocalcemia (78%). $Alpha_1$-AT clearance in stool and lymphocytopenia was not assessed. When comparing the clinical and hemodynamic data in G1 with that in a subgroup of 20 pts without PLE (G2) with similar gender, type of CHD and op, significant differences emerged: pts with PLE (G1) were older at FO [mean age G1 (9 yrs) vs G2 (6.5 yrs)] and post-FO increase in diastolic RA pressure was higher in G1 (16 vs 11.7 mm Hg). Furthermore, PLE was prominent in pts with prior-PAB (9/23) and mortality post FO was higher in G1 than in total group (11/23 vs 36/181) (p=0.02). In summary, the risk for PLE after FO was significantly related with excessive size, high diastolic pressure and non-pulsatile, turbulent flow in RA and long-standing prior-PAB. Right-sided obstruction was not prevalent. Surgical implications to avoid PLE include: 1) Early FO, especially in pts with prior PAB; 2) Undelayed reoperation in selected pts, e.g., with PLE, designed to decrease the size of RA and restore laminar blood flow.

P 29

Pulmonary venous blood flow in the human fetus

Paladini D, Palmieri S, Morelli P, Morra T, Martinelli P, Calabrò R
Fetal Cardiology Unit, Department of Gynecology and Obstetrics, Federico II University of Naples and Pediatric Cardiology, 2nd University of Naples, Naples, Italy

In the adult, the study of pulmonary venous blood flow has become part of left ventricle diastolic function evaluation. In the fetus, several reports have dealt with cardiac function, but pulmonary venous blood flow throughout gestation has never been evaluated. We studied 96 singleton pregnancies between the 17th and the 41st week of gestation. In all of them, pulmonary venous blood flow was investigated by 2D, color Doppler and pulsed Doppler echocardiography. On color Doppler, the upper right pulmonary vein could be imaged in 89.6% of cases in its peri-atrial tract and in 75% of cases within the lung. In contrast, the upper left pulmonary vein could be imaged only in 8% of cases close to the atrium and in 41% of cases intraparenchymally. Reliable velocity waveforms were obtained in 91% of cases. Pulmonary venous velocity waveforms resemble those obtained in the adult, despite the fetal circulation. Mean systolic peak velocity was 22.6±8.5 cm/sec and mean diastolic peak velocity 22.4±8.8 cm/sec. Both increased significantly with gestational age (p<0.00001 and p<0.001, respectively). As a result, their ratio remained constant throughout gestation (1.04±0.18). Reverse blood flow was present in 18% of cases, regardless of gestational age and fetal heart rate. Expressed as percentage of forward flow velocity time integral, its value was 7.65±5.2%. There was no difference with the results obtained in ten of the 96 fetuses followed longitudinally. We conclude that these data seem to confirm the presence of pulsatile pulmonary venous blood flow in the second and third trimester fetus. Systolic and diastolic peak velocity are lower than in the adult, but they increase steadily with advancing gestational age. Normative data have been established for the second and third trimester of pregnancy. Further studies are needed to evaluate pulmonary venous flow in pathologic conditions.

P 30

Growth of pulmonary artery after arterial switch operation for simple transposition of the great arteries

Massin M, Nitsch G, Däbritz S, Messmer B, von Bernuth G
Department of Pediatric Cardiology and Cardiovascular Surgery, RWTH Aachen, Aachen, Germany

Forty-nine patients with complete transposition of the great arteries who underwent arterial switch operation with the Lecompte maneuver during the neonatal period were investigated by cardiac catheterization and cineangiography an average of 13.4±3 (SD) months after the operation. The aim of this study was to assess the size and the growth pattern of the pulmonary artery. The diameter of the main pulmonary artery (MPA) at different sites (annulus, sinus, trunk) was measured in posteroanterior and lateral projections, and that of the proximal right (RPA) and left (LPA) pulmonary arteries was measured in posteroanterior projection on the right ventricular angiocardiogram. The values were compared with those of corresponding sites of age-matched normal children taken from the literature. An underdevelopment of the annulus (9.87±1.62 mm), the sinus (12.83±2.48 mm) and the trunk (10.15±2.1) in the lateral projection was established (p<0.01), but the values at the same sites were 37, 34 and 31% larger in the posteroanterior projection, so that the cross-sectional area was normal for the two first sites but smaller than normal for the trunk (110±42 mm^2; p<0.01). An underdevelopment of RPA (8.12±1.71 mm) and LPA (7.83±1.46 mm) was also established (p<0.01). No correlation could be demonstrated with neonatal values in the 24 patients who had a preoperative investigation (r=0.02/0.13). Mild gradients at the bifurcation were regularly diagnosed. A negative correlation was found between the gradient at the origin of RPA and LPA and the diameter/m^2 BSA (r=0.66/0.60, p<0.01) of the concerned branch. The ratio of the gradients over both branches RPA/LPA was inversely proportional to the growth ratio of both branches (ratio of post- and preoperative values for RPA divided by the same ratio for LPA) (r=0.79; p<0.001). It is concluded that the Lecompte maneuver induces a flattening of the MPA with concomitant reduction of its cross-sectional area.

P 31

Bidirectional cavopulmonary anastomosis with additional flow sources to the lungs—clinical experience in 16 cases

Borini I, Dalmonte P, Gorrieri PF, Marasini M, Grasso P, Zannini L, Bertolini A
Institute G. Gaslini, Departments of Cardiovascular Surgery, Cardiology, and Anesthesiology, Genoa, Italy

Bidirectional cavopulmonary anastomosis (BCPA) is utilized in the following clinical situations: 1) as staging procedure in complex congenital heart diseases with univentricular physiology; 2) as palliative procedure in cases with high risk factors for Fontan operation; 3) as part of a biventricular repair when the right ventricle is diminutive. From November 1992 to October 1994, we performed the BCPA with additional flow sources to the lungs in 16 cases (age 4 mos-12 yrs, 37.76±35.6 mos; weight 4-24.4 kg, 11.23±5.15 kg). The rationale for this approach was to try to increase arterial oxygen saturation beyond the limits of isolated BCPA, to ameliorate pulmonary flow distribution to both lungs and to obtain a more "pulsatile" pulmonary flow. The indication for BCPA was: staging procedure in two cases (1 HLHS, 1 univentricular heart with subaortic obstruction) and palliative procedure in the other cases. There was no hospital death. At follow-up (2-23 mos, 11.5±8.5 mos), we had 1 death four months after the operation for worsening AV valve incompetence. This was the only case with significant morbidity (repeated pleural and pericardial effusions). Percutaneous O_2 saturation was higher than 85% in 12 cases, and between 80 and 85% percent in the other cases. At operation, pulmonary artery pressure was moderately "pulsatile" in all cases. Only one patient has actually shown a reduction in heart size with two-D echo and chest x-ray. BCPA with additional flow sources can increase O_2 saturation beyond the limits of simple BCPA and obtain a "pulsatile" pulmonary flow, but could neutralize diastolic unloading of BCPA, without significantly modifying blood flow distribution to the lungs.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 32

Should we close all perimembranous subaortic ventricular septal defects with aortic valve prolapse?

Skovránek J, First T, Hucín B, Marek J, Samánek M
Kardiocentrum, Praha, Czech Republic

Subpulmonary VSD with AOVP is the indication for an early surgery in order to prevent aortic regurgitation (AR). Indication and timing of surgery in subaortic VSD with AOVP remains an open question. An echo/Doppler follow-up study was undertaken to assess the pre- and postoperative incidence and severity (three-grade scale) of AR in 109 pts with surgically verified subaortic VSD with AOVP. The first examination was carried out at age of 1-219 (mean 68) mos and studies were repeated over a period of 1-129 (mean 26) mos before and 12-135 (mean 54) mos after surgery. AR was not found at first echo/Doppler in 67 repeatedly examined pts. The 10-year probability of AR grade 2-3 developing was 51% (95% CL, 42-60%). In 20 pts with associated right ventricular outflow tract obstruction (RVOTO), the probability was 83 (75-92)% as compared to 43 (36-49)% in 47 pts without RVOTO ($p=0.002$). In total, 78 pts were operated on without preoperative AR. The 9-year probability of postoperative AR developing was 10 (3-18)% compared to 51 (42-60)% in pts not subjected to surgery ($p<0.001$). In conclusion, pts with subaortic VSD with AOVP (especially those with associated RVOTO) are at high risk of developing AR. VSD closure significantly decreases the risk. Subaortic VSD with AOVP should be closed to prevent aortic valve damage.

P 33

The assessment of phrenic nerve function after cardiac surgery

Dinarevic S, Al Marsafawy H, Bettles N, Shinebourne E
Royal Brompton Hospital, London, UK

The aim of this study was to evaluate the efficacy and utility of a new battery operated system-Phrenos in the assessment of phrenic nerve function pre and post paediatric cardiothoracic surgery. We undertook phrenic nerve stimulation study on 55 children (ages 2 weeks to 11 years) before and after cardiac surgery and at outpatient follow-up 3 months to 1 year later. A portable battery operated device was used which gives a constant voltage output from 0-300 v with a fixed duration of 100 msec. In most patients, a 1 mv/div electrical stimulus was applied over the phrenic nerve at the posterior border of the sternomastoid at the level of the upper border of the thyroid cartilage. A diaphragmatic electromyogram was displayed on a storage oscilloscope as the average of 10 sweeps. Mean phrenic latency time was 5.2±0.03 msec on the left side and 5.1±0.04 msec on the right side. Prolongation of phrenic nerve latency by more than 2 msec was found in 4 of 55 postoperative measurements and in 4 patients there was no response, due to phrenic nerve injury. Consequent to confirmation of unilateral phrenic nerve plasy in 4 infants, diaphragmatic plication was undertaken within 5 days of surgery in all patients contributing to a shortened duration of stay in the intensive care unit as compared to previous reports of this entity. In conclusion, we report a simple non-invasive device which can easily and safely be used at the bedside in any cardiac surgical unit. It offers a practical contribution to the rapid diagnosis and management of post operative phrenic nerve injury in pediatric patients.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 34

Angiographic study of the aorta, coronary arteries and left ventricular performance after arterial switch operation for simple transposition of the great arteries

Massin M, Däbritz S, Messmer B, von Bernuth G
Department of Pediatric Cardiology and Cardiovascular Surgery, RWTH Aachen, Aachen, Germany

Forty-one patients with complete transposition of the great arteries who underwent arterial switch operation (ASO) with Lecompte maneuver during the neonatal period were investigated by cardiac catheterization and cineangiography an average of 13.5±9 (SD) months after the operation to assess the growth pattern of the aorta and the coronary arteries and left ventricular function. The diameters of the aortic annulus, aorta at different sites (sinus, sino-tubular junction, shoulder, anastomosis, distal ascending aorta, isthmus, 1 cm distal to isthmus, thoracoabdominal junction) and coronary arteries were compared with corresponding diameters of age-matched normal children taken from the literature. Parameters of myocardial performance were also calculated. The neoaortic annulus was 1.2 SD larger and the neoaortic root at the 3 sites was 3, 2.6 and 0.6 SD larger than corresponding structures in control subjects. Pressure gradients were not observed. The anastomosis was 0.35 SD smaller ($p<0.01$) than the ascending aorta in control subjects. Except in one patient who underwent an internal mammary bypass graft for obstruction of the right coronary artery in the neonatal period, the coronary arteries, visualized by aortic root angiography, showed no stenosis. The size of the proximal coronary arteries was normal. The size reduction in the distal vessels was comparable with that of adults, except in two children who had a hypoplastic left anterior descending coronary artery (LAD). Except in the bypass child, the global LV function was normal with ejection fraction of 66.2±6.3%, end-diastolic volume of 63±8 ml/m² BSA, end-systolic volume of 21.7±5 ml/m² BSA and cardiac output of 3.9±0.5 l/min/m² BSA. The midterm results for ASO are encouraging. Progressive aortic dilatation, aortic valve regurgitation, coronary insufficiency or myocardial dysfunction in the future remain possible, so that long-term follow-up studies are important.

