

Abstracts

Southeastern Pediatric Cardiology Society—25th Annual Meeting

Organized by Peter L. Ferrer

Miami, December 1992

AV valve competence after takedown to improve exposure during VSD repair—an echo-Doppler study

Pridjian AK, Culpepper WS, Williams LC, Ochsner JL, Pearce BF
Ochsner Clinic, New Orleans

Although the AV valve and its attachments can sometimes obscure the superior margin of a VSD, concern for valvar competence has made surgeons hesitant to take down the AV valve. From May 1982 to the present we began to take down the right AV valve to improve exposure for VSD repair in selected patients. Medical records of the 30 patients repaired in this manner were reviewed, and follow-up echocardiographic studies, available in 25 of the patients, were examined to determine the degree of tricuspid regurgitation and adequacy of VSD repair. Patients ranged in age from two months to 4.5 years (mean 18 months) having weights of 2.7 to 26 kg (median 5.5 kg). There were 25 patients with isolated perimembranous defects, three with subaortic defects and DORV, and two with perimembranous defects and DORV. Mean preoperative pulmonary to systemic blood flow ratio was 4.5:1, and pulmonary vascular resistance ranged from 0.5 to 6.6 Woods units (mean 2.9). Contiguous portions of the septal and anterior leaflets of the valve were taken down leaving a small cuff of leaflet tissue on the valve annulus. Valve leaflets were resuspended after VSD repair with running polypropylene. The degree of residual VSD and valvar regurgitation determined by echo were graded as none, trivial, moderate, or severe based on the area of the color-flow jet. Valvar regurgitation was graded as none in 16, trivial in eight, and mild in one. No patient had moderate or severe regurgitation. Eighteen patients had no residual VSD, and tiny residual VSD's were found in seven. There were no early or late deaths in any of the patients undergoing VSD repair by this approach. Takedown and resuspension of the AV valve is a safe and effective technique which improves exposure for VSD repair and does not adversely affect valve competence.

Echocardiographic evaluation of CPR in infants

Groud JM, Cavallaro DL, Saltiel A, Nichter MA, Quintessenza JA
All Children's Hospital, St. Petersburg

The mechanism of blood flow during cardiopulmonary resuscitation (CPR) remains controversial. External chest compression (ECC) may generate blood flow as the result of either direct compression of the heart between the sternum and spine (cardiac pump) or a generalized increase in intrathoracic pressure (thoracic pump). Several investigators have proposed that cardiac compressions occur in infants due to their compliant chest walls. In a retrospective study in infants, we used clinically indicated two-dimensional echocardiography (TDE) to determine whether ECC causes changes in left ventricular (LV) geometry. TDE was being performed on four postoperative cardiac patients when they suffered cardiac arrest. Chest compressions were then carried out simultaneously with TDE and recorded on videotape. Percent change in anterior-posterior dimension of the LV (LVD) and the distance from the apex of the image to the LV posterior wall (PW) were measured during ECC. In conclusion, this study suggests that in infants with a closed sternum, chest compression does not cause cardiac compression. This study supports the thoracic pump theory in this group of patients. Maneuvers that increase intrathoracic pressure may be of benefit by increasing blood flow during CPR in infants.

Detection of cardiac allograft rejection in children using echocardiographic indices of left ventricular size and function

Tantengco MVT, Dodd D, Garrison D, Davis A, Boucek RJ Jr.
Vanderbilt University, Nashville

Cardiac transplantation has now become an accepted management option for children with end-stage heart disease. Detection and early treatment of graft rejection remain the key factors determining short- and long-term survival. Adequate noninvasive echocardiographic (ECHO) monitoring for rejection in infants has been described using a multiparametric computer program. We hypothesize that abnormalities in left ventricular (LV) mass and function are associated with acute allograft rejection and that ECHO analysis using a multiparametric scoring system could facilitate monitoring rejection in an older pediatric population. A scoring system incorporating multiple ECHO parameters (derived retrospectively) was applied in a prospective fashion to 12 pediatric patients (age range at study: 1.03 to 13.1 yrs). A total of 21 ECHO analyses were performed within 24 hours of obtaining RV endomyocardial biopsies, 0.33-64.4 months after transplantation. 5/12 patients developed moderate to severe rejection (biopsy > Gr. II), 1.3 to 37.7 months after transplantation, and were treated. 4/5 of these patients had signs and/or symptoms suggestive of rejection at the time of their ECHO and biopsy. Negative biopsy findings (normal to mild rejection) were found in 10 patients. No single ECHO parameter has proven to be sufficiently sensitive to detect moderate to severe graft rejection. A multiparametric ECHO scoring system was applied prospectively. The mean±SEM scores were 2.2±0.4 vs. 5.8±0.9 for the absent/mild vs. moderate/severe rejection groups, respectively (p<0.01). Rejection was defined as an ECHO score >4 and/or a subnormal shortening fraction. This multiparametric ECHO scoring system was 100% sensitive and 81% specific for the diagnosis of moderate/severe rejection. We conclude that abnormalities in LV mass and function are found with rejection, and a multiparametric ECHO scoring system correlates with the histopathologic diagnosis of moderate to severe graft rejection in this pediatric patient population.