P 35

The optimal timing of primary anatomic total correction of transposition of the great arteries with ventricular septal defect

Urban AE, Wetter J, Brecher AM, Sinzo N
Kinderklinik, St. Augustin, Germany

The timing and strategy of surgical therapy in patients with transposition of the great arteries (TGA) complicated by large ventricular septal defect (VSD) remains controversial. The purpose of this study was to determine the risk of very early (neonatal) arterial switch (ASO) and patch-closure of VSD in this subset of patients. Between 1984 and 1994, 61 patients underwent ASO and patch-closure of VSD. Twenty-eight infants had total correction at a mean age of 135 days, 33 neonates at a mean age of 11 days respectively. Concomitant cardiac pathology was repaired at ASO. Of the 61 patients, two infants and one neonate died in the hospital with an early mortality of 4.9%. Four patients, all after ASO in infancy, had successful reoperations for various causes. The late mortality was 3.4% (2/58) and of unrelated reason. Postoperative follow-up reveals sinus rhythm and normal ventricular function in all. Five patients have significant RVOT gradients. In conclusion, this study suggests, that neonatal primary anatomic correction in patients with TGA complicated by a large VSD carries a low risk. Anatomic repair in this subset of TGA therefore should not be deferred into later age; long-standing palliative operations are not indicated.

P 36

Amrinone in postoperative treatment of infants and newborns after surgery for congenital heart defects

Laitinen P, Happonen J-M, Peltola K, Lundström U, Sairanen H, Leijala M
Helsinki University Children's Hospital, Helsinki, Finland

Twenty-five patients with reconstructive surgery for AVSD (14 pts) or arterial switch operation for TGA (11 pts) were randomized for postoperative treatment with amrinone 7.5 µg/kg/min (11 pts) or dopamine (5 µg/kg/min) combined with nitroglycerin (1 µg/kg/min) (14 pts), in a double-blind manner. The circulatory status was evaluated two-hourly on the first postoperative day and six-hourly on the second postoperative day. Epinephrine was added if more inotropic support was needed. Pulmonary and systemic flow indexes and corresponding resistance indexes (PVRI, SVRI) were calculated using the Fick principle, with oxygen contents measured from superior caval vein, pulmonary artery, left atrium and peripheral artery. Oxygen consumption was measured using Deltatrac II® metabolic monitor (Datex, Helsinki, Finland). The two study groups did not differ in age, diagnosis, perfusion time, length of circulatory arrest, postoperative mean arterial pressures or the incidence of postoperative complications. No adverse effects attributable to the medications were noted. The results for the first postoperative day are presented in the table.

	Amrinone		Dopamine + Nitroglycerine		
	Mean	95% CI	Mean	95% CI	p value
PVRI (U · m²)	4 9	4.6-5.2	7 5	6 8-8.2	0.031
SVRI (U · m²)	24 5	23 1-25.9	29.7	27 8-31.6	0 087
PVRI/SVRI	0 22	0.20-0.24	0.27	0.25-0 29	0 211
ICU stay (days)	5.8	3 8-7.8	7 5	5 7-9 3	0 233

The lower PVRI with amrinone during the early postoperative period together with systemic inotropic support comparable to dopamine, are clearly beneficial in this selected group of patients prone to pulmonary hypertension. Maintaining low PVRI may be a major factor in optimal postoperative recovery.

P 37

Left atrioventricular valve incompetence after repair of common atrioventricular canal defects

Stellin G, Rubino M, Michielon G, Bianco R, Milanesi O, Svaluto Moreolo G, Daliento L, Casarotto D
Departments of Cardiac Surgery, Pediatrics and Cardiology, University of Padova, Padova, Italy

Since 1982, 207 patients with common atrioventricular canal defects (CAVCD) underwent surgical repair at our institution. A complete form (CCAVCD) was present in 82 pts, a transitional form in 23 pts and a partial form in 102 pts. Median ages at operation were 6 mo, 10 mo and 10.7 yrs respectively. Preoperative moderate to severe AV valve incompetence (AVVR) was present in 80 pts (38%) with a lower incidence in the CCAVCD group (26/82, 32%) versus the transitional (10/23, 43%) or the partial forms (44/102, 43%). The incidence was even lower within the CCAVCD group for pts younger than 4 mo (4/29, 13%). Repair of the left AV valve included reconstruction of the septal commissure and annuloplasty for central valve leaking. Moderate to severe AVVR occurred by echocardiography in 55 pts postoperatively. This was a contributing cause of death in 9 pts and the reason for reoperation in another 9. In 78%, the mechanism for AVVR requiring reoperation was the asymmetric apposition of the superior and inferior septal leaflets. At reoperation, valvuloplasty was always performed, with one late death. AVVR improved with surgery in all CCAVCD pts younger than 4 mo. Early age (≤4 mo) was not a risk factor for new onset of AVVR postop. In conclusion, repair of CAVCD remains a surgical challenge and should be addressed in an individual fashion. Postoperative AVVR can still be corrected by valvuloplasty with low mortality. Early repair of CCAVCD could prevent progression of valve displasia, therefore reducing the overall incidence of AVVR postoperatively.

P 38

Staged correction for hypoplastic left heart syndrome

Stellin G, Michielon G, Rubino M, Milanesi O, Bianco R, Daliento L, Casarotto D
Departments of Cardiac Surgery, Pediatrics and Cardiology, University of Padova, Padova, Italy

Between October 1992 and October 1994, 16 patients (pts) with hypoplastic left heart syndrome (HLHS) underwent staged surgical correction at our institution. Mean age at time of stage I was 7.6 days (1 to 56 days) and weight varied between 2.2 and 3.8 kg (mean 3.1). Aortic atresia/mitral atresia was diagnosed in 11 pts, and aortic atresia/mitral hypoplasia was present in 5 pts. Preoperative management included PGE_1 infusion in all pts and CO_2 inhalation (between 7 and 14 Torr) in the last 3 pts. All the neonates but one were managed to obtain a relative balanced hemodynamic and metabolic arrangement at time of stage I palliation. Associated cardiovascular lesions were isthmic coarctation of the aorta (or intimal ridge) in 8 pts, intact atrial septum in 1, persistent left superior vena cava to the coronary sinus in 2 and single coronary ostium in 1. A balanced systemic/pulmonary blood flow after stage I was obtained by means of vasodilators, hypoventilation and most recently by inhalation of CO_2. Nine pts (56%) survived stage I paliation and were discharged home between 12 and 32 days postoperatively. Five pts underwent stage II palliation at 6-7 months of age with no deaths. A bidirectional SVC-RPA anastomosis was performed in 2 pts and a hemi-Fontan (Norwood stage II) in 3 pts. Balloon dilation of the aortic isthmus was performed in 2 pts before stage II repair. A modified Fontan operation (Norwood stage III) was successfully performed in one pt at 19 months of age to maximize arterial oxygen saturation. In our limited experience, we conclude that the Norwood staged approach to HLHS represents the ideal form of surgical therapy. Unloading of the right ventricle at 6 months of age with a hemi-Fontan allows geometric and morphologic changes of the ventricle in preparation for the modified Fontan operation.

P 39

Pulmonary banding on probation—a diagnostic criterium for "one-stage" arterial switch operation in infants with simple transposition of the great arteries

Engelhardt W, Däbritz S, Messmer BJ, von Bernuth G
Departments of Pediatric Cardiology and Cardiovascular Surgery, RWTH Aachen, Aachen, Germany

Patients with simple TGA destined for arterial switch operation ideally are corrected within the first 2 or 3 weeks of life because left ventricular work capacity tends to be insufficient thereafter. Six of 193 patients with simple TGA were admitted to our institution for surgical management at four weeks of age or later due to delayed diagnosis (5) or perinatal cerebral hemorrhage (1). We report on 4 of them who showed low left ventricular pressure at cardiac catheterization and a banana-shaped left ventricular at echocardiography. The patients were operated on between 28 and 68 days of life by a sternal approach, leaving the option of an arterial switch procedure. Left ventricular pressure was measured intraoperatively to be half systemic or less (pulmonary-to-systemic systolic pressure ratio 0.2-0.53). Therefore, probational banding of the pulmonary trunk was performed, achieving a pulmonary-to-systemic systolic pressure ratio of about 0.8-1.0 and kept for 15 to 30 minutes. All infants tolerated the increase in left ventricular pressure work well. It was therefore decided to remove the pulmonary band and to proceed with an arterial switch. No serious complications occurred postoperatively. Weaning from mechanical respiration (2-7 days) was delayed in the oldest infant, but all patients could be dismissed home within 15 to 20 days. In the oldest infant (preoperative pressure ratio 0.2), a dramatical increase in the left ventricular posterior wall thickness and a normalization of left ventricular shape was observed within ten days. We conclude that, in infants up to 2 or 3 months of age with simple TGA and low preoperative left ventricular pressure, pulmonary banding on probation may be helpful to decide on the surgical management. If no left ventricular failure occurs intraoperatively, the banding can be removed and an arterial switch procedure performed.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 40

Long-term follow-up after acute myocarditis in childhood

Di Filippo S, Gouton M, Sassolas F, André M, Stamm D, Floret D, Bozio A
Hopital Cardiologique, Lyon, France

We reviewed the records of 94 patients (52 girls and 42 boys), aged 3 days to 14 years (median 14 months) admitted for acute myocarditis from 1970 to 1994. Severe heart failure (HF) was present at admission in 28 patients (pts) and 23 needed ventilatory assistance. Fifty-three pts had moderate HF and 3 had inaugural arterial emboli. Seven pts displayed severe arrhythmias. Initial left ventricle systolic shortening fraction varied from 5 to 35% (mean 16.5%). Most pts (73) had a recent infection, mainly in the upper tract airways. Viral serologies were positive in 43 cases. On follow-up (FU) (4 months to 24 years, mean 8.9 yrs), 33 pts died, 48 recovered, 10 improved and 3 were transplanted. Death occurred 1 day to 9.5 years (yrs) after onset (mean 11.3 months, median 1.5 mos), mainly by refractory HF (22 pts) or by sudden death (5 pts). Normalization of clinical and echo data occurred in 51% of the pts within 8 days to 15 yrs after onset (mean 18.6 mos). Most of these pts recovered within 2 yrs (35.5% at 6 mths, 56.4% at 1 year, 73% at 2 yrs), but cure could be observed much more later (until 15 yrs FU). Acute myocarditis in childhood is a severe heart disease with maximal mortality during the first year FU. In our experience, recovery can occur even very late and long-term prognosis seems to be more favorable after one year FU.

P 41

Improving results from the Fontan operation for patients with asplenia and polysplenia

Mair D, Cetta F, Feldt R, O'Leary P, Warnes C, Offord K, Puga F, Danielson G, Schaff H
Mayo Clinic, Rochester, USA

The asplenic and polysplenic syndromes characterized by heterotaxy are frequently associated with very complex congenital heart disease for which the modified Fontan (MF) procedure is the most definitive surgery available. Operative mortality (OM) after MF was high during the early years of our experience but has improved dramatically during the more recent time period. We reviewed all asplenic and polysplenic patients who had a MF at our institution between 1973 and 1992 (n=111). These 111 patients represented 13% of the 839 MF procedures performed at our institution during that time period. Fifty-four patients were polysplenic and 57 asplenic. Median age at the time of operation was 11 years. OM for the time period 1973-1986 was 43% (21/49) as compared to 15% (9/62) having surgery between 1987 and 1992 ($p \leq 0.001$). In the early experience, asplenic patients had a significantly increased OM compared to polysplenic patients, but in the more recent time period, they have been similar (asplenia 5/34,15% vs polysplenia 4/28, 14%). Many factors involving improved preoperative diagnosis, operative techniques, and perioperative intensive care have contributed to the improved results. The risk of the MF remains higher than for tricuspid atresia (OM, 4%) or double inlet left ventricle (OM, 4%). This appears likely due to increased frequency of complicating features in these heterotaxy patients particularly atrioventricular valve incompetence. AVV repair/replacement was more frequent in heterotaxy patients than the nonheterotaxy group (45 vs 14%). In heterotaxy patients who did not have simultaneous AVV procedures, OM was 9% (3/34) compared to 21% (6/28) who did have AV valve surgery. The 9% OM rate observed in these patients without AVV surgery is not significantly different from the 4% OM rate observed in nonheterotaxy patients (p=0.2).

P 42

Isolated subaortic stenosis in children—clinical evolution in mid-term follow-up

Kaneva A, Velkovski I, Pavlova M, Pilossoff V, Tzonzarova M, Arnaudov N
National Center for Cardiovascular Diseases, Sofia, Bulgaria

We reviewed the clinical history of 48 pts with isolated fixed subaortic stenosis (SAS) to assess probable correlation between morphology, clinical course and evolution Clinical criteria for the diagnosis and severity of LVOT obstruction were established. Criteria were selected from the clinical course, serial ECG and echocardiogram in 39 pts (range 1-11 years, mean 4 years) followed for more than 1 year. The mean age at admission was 6.6 years, range 2-15 years. In 70% of cases, the ECG showed left ventricular hypertrophy and in 30% it was normal. Two-D echo classified the following morphology at the presentation: complete ring-stenosis in 19 pts and discrete shelf (membrane or semilunar forms) in the remaining 20. Trivial aortic regurgitation existed in 26 of 39 pts (66%). The range of the LVOT peak systolic gradient on Doppler examination was 20-105 mm Hg (mean 40 mm Hg). Two types of evolution were assessed during the midterm follow-up. Nineteen pts showed rapid progression of the LVOT obstruction with clinical and ECG deterioration, 5 of them up to NYHA class II and LV strain. Pressure gradient rose from 50 to 72 mm Hg (mean). All of them had circular ring stenosis with incorporation of fibrotic tissue on the basis of anterior mitral leaflet (AML) as a main echo finding. In the remainder, the subaortic substrate seemed like homogeneous thin membrane or fibrotic ring-stenosis. Fourteen of these 19 pts were operated on and surgery confirmed the described morphology. Twenty pts formed a group with no significant deterioration of clinical, ECG and echo findings during the follow-up. The pressure gradient at the end of follow-up was found practically unchanged: from 23 to 27 mm Hg (mean). In all of them, we found discrete septal shelf without visible mitral part of the SAS or deformation of LVOT geometry. This study demonstrated the widely differing clinical histories of individual SAS during the mid-term follow-up.

P 43

Repair of tetralogy of Fallot in the first six months of life—transatrial versus transventricular repair

Stellin G, Milanesi O, Rubino M, Michielon G, Bianco R, Daliento L, Casarotto D
Departments of Cardiac Surgery, Pediatrics and Cardiology, University of Padova, Padova, Italy

Between January 1978 and October 1994, 51 pts, 22 males and 29 females, underwent correction of TOF <6 mos of age (mean age 4.8 mos). Three were neonates. Repair was accomplished through the right ventricle in 21 pts (42%), while more recently a combined transatrial-transpulmonary approach was used in 30 (58%). Transannular patch enlargement was necessary in 35 (68%), while in 4 a combined LPA patch augmentation was performed. Associated cardiac lesions as ASDII (2), multiple VSD (2) and PDA (1) were corrected simultaneously. One pt died (2%) on postop day 1 from possible major infundibular coronary branch distortion. There were no late deaths. Three pts required reoperation 6 days, 30 days and 3 yrs after repair for systemic-PA collateral ligation (1), permanent pace-maker implantation (1), fixed subAo resection + MV plasty (1), respectively. Forty-four pts are asymptomatic 2 mos to 16 yrs after repair (mean 7.3 yrs). Two-D Echo and Doppler disclosed good RV and LV function in all and absence of residual VSD in all but 1 pt with multiple VSDs. We have found a mild-moderate stenosis of LPA in 4 pts (2 of them successfully treated with balloon dilatation), of RPA+LPA in 3 (1 peripheral, treated with LPA stent insertion) and of MPA in 2. In conclusion, early correction of TOF carries low operative risk, low postoperative morbidity and good mid- and long-term survival. Transatrial-transventricular approach seems to be superior to transventricular repair because of decrease incidence of early RV failure and late RV dilatation/dysfunction. Residual LPA stenosis seems to be related to an anatomical feature of LPA in TOF rather than to surgical sequelae.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 44

Myocardial anomalies in fetal life

Fesslova V, Bellotti M, Villa L
Division of Maternal-Fetal Cardiology, "G. Adler," Department of Cardiology, ICP Milano and the Department of Obstetrics and Gynecology, University of Milan, Ospedali S. Paolo, Milan, Italy

To evaluate retrospectively the frequency and outcome of myocardial morphological and/or functional anomalies (MA) found at fetal echocardiography (F) during the 2nd level screening for congenital heart disease (CHD) in pregnancies at risk for this condition, out of 2100 fetuses at risk for CHD examined by FE (2-D and Doppler) at 16-34 weeks gestation, 51 cases presented MA, not associated with CHD (2.9% of the population and 15.9% of the total number of 320 cardiac anomalies diagnosed). Fifteen fetuses (group 1) presented a pattern of dilated cardiomyopathy (DCMP) with dilated left and/or right ventricle (LV, RV) with reduced contractility. Ten cases had probable myocarditis with both ventricles dilated (CMV infection was documented in 5); all were in heart failure. A pattern of myocardial hypertrophy, usually of the LV, with normal contractility (HCMP) with or without obstruction was observed in a total number of 36 fetuses (group 2); 9 were isolated, with familiarity for HCMP in 2/15 in association with extracardiac anomalies (ECA), mainly renal or CNS. Moreover, the LV hypertrophy with highly echodense myocardium was observed in 9 recipient twins with twin-to-twin transfusion syndrome (TTTS). Out of fetuses with DPMC, 12/15 (80%) died *in utero* and 3 after birth. Two cases with myocarditis survived, and in one case the pregnancy was terminated. From group 2, 5/9 fetuses with isolated HCMP died and 4 are stable at 6 mos-3 yrs. Eight of 15 fetuses with ECA had termination, 2 died *in utero*, 3 after birth and 5 are alive, with subsequent normalization of the pattern within 6 mo. In conclusion, our data show a poor prognosis for fetuses with DCMP, while variable entities of HCMP were observed *in utero* with different outcome. A relatively high frequency of secondary forms was observed and a rapid development of cardiac hypertrophy was documented. The possibility of spontaneous regression in secondary forms is confirmed.

P 45

Early mortality in infants with hypertrophic cardiomyopathy—1969-1993

Skinner JR, Martin RP, Hayes A, Joffe H, Jordan SC
Royal Hospital for Sick Children, Bristol, UK

Mortality in infants presenting in the first year of life with hypertrophic cardiomyopathy (HCM) has previously been reported to be roughly 50%, but our clinical impression has been that it is lower. Using the UK South West regional cardiac database, we reviewed the mode of presentation, associated features, and mortality among all infants (<1 year) presenting with HCM between 1969-1993. Sixty cases were identified. Nineteen (32%) had some (mostly minor) associated congenital heart disease. Thirteen (22%) were infants of mothers with diabetes during pregnancy. Nine (15%) patients (pts) had Noonans' syndrome, one had trisomy 18, and one had generalized muscle disease. Incidence increased over the study period—until 1978, 10 pts; '79-'83, 7 pts; '84-'88, 14 pts; and '89-'93, 29 pts. Mode of presentation was analyzed in the 57 cases with adequate documentation: cardiac failure (29, 51%), heart murmur (17, 30%), cyanosis (5, 9%), family history (4, 7%), other (2, 4%). Twenty-three pts (40%) presented in the first 24 hours (including all 5 pts with cyanosis), 16 pts (28%) between 2 and 28 days, and 18 pts (32%) after 28 days. Excluding infants of diabetic mothers (one of whom died), there were 8 deaths; 6 were cardiac related (13%); all 6 were due to progressive cardiac failure; 5 occurred within in the first year, and 1 at 4 yrs. Of all the infant deaths, 5 occurred before 1986 (5/21, 24%), and 1 after 1986 (1/39, 3%), $p<0.05$. In conclusion, early mortality in infants with HCM is much lower than previously reported and is falling. This may be related partly to increased recognition of more minor forms, and possibly also due to the success of aggressive medical management including ß-blockade.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 46

Tachycardia-induced cardiomyopathy in children—treatment and prognosis

Villain E, Iserin L, Aggoun Y, Bonnet D, Kachaner J
Hôpital Necker, Enfants-malades, Paris, France

Dilated cardiomyopathy (DCM) may be induced by prolonged non-sinus tachycardia only. The aims of this study were to assess the reversibility of ventricular dysfunction by controlling the arrhythmia, and to establish a therapeutic strategy. We reviewed 12 patients (pts) referred for heart failure beyond the neonatal period at 8-154 months, with 6 infants <2 years, and 6 children >6 years. Echocardiography showed normal heart structures and severe DCM. Heart rate ranged from 150 to 300 bpm and was due to atrial ectopic tachycardia in 6 pts, reentrant tachycardia over accessory pathways in 3, and ventricular tachycardia in 3. The oldest pt underwent radiofrequency ablation of an accessory pathway. All other were given antiarrhythmic drugs, oral amiodarone being our first line therapy at a loading dose of 500-1000 mg/m^2. It was effective in 10 pts, alone or in association with propranolol, but one died of ventricular tachycardia despite prompt return to sinus rhythm. It failed in the last pt who was successfully treated with flecainide. Echocardiographic parameters returned to normal within 1-12 months (median 6 months) after restoration of a stable sinus rhythm, except in the pt who was ablated and who still has ventricular dilatation 14 months after the procedure. At follow-up (11-90 months, median 49), 7 patients have been weaned from therapy, but 3 (with atrial tachycardia) are still on beta-blockers or flecainide. In conclusion, 1) children with 'unexplained' DCM should be investigated for chronic non-sinus tachycardia; 2) the prognosis of tachycardia-induced DCM is good as control of the arrhythmia leads to resolution of the left ventricular dysfunction; 3) drug therapy, including amiodarone, is very effective and should be considered as the treatment of choice since young children may grow out of their arrhythmias. Ablation should be indicated only when antiarrhythmic drugs have failed or in teenagers who have DCM due to atrioventricular reentry.

P 47

Hypertrophic cardiomyopathy—more severe course in Noonan syndrome?