Intraoperative biplane transesophageal echocardiographic evaluation of coarctation of the aorta

Fyfe DA, Lau YR, Sade RM
Medical University of South Carolina, Charleston

Biplane transesophageal echocardiography (TEE) was utilized for preoperative and immediate post-repair intraoperative evaluation in four patients undergoing surgical correction of congenital coarctation of the aorta. Patients were from three to 16 (median 12) years weighing 14 to 81 (median 20) kilograms. TEE probes were seven and 13 mm diameter and 5 MHz frequency. Preoperative imaging defined the location and morphology of the coarctation site and the diameter of the proximal and distal aorta. Color flow and range gated pulsed Doppler detected and localized abnormal blood flow patterns through the coarcted segment in each case. Collateral vessels to the descending aorta were defined in the two cases in which they were present. Postoperatively, two patients had widely patent anastomoses seen with normal pulsatile flow in the descending aorta. Collateral blood flow in the aorta was eliminated. Two patients were noted to have residual stenosis with aliasing blood flow patterns beginning at the site of end-to-end anastomoses. Arm-leg blood pressure measurements confirmed the presence of residual coarctation as did surface echocardiography. In conclusion, biplane TEE provides excellent intraoperative evaluation of coarctation of the aorta and predicts postoperative results.

Congenital heart disease and associated non-cardiac abnormalities

Perry LW, Ferencz C, Neill CA, Brenner JI, Martin GR, Downing JW, Mardini MK

The Baltimore-Washington Infant Study Group (BWIS), Georgetown University, Washington, D.C. and the University of Maryland, Baltimore Between 1981 and 1989, 4390 cases of congenital heart disease (CHD) and 3572 controls without CHD were registered by the BWIS, a case-control study of environmental risk factors in CHD. Of the cases, 75% had isolated CHD and 25% had associated noncardiac abnormalities (NCA). Of the NCA, 12% were chromosomal abnormalities, 8% were syndromes and 5% were other organ abnormalities. Only 2% of controls had NCA. Less than 10% of cases with complete transposition of great arteries (TGA), tricuspid atresia, pulmonary atresia and pulmonary stenosis had NCA. Eleven to 20% of cases with hypoplastic left heart (HLH), aortic stenosis, ventricular septal defect (VSD), coronary artery/mitral valve abnormalities had NCA. Twenty-one to 30% of cases of corrected transposition of great arteries (CTGA), total anomalous pulmonary venous return (TAPVR), Ebstein's malformation, coarctation of aorta (COAO), bicuspid aortic valve, branch stenosis of pulmonary arteries (BSPA), atrial septal defect (ASD) and patent ductus arteriosus (PDA) had NCA. Thirty-one to 40% of cases with truncus arteriosus, double outlet right ventricle (DORV), tetralogy of Fallot (TOF), partial anomalous pulmonary venous return (PAPVR) and cardiomyopathy had NCA. Forty-eight percent of cases with interrupted aortic arch (IAA) had NCA. Seventy-eight percent of cases with heterotaxia and/or atrioventricular septal defects had NCA. Major organ defects—kidney, brain, gut—occurred in 22% of cases with PAPVR, 15% of cases with HLH, 13% of cases with TOF, 10% of cases with heterotaxia, and 10% of cases with TAPVR. Only double inlet left ventricle and cor triatriatum had no associated major organ defects. In the remainder of the cases less than 10% had associated major organ defects. Chromosomal abnormalities were encountered in 71% of AV septal defects, in 14% of ASD, in 14% of PDA, in 12% of COAO and DORV and in 11% of TOF. Double inlet left ventricle, cor triatriatum, PAPVR, tricuspid atresia and CTGA had no associated chromosomal abnormalities. Chromosomal abnormalities were noted in only 0.5% of cases with TGA and in 1.2% of cases with cardiomyopathy. Of the remaining types of CHD, 3-8% of cases had associated chromosomal abnormalities. Syndromes were encountered in 65% of cases with heterotaxia, 42% with IAA, 32% with cardiomyopathy, 26% with truncus arteriosus, 23% with CTGA, 20% with BSPA, 16% with DORV, 12% with Ebstein's malformation, 11% with PAPVR and 10% with TAPVR. Double inlet left ventricle, cor triatriatum and coronary/mitral abnormalities were not associated with syndromes. In the remainder of the cases, less than 10% were associated with syndromes. Compared with controls, cases with CHD have a highly significant risk of having NCA. The BWIS has outlined the population-based prevalence of chromosomal abnormalities, major organ defects and syndromes with specific types of CHD. Noncardiac abnormalities should be considered in the evaluation of patients with CHD.