Kehl HG, Kececioglu D, Pröse K, Nekarda T, Gehrmann J, Kotthoff S, Asfour B, Vogt J
University of Münster, Münster, Germany

The purpose of this study was to determine the incidence of hypertrophic cardiomyopathy (HCM) in Noonan syndrome (NS) and to compare its clinical course with that of familial HCM. During a period of 23 years, 201 patients were suspected to have NS or familial HCM. The diagnosis of HCM was based on echocardiographic, angiographic or pathologic criteria and exclusion of systemic hypertension. Familial HCM was assumed by familial history and by exclusion of known conditions due to secondary HCM. The diagnosis of NS was made in the presence of typical somatic features according to previously published criteria. The diagnosis of NS or familial HCM could be confirmed in 124 patients. NS was diagnosed in 69 (40 male, 29 female patients; age at diagnosis 1 week to 27 years, mean 7.9 years). Heart defects were found in 49 patients (71%) with 81 major cardiac lesions (pulmonary stenosis in 28 of 69 (40%); septal defects in 25 (36%); aortic stenosis and patent ductus arteriosus in 3 (4%) respectively; pulmonary atresia and isolated anomalous of pulmonary venous connection in 2 (2%) respectively). NS with HCM was present in 13 of 69 patients (18%) (7 male, 6 female); 9 (13%) had an obstructive form (HOCM), 4 (5%) had an nonobstructive form (HNCM). Familial HCM could be confirmed in 55 patients (28 male, 27 female; age at diagnosis 2 weeks to 18 years); HOCM was present in 54 (98%), HNCM in 1 patient. In NS severe cardiac symptoms occurred significantly earlier with HOCM compared to familial HOCM; neonatal diagnosis was made in 4 of 9 HOCM with NS versus 2 of 54 familial HOCM ($p<0.001$). Deaths occurred in 5 of 9 HOCM with NS versus 6 of 54 familial HOCM and versus 2 of 60 NS without HOCM ($p<0.001$ respectively). We conclude that NS has a high incidence of HCM. HOCM was the most important risk factor in the outcome of NS.

P 48

Early identification of anthracycline cardiomyopathy—possibilities and implications

Bu'Lock FA, Mott MG, Oakhill A, Martin RP
Bristol Royal Hospital for Sick Children, Bristol, UK

Data from 451 serial echocardographic studies performed during treatment of 125 children for malignant disease were examined for early markers of functional status at end-of-treatment (END). They were treated to total anthracycline doses of 45-1150 (mean 320) mg/m^2. Cardiac function was assessed from the left ventricular (LV) shortening fraction (SF). A SF of <30% is associated with a significant risk of cardiac decompensation. SF fell with increasing cumulative anthracycline dose at an average rate of 1% per 100 mg/m^2 ($r= -0.29$, $p<0.0001$). Twenty-four patients (19%) had SF <30% by the end of treatment; the incidence of abnormal SF rose with increasing anthracycline dose ($p=0.0001$). When data from patients with abnormal ENDSF at end-of-treatment were compared with those with ENDSF>30%, potentially important differences in LV function earlier in treatment were apparent. Median and interquartile range (IQR) SF for patients in 100 mg/m^2 dose groups through treatment, subdivided by SF at end-of-treatment, show marked divergence in SF between the two groups from early in treatment, largely due to changes in LV systolic diameter rather than diastolic dilatation. Median rate of fall of SF was also significantly steeper in those with ENDSF <30% (2%, 1QR 1 to 3) per 100 mg/m^2 than those with ENDSF >30% (1%, -0.8 to 3) per 100 mg/m^2 ($p<0.01$). Regular monitoring of LV shortening fraction during anthracycline treatment can identify patients at higher risk of important cardiotoxicity (eg, with SF<32% at >200 mg/m^2, or perhaps a fall in SF of >2-3 absolute percent per 100 mg/m^2). Modification of therapy and/or use of cardio-protective agents might be indicated in these patients. However, pretreatment and frequent low dose scans are required, which has significant resource implications.

P 49

Left ventricular systolic dysfunction late after anthracycline therapy for childhood cancer

Rübo J, Klement D, Krogmann ON, Heusch A, Friedland C, Körholz D, Goebel U, Bourgeois M
Pediatric Cardiology and Pediatric Oncology, Heinrich-Heine University, Düsseldorf, Germany

An echocardiographic study was designed and 52 pts (age 16.3±2.3 years) were reevaluated 6.9±2.5 years after completion of anthracycline-containing combination chemotherapy. Pts were grouped by their underlying disease: leukemia in 34 pts (P1) and sarcoma in 18 pts (P2). All pts were in functional NYHA class I. Standard M-mode echocardiography was performed simultaneously with the carotid pulse tracing. Blood pressure was measured by the oscillographic method. LV end-systolic wall stress (Ses) was calculated according to Brodie. Data were compared to a control group of 25 healthy volunteers (C). P2 had a higher anthracycline dose than P1 (367 vs. 235 mg/m^2; $p<0.001$). LV end-diastolic diameter, LV muscle mass index and Ses were not different between patients and controls. For all groups, a significant correlation between Ses and velocity of circumferential fiber shortening (Vcf) was found. There was a downward displacement of the Ses-Vcf relationship in pts compared to controls. Vcf for a given Ses was significantly lower in P1 and in P2 than in C (-0.23 l/s in P1 and -0.30 l/s in P2 vs 0.01 l/s in C; $p<0.001$). There was also a significant correlation between the anthracycline dose and lowering of the Vcf for a given Ses ($r=0.58$, $p<0.001$). Fifteen pts showed LV systolic dysfunction (2 SD below control value): 21% of P1 and 53% of P2 ($p<0.05$). Pts with depressed LV function had received a higher mean dose of antracyclines; however, in 4 the dose was below 200 mg/m^2. In conclusion, LV systolic dysfunction is seen 6.9 years after anthracycline treatment in 20-50% of asymptomatic pts. There is a correlation between dose and depression of LV function and pts with sarcoma and larger doses of antracyclines are at a higher risk. But even pts after a low dose (<200 mg/m^2) may have LV systolic dysfunction late after antracycline treatment.

P 50

Ethical impacts of interventional pediatric cardiology—how to accept morbidity and mortality?

Hausdorf G, Schneider M, de Vivie J, Distler B, Goeldner B, Gohlisch K, Kampmann C, Kargus K, Will J, Herweg V, Konertz W
Charité, Pediatric Cardiology and Cardiac Surgery, Berlin, Germany

With increasing experience and development of novel techniques, the number of children with congenital heart disease susceptible to transcatheter therapy is steadily increasing. While a surgical hazard of death, chronic disability/morbidity and the morbidity of thoracotomy and extracorporal circulation are generally accepted, these hazards are an ethical problem for the pediatric cardiologist. We analyzed the 275 interventional procedures between 10/93 and 11/94 for their hazards and compared them with the estimated surgical hazards. The estimated surgical risk was calculated for each intervention (VSD-closure 1%, pulmonary atresia 5%, coarctation repair 2% etc.), resulting in an overall hazard of death for surgery of 2.06% compared with an transcatheter hazard of 0.35%. The increased risk of reoperations (12%) was not taken into account. Complications occurred in 2.52% of the interventional procedures, however, morbidity due to complications in only 1.8%. Acute surgical intervention was necessary in 0.72%. It is concluded, that in experienced centers the hazards of extended transcatheter therapy can be lower than that of surgery.

P 51

Multitrack—a versatile and less costly approach for dilation of mitral stenosis in children

Bonhoeffer P, Piechaud JF
Hopital Necker/Enfants-Malades, Paris, France

Mitral stenosis in pediatric age is uncommon in the developed countries but not rare in countries where rheumatic heart disease persists. Over the last years, percutaneous mitral valvotomy has largely replaced surgical intervention in the treatment of mitral stenosis in the adult population. The most frequently used catheters are the Inoue and Bifoil. We developed a new technique (Multitrack) with two separate balloon catheters positioned on a single guidewire. The first catheter with only a distal guide wire lumen and a proximal balloon is first introduced over the guidewire into the vein and positioned in the mitral valve orifice. Subsequently, a normal balloon-catheter, running on the same guide wire, is inserted and lined up with the first catheter in order to position the two catheters side by side. Then the balloons are inflated contemporarily, similar to the other double balloon techniques. This technique was applied in 4 children with severe rheumatic mitral stenosis between 10 and 17 years of age (mean 13) and 24-54 kg (mean 34). Valve area increased from 0.6 cm^2 (0.3-0.9 cm^2) to 2.0 cm^2 (1.8-2.3 cm^2) and mean left atrial mean pressure dropped from 31 mm Hg (18-52 mm Hg) to 10 (7-20 mm Hg). The balloon choice was 15+15 mm in two cases, 15+18 in one and 18+18 in one. Mitral dilatation with the Multitrack system gives the possibility to choose smaller balloons giving comparable results to the previously described techniques. The catheters are much simpler and less costly. It is more versatile than the procedure with double balloon on a single shaft and is technically easier than the approach with two balloon catheters on two separate guidewires.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 52

Balloon dilatation of aortic valve stenosis with transesophageal echocardiography

Gildein HP, Kleinert S, Edis BD, Goh TH, Menahem S, Wilkinson JL
Department of Cardiology, Royal Children's Hospital, Melbourne, Australia

Balloon dilatation for aortic valve stenosis is still controversial, and severe aortic regurgitation as a complication of the procedure has been reported. To minimize the risk of regurgitation and to maximize the result by using gradational balloon dilatation with oversized balloons when necessary, we used transesophageal echocardiography (TOE). The intention of TOE was to confirm the correct position of the stiff dilatation wire and to estimate aortic regurgitation. If the reduction of gradient was not sufficient, and there was no increase in aortic regurgitation on TOE, a larger (and even oversized) balloon catheter was used. This technique was employed in 8 children (age 3-11.5 years). Initially, the ratio of balloon diameter to aortic valve diameter was 85-90% in 3 patients. For the remaining 5 patients the ratio was 100-130%. Up to 4 different balloon sizes were used. TOE, in all cases, showed correct position of the dilatation wire. Aortic regurgitation was absent or mild and did not increase following the procedure. The gradient decreased from 60±18 to 35±11 mm Hg ($p<0.01$). Balloon valvuloplasty of aortic valve stenosis may be more effective under TOE which can delineate increasing regurgitation. With this technique it may be safe to use oversized balloons when gradational balloon dilatation is performed.

P 53

Detachable balloons for closure of congenital coronary fistulas and small ductus arteriosus

Meijboom EJ, Hutter PA, Mast EG, Ernst JMPG, Overtoom T, Borman F, Plokker HWM, Harinck E
Departments of Cardiology, Wilhelmina Children's Hospital, Utrecht and St Antonius Hospital, Nieuwegein, The Netherlands

Detachable balloons (DB) play an important role in radiological interventions. In this study, these DBs (Nycomed) are used in the cardiovascular setting for the closure of coronary fistulae and a small, but elongated PDA. Surgical closure of congenital coronary fistulae can be difficult and tedious with a success rate of less than 100%. Transcatheter techniques to close these coronary fistulae seem an appropriate alternative. In 5 pts (6 procedures) with congenital coronary fistulae, transcatheter DBs were used. One patient had a large right coronary to right ventricular fistula through an aneurysmal sack, requiring a combination of coils and a DB to obliterate the fistula. The 4 other pts had coronary to pulmonary artery fistulae and required several coils to be delivered initially through selective engagement of the coronary artery. In all 5 pts, flow through the fistulae was strongly reduced to hemodynamically insignificant proportions immediately following the procedure and total closure developed subsequently. ECGs in one patient showed a transient ST-T segment change during the procedure, although pre-procedure balloon occlusion (5 min) showed no ECG changes. No complications were encountered during or after the procedures. In 1/36 pts with PDA, the aortic ampulla was long and narrow, preventing the placement of a Rashkind occluder system. From the pulmonary artery side, a DB could be safely deposited and remained in perfect position after detachment. No residual shunting was encountered. We conclude that selective closure of congenital fistulae and narrow PDAs with DBs is a safe and promising treatment, warranting serious consideration when treatment is discussed.

P 54

Intermediate-term results of balloon dilation of isolated aortic stenosis in the first year of life

Carminati M, Giusti S, Spadoni I, Borghi A, Bonhoeffer P
Ospedale Pediatrico Apuano, Massa and Ospedali Riuniti, Bergamo, Italy

The results of balloon dilation or surgical aortic valvotomy in infancy are greatly influenced by the severity of associated anomalies. The aim of this study is to evaluate the outcome of 25 patients with isolated aortic stenosis who underwent balloon dilation from April 1989 to September 1993. The age ranged from 1 to 319 days (mean 67 days) and the weight from 2.1 to 10 kg (mean 4.5 kg). Nine (36%) were in the first week of life. Eleven (44%) presented with severe cardiac failure. Morphology of the valve was bicuspid in 17 cases, tricuspid in 6, undetermined in 2. One patient died (4%) from complications related to the procedure. All other patients had a very good immediate result with significant reduction of the gradient (from 85±32 to 27±16 mm Hg) and an increase in left ventricular ejection fraction (from 51±25 to 67±12%). In no cases did balloon dilation produce severe aortic regurgitation. At follow-up (13 to 54 months), all patients are doing well without any therapy. Aortic restenosis occurred in 3 cases (12. 5%)—1 was treated by surgical valvotomy and 2 by repeat balloon valvotomy; in the other 2 cases, a subvalvular aortic obstruction developed and was relieved by surgical resection. There were no significant changes in the degree of aortic insufficiency during the follow-up period. No late mortality was observed. Based upon our results, the balloon dilation can be considered as the first choice procedure for treatment of isolated aortic stenosis in infancy.

P 55

Preliminary experience with PDA closure using thoracoscopy

Borini I, Cervo G, Calza G, Camassa N, Gorrieri F, Dolcini G, Bertolini A, Zannini L
Departments of Cardiovascular Surgery and Anesthesiology, Institute G. Gaslini, Genova, Italy

In 1993, Laborde and colleagues reported an innovative technique for PDA closure, using video thoracoscopy. The advantages of this method are: 1) mini-invasivity (three small holes in the thoracic wall instead of thoracotomy, with complete muscle sparing); 2) versatility, almost all non-complicated PDA, irrespective of size and length, can be closed; 3) efficacy, instantaneous and complete PDA closure; 4) absence of post-surgical pain (patients begin to move and breath normally after a few hours); and 5) good cost-to-benefit ratio, the procedure is very inexpensive and utilizes the same hemoclips as in the surgical approach. The drawbacks could be: 1) high initial cost for video thoracoscopic instrumentation; and 2) the risk of emergency thoracotomy for major bleeding. In 1994, we performed 5 thoracoscopic PDA closures (age of the patients 8 mos-8 yrs, 35.2±6.6 mos; weight 6-22 kg, 13±4.3). All cases were diagnosed by two-D echo color Doppler. PDA closure was controlled by two-D echo immediately after the procedure. The operation was successful in all cases, without any morbidity nor mortality. Thoracoscopic PDA ligation can be considered as a valid method when compared with current surgical and cardiological approach to this problem.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 56

Percutaneous fenestration of the atrial septum with a stent—an experimental study

Gewillig M, Mertens L, Stockx L
University Hospital Gasthuisberg, Leuven, Belgium

A restrictive communication between the systemic venous and pulmonary venous atrium in a Fontan circulation has been shown to reduce early mortality and morbidity. Such a fenestration reduces systemic venous pressure and improves cardiac output at the expense of a slight arterial desaturation. A secondary fenestration created with balloon dilatation has a high tendency to close over a period of several months. This study was set out to evaluate the feasibility, safety and efficacy of creating a stented fenestration of the atrial septum in mongrel dogs. After puncturing with a Brockenbrough needle, the atrial septum was crossed with a 7 Fr Mullins sheath. A self-expandable wallstent was deployed across the septum. The animals were sacrificed after 0-12 weeks (mean 6.5±4.9 weeks). Macroscopic and microscopic examination was performed. In 3 dogs, the septum could be punctured in its mid-portion; in all 3, the stent was deployed symmetrically across the septum. In 1 dog, the septum was crossed in a more posterior position but stent deployment was symmetrical. When an 8 mm stent was used, the final fenestration was of a clinically desirable size (3.7±0.9 mm); a 10 mm stent resulted in a too large an opening (7±2.8 mm). No thrombi or peal formation could be observed in these 4 stents during the follow-up period. In 2 dogs, poor positioning of the stent was obtained with the stent being too high posterior and asymmetrical across the septum. This resulted in peal formation either because of low flow or because of subobstruction of the stent. We conclude that if the septum can be punctured in its mid-septal portion, a wallstent can safely be deployed. Over a period of 0 to 12 weeks, no adverse effects such as thrombi or peal formation were observed in well-deployed stents.

P 57

The adjustable buttoned device for the occlusion of patent ductus arteriosus—international experience

Rey C, Walsh K, Kim SH, Worms AM, Wang HC, Leung M, Rao PS, Sideris E
Pediatric Cardiology, Lille, France

One hundred-twenty cases of patent ductus arteriosus (PDA) occluded by the adjustable buttoned device were performed to October 1994. They included cases of conical, tubular, and short ductus as well as Rashkind occluder residual shunts.

					Occlusion	
n	Duct (mm)	Device (mm)	Age (yrs)	Weight (kg)	24 hours	1 month
120	2-15	15-35	0 9-58	6 5-62	69%	80%

All devices were introduced through 7-8 Fr sheaths. There was one unbuttoning, no hemolysis, no device embolization, no left pulmonary artery stenosis or aortic stenosis were noticed. In 5 patients, devices were pulled through the ductus in the pulmonary artery and they were either replaced by others (3) or sent to surgery (2). The adjustable buttoned device appeared effective and safe for most types and sizes of PDA with the exception of infants. The newly introduced infant PDA device is expected to address the infant age group and improve on the full occlusion rate of the adjustable buttoned device.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 58

Transcatheter occlusion of atrial septal defects in pigs using the ASDOS-device

Schneider M, Babic UU, Bloch-Thomson A, Hasenkam M, Bagger JP, Hausdorf G
Charité, Berlin, Germany and University of Aarhus, Aarhus, Denmark

Several devices are under investigation for the transcatheter occlusion of atrial septal defects. The ASDOS device is made of nitinol, a memory metal, which is covered by a microporous polyurethane-membrane. Two umbrellas in each atrium are connected with each other using a screwing mechanism. The applicability and biocompatibility of the device were investigated in 12 pigs. After transseptal puncture of the fossa ovalis, an atrial septal defect was created by blade-septostomy and balloon dilation of the defect. An arteriovenous loop from the femoral vein through the defect to the femoral artery was established and the device implanted using a screwing catheter. In one pig a large Eustachian valve was caught by the right-sided umbrella, resulting in incomplete occlusion of the inferior vena cava. After training on the technique in 4 pigs, implantation of the device was performed without complications. Even after screwing them together, both umbrellas can be retrieved. Two and 3 months after the implantation, the pigs were sacrificed; the polyurethane was found to be completely covered with fibrous tissue.

P 59

Myocarditis in children—clinical and immunohistological findings, influence of therapy

Heusch A, Kühl U, Rammos S, Schultheiss HP, Rubo J, Bourgeois M
Departments of Pediatric Cardiology and Internal Medicine-Cardiology, Heinrich-Heine University, Düsseldorf, Germany

The purpose of the study was to compare the sensitivity of common hematoxylin-eosin (HE) staining and immunohistological methods in myocarditis and to assess the influence of steroid therapy on the outcome of the disease. From 1991 to 1994, endomyocardial biopsies were performed in 33 children (mean age 4.7 years, range 0.1-18) with suspected myocardial disease. Biopsies were examined after HE-staining and after incubation with various antibodies marking the specific lymphocytic surface antigens CD2, CD3, CD4, CD8 and HLA-expression. In 16 patients with acute onset of the disease, immunohistological techniques proved severe lymphocytic infiltration. CD2/CD3 was positive in all of them; HLA-expression increased in 14 patients. Leading clinical manifestations were severe reduction of myocardial function (8) (NYHA 2-3), 2-3° AV-conduction disturbance (5), recurrent pericardial effusion (1), ventricular tachycardia (1) and changes of repolarization (1). Serological markers detected mycoplasma pneumonia only in 1 patient. Seven patients with impaired hemodynamics and persistent inflammation were treated with prednisone (12 mg/kg/d). Additional medication with diuretics, digoxin and vasodilators was necessary in 6. Six of 8 patients showed clinical, improvement and disappearance or reduction of infiltration in biopsies after prednisone therapy (duration 2-12 months). In contrast, 5/6 patients without therapy who had biopsies, showed persistence of the infiltration even if clinical symptoms improved. Two patients died, one with severe diastolic dysfunction during prednisone therapy (10 days), a second patient (ventilated, NYHA 4) before the histological diagnosis was proven. In conclusion, immunohistological techniques can prove lymphocytic myocardial infiltration, even if HE-staining is negative. Prednisone can reduce inflammatory infiltration and seems to have a supportive effect on the clinical course, especially if LV-function is impaired.

P 60

Severe heart failure as a predictor of poor outcome after neonatal balloon aortic valvuloplasty (BAV)

Cazzaniga M, Abraira V, Fernández Piñeda L, Herraiz I, Bermúdez R, Quero M, Maitre M, Quero M
Hospital Ramón y Cajal, Madrid, Spain

The influence of the clinical picture and 2-D echo variables on survival were retrospectively analyzed in 27 neonates who underwent balloon therapy at 18±3.7 days of life. Age, weight, gender, ductal patency (Duct), presence of Ao coarctation (AoC), left ventricular diastolic diameter (LVDD), shortening fraction (SF), aortic "annulus" (AoA), balloon/"annulus" ratio (B/A) were measured in all pts. Cardiac failure (CF) was coded as 0 (absent), 1 (mild), and 2 (severe: drugs + ventilatory support). In addition, mitral annulus (MA), ascending aortic dimension (AAoD) and LV area were categorized in 19 of the 27 pts. The immediate and mid-term (30±17 months) results of BAV on the LV/Ao gradient were measured by Doppler method. Wilcoxon and Fisher's exact tests were used to compare means and proportions; unconditional logistic regression ("backward" strategy) was applied to define a model to predict the outcome after BAV. Seven pts died (26%), and 1 of the 2 pts with severe aortic regurgitation required a mechanical prosthesis. The LV/Ao gradient decreased from 70±30 to 25±17 mm Hg ($p<0.01$) after BAV, and did not change in the follow-up period. Severe CF (grade 2) was an independent factor with high influence on the mortality ($p<0.001$). The odds ratio to CF 2 was 36 (1.8-732 CL, $p<0.019$) while the odds ratio to CF1 was 1.8 (0.13-23 CL, $p<0.658$). Others variables that individually affect the success of the BAV were: presence of AoC and Duct patency, LVDD <12 mm, LV area <2.0 cm^2, MA <9 mm, AoA <6.7 mm and AAoD <7.9 mm. Four of the 5 pts (80%) with CF grade 2 had the lowest 2-D echo left heart dimensions. Severe heart failure as a result of the most profound compromise of the left heart structures in the setting of severe/critical AoS is a predictor of poor outcome when the balloon therapy is planned. In this debatable modality of treatment, neonates without cardiac failure or with mild impaired left cardiac function emerge as the better candidates for balloon valvuloplasty.

P 61

Incessant ventricular tachycardia in children—prognosis and follow-up

Vignati G, Drago F, Mauri L, Guccione P, Aragonese P, Figini A
Cardiologia Pediatrica, Ospedale Niguarda, Milano and Ospedale Pediatrico Bambino Gesù, Roma, Italy

The aim of this study was to assess the prognosis and clinical course of incessant ventricular tachycardia (VT) in a group of consecutive 11 children (mean age 8±4 yrs) admitted to our institutions between 1983-1993. All patients (pts) had frequent (more than 80% of any day) non-sustained VT runs. No child had demonstrable heart disease and 5 pts were symptomatic for palpitations. VT was monomorphic with left bundle branch block and inferior axis in all. The VT mean rate was 132±20 bpm. In all but 1 pt, VT disappeared with exercise and returned during recovery. Cardiac catheterization was normal in all but 1 child who presented with dilation of the right outflow tract. The same pt had an increase in epicardial fat at MRN evaluation. MRN controlled in 5 pts was normal in 4. VT arose (intracardiac mapping 8/11 pts) from right outflow tract in 7 and from superior interventricular septum in 1. Chronic treatment was started in 8/11 pts, effective in half of them. The most successful drug was amiodarone. During follow-up (52±10 mos), VT disappeared in 4 children; the others had no variation of arrhythmia and remained asymptomatic. In conclusion, incessant VT seems mostly to arise from the right outflow tract, rarely to be an expression of right ventricle dysplasia, to be often insensitive to medical treatment and to have a good mid-term prognosis.

P 62

Pulmonary artery banding and arterial switch for failed atrial switch procedures for transposition of the great arteries echocardiographic observations

De Giovanni JV, Stümper O, Silove ED, Brawn WJ
Birmingham Children's Hospital, Birmingham, UK

Atrial switch (AtS) operations for transposition of the great arteries provides long-term palliation in the majority but fails in some patients due to progressive right ventricular failure and tricuspid regurgitation. Current surgical practice has replaced the AtS by the arterial switch (ArtS) procedure as this is more physiological. Ten patients who have previously had an AtS were evaluated because of right ventricular dysfunction. Eight male and 2 female patients aged between 10 and 25 years (mean 19.3 years) were investigated prior to and following pulmonary artery banding designed to train the left ventricle prior to a planned ArtS and take-down of AtS. Pulmonary artery bands were successfully placed in 9 patients, 3 of whom have proceeded to an ArtS. Ventricular dimension, septal and left ventricle posterior wall thickness and motion were evaluated by echocardiography. Intraoperative transesophageal echocardiography at the time of pulmonary artery banding was performed in some patients and proved helpful in determining the tightness of the band. Preliminary results indicate that older patients respond less favorably to pulmonary artery banding due to lack of left ventricular hypertrophy.

P 63

Balloon angioplasty of native coarctation of the aorta, immediate and long-term results

Thanopoulos BD, Anagnostakis JS, Tsaoussis GS, Stinios JJ, Simeunovic SS
"Aghia Sophia" Children's Hospital, Athens, Greece and University Children's Hospital, Belgrade, Yugoslavia

Several investigators have reported early and intermediate results following balloon angioplasty (BA) of native coarctation of the aorta (Na-CoA). However, there are little data available from long-term follow-up of these patients. We studied 35 pts who had undergone BA for Na-CoA, at a median age of 3.6 years (range 2 days to 14 years). Length of FU (clinical examination, Doppler ultrasound, magnetic resonance imaging) ranged from 24 to 84 months (median 48.2 months). Repeat cardiac catheterization was performed in all pts 6 months to 4 years after BA. Mean peak systolic pressure gradient (MPSPG) across Na-Coa decreased from 43.4±10.3 mm Hg before BA to 8.5±6.9 mm Hg immediately after the procedure. Mean diameter of the stenotic aortic isthmus increased from 3.6±2.2 to 8.9±3.6 mm following initial angioplasty. There was no mortality. Aneurysm formation occurred in 2 pts (5.7%). Restenosis occurred in 10 (28.5%) of the 35 pts (6/9 neonates, 3/11 infants and 1/15 children) and was treated successfully either surgically (5 pts) or by repeat BA (5 pts). MPSPG across the area of the coarctation was 8.4±4.7 and 9.2±5.0 mm Hg after final angioplasty and at FU, respectively; 91.6 and 84.3% of the pts are presently normotensive at rest and at exercise, respectively. In conclusion, balloon angioplasty offers an effective means of relieving native coarctation with low morbidity and immediate results maintained on long-term follow-up. Restenosis usually occurs after neonatal balloon angioplasty.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 64

Percutaneous closure of ostium secundum with Sideris device—echocardiographic follow-up

Lambert V, Losay J, Chevalier B, Piot JD, Brenot P, Angel C
Centre Chirurgical Marie Lannelongue, Le Plessis-Robinson, France

Efficacy and safety of atrial septal defect (ASD) percutaneous closure has been evaluated in 20 consecutive patients (pts) who had 21 umbrellas placed between June 1992 and June 1994. Median age was 19 years and median weight 46 kg. Median stretched size of ASD was 20 mm and median device/umbrella ratio 2.31. Each pt had aspirin for 6 mos. The umbrella was assessed by chest x-ray, transthoracic echocardiography (TTE) and, at the last follow-up (FU), by direct vision during surgery or transesophageal echocardiography (TOE). No patient was lost to FU. Immediately, unbuttoning occurred in one patient and immediate migration in another; a shunt was absent in 13, ≤3 mm in 2 pts, and >3 mm in 3 pts. The device was well seated in all pts but one. At 3 days, the shunt increased and was larger than 3 mm in 7 pts. At one month FU, 9 pts had a shunt larger than 3 mm, and on TTE, the device was inadequately positioned in 6 pts. At last FU, displacement was evident on TOE or surgery in 10 pts. One of these pts had a massive systemic embolization at 15 mos. This patient and 4 others had surgical closure of the residual ASD, and in another pt a left atrial thrombus was discovered. Perfect closure was obtained in 35% of the pts, but the Sideris device can be partly displaced early after ASD closure, causing significant residual shunt in 35% of the pts and possibly thrombus formation. TOE is more accurate to visualize the placement of the device. Unsatisfactorily positioned devices should be retrieved.

P 65

Coil vs umbrella vs surgical closure of the small patent ductus arteriosus

Dahdah N, van Doesburg NH, Guérin R, Chartrand C, Fouron JC, Miró J
Hopital Sainte-Justine, Montreal, Canada

The aim of this study is to compare the three techniques used in our institution in the treatment of small (<3.0 mm minimal diameter) patent ductus arteriosus (PDA). From 9/92 to 10/94, 14 patients (wt 5.9-53.4 kg) underwent surgical treatment (Sx), 13 patients (wt 10.8-50.4 kg) had a 12 mm Rashkind umbrella placed (Um) and 9 patients (wt 8.8 to 20 kg) underwent embolization with Gianturco coils (Co). The procedure was successful in all cases, although one Um patient required a second attempt. Complications occurred in 1/13 Um patients (device embolization to LPA, successfully retrieved percutaneously), in 2/9 Co patients (coil embolization to femoral arteries both retrieved, one of them resulting in femoral artery thrombosis, completely recanalized with thrombolytic therapy) and in 2/14 Sx patients (chylothorax in one, prolonged intubation due to atelectasis in one). Hospital stay was significantly longer in the Sx group (5.4±3.6 days, range 3-18) than in the Umb or Co groups (2.15±0.53 and 2.2±0.63 days respectively). Total hospital cost per patient was comparable for the Sx (6199±3222 CanD) and the Um groups (5667±1973 CanD) and significantly lower for the Co group (3015±513 CanD). Small or trivial shunts at last echocardiographic follow-up were found in 4/13 Um patients, in 1/9 Co patients and 0/14 in Sx patients. We conclude that both percutaneous techniques are safe, reliable and cost-effective alternatives to surgical treatment of small PDA, with Gianturco coils being particularly attractive.

P 66

Late systemic desaturation after total cavopulmonary shunt

Stümper O, Wright JGC, Sadiq M, Silove ED, Sethia B, Brawn WJ, deGiovanni JV
The Children's Hospital, Birmingham, UK

The creation of a total cavopulmonary shunt (TCPS) offers very good palliation in pts with left atrial isomerism, azygos continuation of the IVC and complex CHD, who are not suitable for biventricular repair. However, long-term results have not yet been reported. Six patients underwent detailed follow-up 3-10 years (mean 4.7 yrs) after TCPS operation. Oxygen saturations (O_2 sats) postoperatively ranged from 89-92% (mean 90%) and decreased to 73-81% (mean 77%) at last follow-up ($p<0.05$). During stress testing (mean 6.7 min), O_2 sats decreased to 62-87% (mean 71%) and during recovery to 68-85% (mean 78%). One pt had a rise in O_2 sats both during exercise and recovery, reflecting ventilation/perfusion mismatch. The other 5 pts with further decrease in O_2 sats underwent detailed catheterization and angiography. No pt had pulmonary artery obstruction or pulmonary arteriovenous fistulae. All four pts (67% of study population) with a drop in O_2 sats of more than 5% during peak exercise (compared to resting values) were found to have systemic-to-portal venous fistulae. These were infradiaphragmatic in 3 and supradiaphragmatic in 1. Fistulae were successfully occluded using detachable balloons (2) or coils (1). Occlusion failed in 1 pt with numerous small fistulae. Oxygen saturations post-procedure improved by 6-10% at rest. (Pre-occlusion range 73-80%, mean 77%, postocclusion 79-88%, mean 84%). Patients with interrupted IVC who underwent a total cavopulmonary shunt operation are at risk to develop sytemic-to-portal venous fistulae. Excercise stress testing allows identification of such pts. Angiography should also assess the infradiaphragmatic systemic veins. Transcatheter occlusion of fistulae provides very good symptomatic relief and thus should be preferred to reoperation.

P 67

WEB 2170 inhibits platelet-activating-factor induced pulmonary hypertension in fetal pigs

Grabitz RG, Coe JY, Chen R, Timinsky J, von Bernuth G
Pediatric Cardiology, University of Alberta, Edmonton, Canada and RWTH Aachen, Aachen, Germany

Elevated platelet-activating-factor (PAF) levels are found in neonates with persistent fetal circulation (PFC). We evaluate the role of PAF and PAF-inhibition by WEB 2170 on fetal circulation using near-term fetal pigs. Under maternal GA, fetal piglets were acutely instrumented with an EM-flow probe around the left pulmonary artery (LPA) and catheters in the PA, aorta (Ao), right and left atria (RA, LA). Part A: PAF (1 to 10 ng/kg) was given over 15 sec in PA and its hemodynamic effect monitored for 15 min. Part B: WEB 2170 (1 mg/kg) given over 15 sec in RA, PAF (5 ng/kg) added at 30 min and hemodynamic effects monitored for an additional 5 min. Results of calculated arteriolar resistance (percent changes ± SEM, time post-PAF) are listed in the table.

PAF (ng/kg)	n	0 5 min	1 min	2 min	5 min	10 min	15 min
1	4	2 ± 5	3 ± 5	-2 ± 6	4 ± 5	0 ± 6	4 ± 3
2 5	2	4 ± 5	25 ± 10	20 ± 12	8 ± 5	5 ± 10	-2 ± 7
5	6	38 ± 22	112 ± 34	170 ± 48	51 ± 27	20 ± 18	5 ± 15
7.5	3	38 ± 19	125 ± 21	185 ± 29	120 ± 30	70 ± 30	30 ± 20
10	4	53 ± 30	133 ± 28	182 ± 33	157 ± 24	154 ± 34	110 ± 45
WEB 2170 (1 mg/kg)							
5 at 30 min	3	3 ± 5	-4 ± 6	5 ± 6	4 ± 3	5 ± 4	-3 ± 6

Results of calculated pulmonary arteriolar resistance show that the vehicle had no effect. PAF caused a dose-dependent rise in Ao and PA pressure and reduction in flow to LPA (up to 80%). Pretreated (WEB 2170) fetuses did not show hemodynamic responses to 5 ng/kg PAF. Unlike fetal lambs, PAF is a pulmonary vasoconstrictor in porcine fetuses regardless of dosage. Its effect is abolished by WEB 2170. Circulatory responses may be more representative of the human. PAF-inhibitors such as WEB 2170 may have a potential role in treating PFC.

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 68

Inhaled nitric oxide reduces elevated pulmonary vascular resistance in infants with congenital heart disease

Lundell B, Winberg P, Gustafsson L
Department of Pediatrics, St Görans Childrens Hospital and the Department of Physiology and Institute of Environmental Medicine, Karolinska Institute, Stockholm, Sweden

Nitric oxide (NO) is an endogenous local mediator of vascular tone and has been shown to reverse hypoxic pulmonary vascular constriction when inhaled. This study was performed to test if chronic non-hypoxic increased pulmonary vascular resistance (PVR) could also be reversed by inhalation of NO. Sixteen infants 3-10 (median 4) months old with CHD and pulmonary hypercirculation were studied during elective heart catheterization. Systemic and pulmonary hemodynamics were measured using oxygen consumption and the Fick principle before and during NO inhalation (40 ppm) for 10 min. In 6 infants with normal PVR (<4 $U \cdot m^2$), NO administration had no significant effect on the pulmonary hemodynamics. In 10 infants with elevated PVR, NO inhalation reduced pulmonary artery pressure from 47.2 to 35.7 mm Hg ($p<0.001$), and increased pulmonary blood flow from 6.1 to 8.6 $l \cdot min^{-1} \cdot m^2$ ($p=0.07$). PVR was reduced from 8.4 to 5.3 $U \cdot m^2$ ($p<0.001$). Systemic hemodynamics were unaltered. We conclude that inhaled NO can induce relaxation of the pulmonary arteries in infants with increased PVR secondary to pulmonary hyperperfusion. This observation may be useful in the treatment of pulmonary hypertensive crisis after corrective heart surgery.

P 69

Nitric oxide inhalation in children with congenital heart disease

Breuer J, Irtel von Brenndorff C, Sieverding L, Baden W, Gass M, Apitz J
Division of Pediatric Cardiology, University of Tübingen, Tübingen, Germany

The purpose of this study was to evaluate the therapeutic efficacy of NO inhalation in infants and children with secondary pulmonary hypertension (PH) or acute respiratory distress syndrome (ARDS) before or after surgery for congenital heart disease. All patients (n=12; age 1 day-6.5 years) were artificially ventilated and NO was introduced into the afferent limb of the ventilator circuit, while the inspired NO and O_2 concentrations were measured continuously. After registration of all hemodynamic parameters during inhalation of 5, 10, 20 and 40 ppm NO, inhalation was continued with 1-30 ppm NO as required to obtain a stable hemodynamic situation. As soon as possible, the applied NO concentration was reduced and then discontinued. The hemodynamic condition and/or arterial oxygen saturation (SaO_2) were significantly improved by NO in 11 patients (92%). SaO_2 increased from 80±3 to 91±2%, probably due to a decreased intrapulmonary right-to-left shunt (32 vs 22%). Mean pulmonary arterial pressure declined significantly from 37±8 to 25±6 mm Hg, whereas mean systemic arterial pressure remained constant (55±3 vs 58±4 mm Hg). This was related to a selective reduction in pulmonary vascular resistance by 43%. The significant, but minor increase in methemoglobin concentration (0.7±0.1 vs 1.4±0.2%) did not affect oxygen transport. In conclusion, NO inhalation in a low dose range selectively reduces pulmonary artery pressure and improves SaO_2 in children with congenital heart disease and ARDS or PH during perioperative care. The hemodynamic improvement seems to be related to an optimized right ventricular performance, since right ventricular afterload is reduced without changes in coronary perfusion pressure as often observed with other vasodilators.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 70

Italian multicenter study on congenital heart disease

Bosi G, DiCamillo A, Scorrano M, Tosato G, Forini E, The IMS-CHD Working Party
Institute of Pediatrics, University of Ferrara, Ferrara, Italy

The general aim of the Italian multicenter study on congenital heart disease (IMS-CHD) is to measure the prevalence rate and the distribution of CHD in Italy and to assess the survival and outcome of affected babies. During 1992-93, the IMS-CHD participating Centers (Milano, Padova, Bolzano, Bologna, Parma, Ferrara, Ancona, Roma, Napoli, Bari, Reggio Calabria, Messina, Palermo, Cagliari) recorded 1,380 cases of CHD among 334,412 livebirths. The average incidence of definite CHD with detailed diagnosis was 3.91 per thousand births; 43.9% of CHD were isolated ventricular septal defects, 7.9% pulmonary artery stenosis, 7.5% atrial septal defect, 3.5% tetralogy of Fallot, and 5.2% endocardial-cushion defects. CHD was first suspected during the first week of life in 70.8% of cases and up to 83% of children were submitted for cardiological examination during the first month of life. Extracardiac anomalies were present among 11.9% of the cases, with chromosomal aberration being the most important subgroup by size (48.2%), followed by gastrointestinal (13.2%), and urinary anomalies (4.02%). Surgery was required in 10.8% of CHD cases in the neonatal period (palliative in 43.6%, corrective in 51.4%). Eighty-two children (6.2%) died, and 87 (8%) of those deaths were CHD-related.

P 71

Congenital heart defects in first degree relatives of 94 infants with hypoplastic left heart syndrome

Sauer U, Burkhart J, Berger G, Bühlmeyer K, Bouvagnet P
Deutsches Herzzentrum München, München, Germany and CRBM, CNRS, Montpellier, France

The incidence and pathogenetic type of congenital heart defects (CHD) in 10 relatives of 94 hypoplastic left heart syndrome patients (HLHS-pts) classified according to Sinha (1968) were determined by questionnaire (1990 & 1993). Subclinical CHD was not excluded. HLHS-probands (63 males, 31 females) presented between 1975 and Dec 1990; 41.5% (39/94) HLHS-pts were first live-born children. CHD occurred in 3.4% (14/415) of 10 relatives: 2.1% (4/188) parents and 4.5% (4/89) siblings born before HLHS-pt were affected, constituting a 2.9% (8/277) precurrence risk vs 4.3% (6/138) recurrence risk in afterborn siblings. The sibling recurrence risk of HLHS plus extracardiac anomalies (12.1%, 4/33) exceeded that of isolated HLHS (3.1%, 6/194). The abortion rate (11.3%, 41/363 pregnancies) was increased. By developmental/mechanistic classification, recurrent CHD were concordant blood-flow defects (Group II: left–10, right–2) in all but 2 families, however, inconsistent by severity, ranging from bicuspid aortic valve to HLHS or ASD II in father and HLHS in first child and tricuspid atresia in 2nd child. Coexistence of pathogenetically discordant CHDs in same sibship—PA+VSD (conotruncal-Group I) and HLHS (Group II) or HLHS (Group II) and DORV+mitral atresia+CoA (conotruncal Group I & Group II-defect) and CoA+membranous VSD (Group II & branchial arch Group I-defect)—suggests a common causative genetic factor. Thus, the incidence of familial CHD in HLHS is higher than predicted by multifactorial mode of inheritance. Counseling on risk has to take into account the possible recurrence of pathogenetically related defects.

P 72

Phenotypic evaluation and genetic molecular study in patients with tetralogy of Fallot

Digilio MC, Marino B, Amati F, Mari A, Mingarelli R, Novelli G, Giannotti A, Dallapiccola B
Pediatric Cardiology and Genetics, Bambino Gesù Hospital, Rome and the Department of Cell Biology, Tor Vergata University, Rome, Italy

Tetralogy of Fallot (TF) is a congenital conotruncal heart defect occurring in DiGeorge (DG) and velocardio-facial (VCF) syndromes. The microdeletion of chromosome 22q11 (del22) is a well established cause of DG and VCF, and it has been demonstrated also in apparently isolated TF. In order to define the proportion of syndromic cases, and to investigate the prevalence of del22 in an unselected population of TF, we studied 157 consecutive children, 88 males and 69 females. The cardiac diagnosis was obtained by echocardiography and cardiac catheterization in all cases, and confirmed at surgery in 129 cases. All patients underwent phenotypical and clinical evaluation. DNA analysis was performed using the HD7k probe. The phenotypical study revealed that 113 patients had isolated TF. A diagnosis of syndromic TF was made in 44/157 (28%) patients: Down (20 cases), VCF (15 cases), DG (3 cases), Goldenhar (3 cases), Noonan (2 cases), Williams (1 case). Del22 was detected in 12/157 (7.6%) patients. All del22 cases were syndromic patients: 3/3 (100%) with DG, 8/15 (53.3%) with VCF, and 1/20 (5%) with Down syndrome. None of the 113 patients with isolated TF had del22. Our results show that del22, occurring in a high proportion of syndromic patients (27%), is always associated with extracardiac anomalies, and it is absent in isolated TF.

P 73

Morphological and genetic differences of atrioventricular canal defect in patients with and without Down syndrome

Marino B, Digilio MC, Santoro G, Formigari R, de Zorzi A, Giannotti A, Mingarelli R, Dallapiccola B
Pediatric Cardiology and Genetics, Bambino Gesù Hospital, Rome, and Department of Cell Biology, Tor Vergata University, Rome, Italy

Atrioventricular canal (AVC) defects are found in children with and without Down syndrome. In order to identify similarities and differences between the two groups in regard to cardiac morphology and genetic causative factors, we reviewed the clinical records of 238 consecutive patients with AVC, 126 with trisomy 21 and 112 with normal chromosomes. We performed molecular studies on two large families with recurrence of AVC without trisomy 21. Cardiac morphology was evaluated by echocardiography, angiocardiography and surgery. Patients with heterotaxia were excluded. Molecular studies included linkage analysis between AVC and markers on chromosome 21. Morphological studies demonstrated that children with AVC and trisomy 21 showed a prevalence of the complete form (80/126, 63.5% $p<0.01$) and of association with tetralogy of Fallot (11/126, 8.7% $p<0.05$). In contrast, patients with AVC without trisomy 21 showed a prevalence of the partial form (86/112, 76.8% $p<0.01$) and of association with left-side obstructions (19/112, 16.9% $p<0.01$). Molecular analysis failed to demonstrate a linkage between chromosome 21 and non-Down AVC. Genes located on other chromosomes may be involved in the pathogenesis of non-Down AVC, determining its peculiar cardiac morphology and associated malformations.

P 74

Myocardial underexpression of copper-zinc superoxide dismutase gene in Down patients with complete atrioventricular canal

Santoro G, Ambrosio G, Formigari R, Guccione P, Chiariello M, Marino B

Department of Pediatric Cardiology, Bambino Gesù Hospital, Rome and the Department of Cardiology, University "Federico II," Naples, Italy

Copper-zinc superoxide dismutase (Cu-Zn SOD) is a major scavenger against oxygen free radicals (OFRs) physiologically generated during cell metabolism. Balance between OFRs generation and scavenging influences endocardial cushion morphogenesis in experimental models. Since Cu-Zn SOD gene is located on the chromosome 21, its tissue expression should be higher in pts with trisomy 21 (Down syndrome). In order to test this hypothesis, we measured Cu-Zn SOD levels in red blood cells and myocardial tissue of Down pts with atrioventricular canal (AVC) (n=11) and compared with homologous tissues of age matched non-syndromic pts without AVC (n=9). As expected, Cu-Zn SOD levels of red blood cells were higher in Down than in normal pts (30.6±1.8 vs 23.8±1.1 IU/g prot, $p<0.01$). Conversely, there was a trend for myocardial Cu-Zn SOD levels of Down pts to be lower than that of normal pts (30.7±8.2 vs 272.7±195.6 IU/mg prot, p=NS). In conclusion, we hypothesize that this selective underexpression of Cu-Zn SOD gene at myocardial level might influence endocardial cushion morphogenesis causing AVC that is a particularly frequent cardiac malformation in Down pts.

P 75

Dual sensor pacemakers in children—what is the choice of sensor blending?

Çeliker A, Tokel K, Lenk MK, Özme S

Department of Pediatric Cardiology, Hacettepe University Medical School, Ankara, Turkey

Dual sensor pacemakers were developed to obtain more appropriate response to activity. We evaluated ten children with dual sensor pacemaker in different sensor blending circumstance by exercise testing to assess which ratio was optimal. Ten patients with several brady-dysrhythmias, ages ranged from 6 to 16 years mean 10.1 years, were included in the study. Eight patients had VVIR (TOPAZ®), and two patients had VDD (SAPHIRE®) pacemakers applied by transvenous route. All patients were in pacemaker rhythm (98.5% pacing). Accurate T wave sensing ranged 81-100%, mean 92%, median 95%. Voluntary exercise testing with CAEP protocol using treadmill in VVIR pacemaker mode, medium activity threshold and three sensor blending ratios (QT=ACT, QT>ACT, QT<ACT) was done in all patients. The mean duration of exercise was not statistically different in three sensors blending ratios. After 90 seconds of exercise, mean pacing rate had increased by 12%, 3%, 5% respectively in three groups. At maximal exercise the increases were 45%, 42%, 54%. Mean heart rates during exercise stage in three ratios were not statistically significant, although we showed statistically significant increase in heart rate during first two stages according to rest period in QT=ACT and QT<ACT sensor blending ratios. This difference was not observed after second stage. In conclusion, QT=ACT, QT<ACT sensor blending ratio seems to be more appropriate in children with dual sensor pacemakers.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press

P 76

Fetal cardiac masses—prevalence, follow-up and difficulties in counselling

Blackburn M, Wharton G, Parsons J, Dickinson D, Gibbs J
Killingbeck Hospital, Leeds, UK

Cardiac masses were seen in 44 of 1354 fetal scans performed in a pediatric cardiac unit over 4 years. In 39 fetuses they appeared as single (36) or multiple (3) discrete, highly echogenic masses 1-5 mm in diameter within the ventricular cavities and associated with the MV apparatus (36), a LV band (1), TV apparatus (1) or RV moderator band (1). Thirteen cases were followed into infancy. All the lesions became smaller, and although 10 were detectable at 4 weeks of age, none were seen after 4 months. Histology in 4 pregnancies terminated for other reasons showed small areas of fibrosis in 2. Four fetuses had larger masses of variable echogenicity, 2 involved the RV myocardium and extended into the cavity and 2 were contained within the ventricular septum. These lesions all had the appearance of rhabdomyomas, suggesting a high risk for tuberous sclerosis. The 2 pregnancies with septal masses were terminated . At autopsy, large fibrotic scars alone were found with dystrophic calcification, consistent with ischemic injury. In the other 2 cases, tuberous sclerosis was confirmed postnatally. A further fetus had a 3 cm mediastinal mass attached to the pericardium; teratoma was confirmed on autopsy following termination. In conclusion, benign masses in the fetal heart are common and recognizable by their echogenicity and their position in the ventricular cavities. Rhabdomyomata appear within the myocardium and are likely to be related to tuberous sclerosis, but masses of similar appearance may be of a more benign nature, possibly related to fetal ischemia. This finding has major importance when counselling parents on the risk of a fetus having tuberous sclerosis.

P 77

Prenatal findings in patients with long-QT syndrome

Hofbeck M, Ulmer H, Beinder E, Singer H
Departments of Pediatric Cardiology and Gynecology, University of Erlangen, Nürnberg and the Department of Pediatric Cardiology, Heidelberg, Germany

The purpose of this study was to describe the findings of fetal echocardiography (FE) in pts with long-QT syndrome (LQTS) who were referred to our institutions for prenatal evaluation. In our centers, 46 pts are followed with the diagnosis of LQTS according to the criteria defined by Schwartz et al (1993). Nine of these pts (20%) were referred to our centers for prenatal evaluation. Indications for FE were fetal bradycardia (5 pts), family history of LQTS (2 pts) or complex arrhythmias (2 pts). FE was performed from 24 to 40 wks of gestation (mean 32 wks). Five pts had persistent sinus bradycardia without further arrhythmias (HR 90-120 bpm). There were no signs of fetal hypoxia or placental dysfunction. One of these pts in addition showed a VSD and a moderate hypoplasia of the RV. In 1 pt we found sinus bradycardia and supraventricular extrasystoles and one pt had severe bradycardia (HR 70-100 bpm) due to intermittent II° AV block. Two pts presented with recurrent VT and intermittent II° AV block resulting in fetal hydrops in one of them. All 7 pts with sinus bradycardia or isolated II° AV-block were delivered spontaneously. Four pts were treated with propranolol postnatally and are asymptomatic. One pt who did not receive treatment died suddenly at the age of 3 wks. The second pt had syncope at the age of 6 wks and is now asymptomatic under treatment. The third pt has been asymptomatic without treatment. Both pts with prenatal VT and II° AV-block were delivered by cesarian section. They died on the first day of life despite aggressive treatment with propranolol and temporary transvenous pacing. In our experience, prenatal persistent sinus bradycardia is not uncommon in pts with LQTS and should alert the physician to perform ECG examinations of all family members and the neonate. Pts with prenatal VT and II° AV-block are a high risk population despite aggressive treatment in the neonatal period.

P 78

Radiofrequency current application at right atrial myocardium in young pigs—involvement of the right coronary artery

Paul T, Bökenkamp R, Zimmermann M, Jacobs M, Mahnert B, Trappe HJ, Maaß A, Sümpelmann R, Gräter T, Kallfelz HC
Pediatric Cardiology, Hannover Medical School, Hannover, Germany

Anatomical sequelae after creation of right atrial myocardial lesions by radiofrequency current (RFC) application have not been studied in detail in young subjects. During general anesthesia, a steerable 6 Fr electrode catheter, equipped with a thermistor at the 4 mm tip electrode, was positioned at the lateral atrial aspect of the tricuspid valve annulus in 10 piglets (german landrace, age 6 weeks, mean body weight 12.5 kg). RFC (500 kHz) was delivered temperature-guided (75 °C) over 30 sec. Right atrial RFC lesions were studied macroscopically and microscopically in 5 animals after 48 hours. The remaining 5 animals were examined after 6 months. After 48 hours, RFC lesions presented as transmural grey-white coagulation necrosis. Lesions were sharply demarcated by a lymphocytic wall. Lymphocytic infiltration around the lesions extended to the right coronary artery in 4/5 animals and to right ventricular myocardium in 3/5 piglets. After 6 months, lesions consisted of transmural white scar tissue extending to the right ventricular myocardium in 2/5 animals. Layers of the right coronary artery were affected in 4/5 pigs. In 2 animals, the lumen of the right coronary artery was obstructed due to intimal thickening by 25% and 40%, respectively. In conclusion, results should be taken into account when radiofrequency current application at the atrial aspect of the tricuspid valve annulus is considered in infants and small children.

P 79

Lesion size early and late after radiofrequency current application in young pigs

Paul T, Bökenkamp R, Zimmermann M, Jacobs M, Mahnert B, Trappe HJ, Maaß A, Sümpelmann R, Gräter T, Kallfelz HC
Pediatric Cardiology, Hannover Medical School, Hannover, Germany

Lesion formation after radiofrequency current (RFC) application at immature atrial and ventricular myocardium has not been investigated systematically yet. During general anesthesia RFC (500 kHz) was delivered temperature-guided (75 °C) over 30 sec in 10 piglets (german landrace, age 6 weeks, mean body weight 12.5) using a steerable 6 Fr electrode catheter equipped with a thermistor at the 4 mm tip electrode. Lesions were created at the right atrial aspect of the tricuspid valve anulus, at the left ventricular myocardium under the lateral mitral valve anulus and at the left ventricular apex, respectively. Lesion size was determined in 5 animals after 48 hours; the remaining 5 animals were examined after 6 months. Results are listed below in mm^3 by location.

Time	Tricuspid valve annulus	Mitral valve annulus	Left ventricular apex
48 hours	48.9 ± 13.5	150.2 ± 15 2	161.7 ± 14
6 months	27.8 ± 4.7	96 7 ± 18.6	164.2 ± 19.9
p	<0 01	<0 01	NS

After 48 hours, atrial and ventricular RFC lesions appeared as mainly transmural coagulation necrosis. After 6 months, lesions consisted as white scar tissue. Atrial and left ventricular apical lesions were still transmural. In conclusion, no increase in lesions size was observed with growth of the animals. Lesions were still mainly transmural late after radiofrequency current application.

P 80

Propafenone—site of action and efficacy in children with atrioventricular reentrant tachycardia

Müller G, Ulmer HE
Division of Pediatric Cardiology, Children's Hospital, University of Heidelberg, Heidelberg, Germany

The aim of this study was to prospectively assess efficacy and site of action of intravenous propafenone for termination of atrioventricular reentrant tachycardia in pediatric patients. We studied 14 patients (9 male, 5 female, age 1 week-20 years; mean age 7.4 years) with orthodromic reciprocating tachycardia (ORT, 9 pts), permanent form of junctional reciprocating tachycardia (PJRT, 1 pt) and AV-nodal reentrant tachycardia (AVNRT, 4 pts). Tachycardia was induced by transesophageal pacing during baseline (10 pts) or orciprenalin infusion (4 pts). Propafenone 0.5 mg/kg iv over 5 minutes was administered during tachycardia. This was repeated up to 4 times (maximum dose of 2 mg/kg) until tachycardia terminated. Propafenone terminated tachycardia in 11/14 pts (79%): ORT 7/9 pts, PJRT 1/1 pt, and AVNRT 3/4 pts. Tachycardia was not terminated in 3 pts. Required dosage for termination was 0.3-1.5 mg/kg iv (mean dosage 1.02 mg/kg). The site of propafenone action in the patients with ORT or PJRT was the accessory connection in 5 pts and the AV-node in 3 pts. In conclusion, the site of propafenone action in patients with ORT or PJRT was the accessory connection in the majority of cases. Overall, propafenone is effective in terminating atrioventricular reentrant tachycardia in a high percentage of pediatric patients.

Notes

https://doi.org/10.1017/S1047951100003127 Published online by Cambridge University Press