Chromosomal abnormalities and congenital heart disease—1981-1989

Perry LW, Ferencz C, Neill CA, Brenner JI, Martin GR, Downing JW, Mardini MK

The Baltimore-Washington Infant Study Group (BWIS), Georgetown University, Washington, D.C. and the University of Maryland, Baltimore Between 1981-1989, 4390 cases of congenital heart disease (CHD) were registered by the BWIS, a case-control study of environmental risk factors in CHD. Of the cases of CHD, 522 or 12% had chromosomal abnormalities. Cases of CHD were divided into developmental mechanism subgroups as follows: (1) situs and looping abnormalities—transposition and heterotaxia; (2) cell migration abnormalities—truncus arteriosus, double outlet right ventricle (DORV), and tetralogy of Fallot (TOF); (3) complete transposition of great arteries (TGA); (4) flow lesions right-sided—pulmonary atresia (PA), pulmonary stenosis (PS), branch stenosis of pulmonary arteries (BSPA), left-sided—hypoplastic left heart (HLH), aortic stenosis (AS), coarctation of aorta (COAO), bicuspid aortic valve (BAV), and septal defects/ductus—membranous ventricular septal defect (VSD), atrial septal defect (ASD), patent ductus arteriosus (PDA); (5) cell death abnormalities—Ebstein's and muscular VSD; (6) extracellular matrix abnormalities—atrioventricular (AV) septal defects; (7) targeted growth abnormalities—total anomalous pulmonary venous return (TAPVR); (8) cardiomyopathy (CM); (9) other rare lesions. Of 385 cases of Down's syndrome, 56% had AV septal defects and 31% had ASD, membranous VSD, or PDA. Six percent had cell migration abnormalities—20 with TOF and 2 with DORV. Left-sided lesions occurred in 2.3%—1 HLH, 1 AS, 1 COAO, 3 BAV. Cell death abnormalities occurred in 2.1%—8 muscular VSD. Right-sided lesions occurred in 1.6%—3 with PS and 3 with BSPA. No situs or looping abnormalities were noted. Of 44 cases with trisomy 18, 12 or 27% had membranous VSD, 9 or 21% had cell migration defects—5 DORV, 4 TOF. Seven or 16% had left-sided lesions—2 HLH, 2 AS, 3 COAO. Right-sided and cell death abnormalities occurred in 2 and 5%, respectively. No situs or looping abnormalities were encountered. Of 25 cases of trisomy 13, 10 or 25% had cell migration abnormalities—2 truncus, 1 DORV, 7 TOF; 6 or 24% had septal defects or ductus—3 membranous VSD, 2 ASD, 1 PDA, and 5 or 20% had left-sided lesions—2 HLH, 2 COAO, 1 BAV. A single case of heterotaxia was reported. There were no cases of TGA, muscular VSD or AV septal defect. Of the 18 cases with Turner's syndrome, all had left-sided lesions—3 HLH, 4 AS, 9 COAO, 2 BAV. Of the 50 cases with other chromosomal abnormalities, septal defects, AV septal defects, left-sided lesions and right-sided lesions were encountered most frequently. Situs and looping abnormalities, TGA, TAPVR and cardiomyopathy rarely were associated with chromosomal abnormalities. Cell migration abnormalities and AV septal defects occurred more frequently in association with chromosomal defects than in our total experience with CHD. Right-sided lesions, cell death abnormalities, and situs/looping abnormalities occurred less frequently in association with chromosomal defects than in our total experience with CHD.

Magnetic resonance imaging of children with dilated aortic roots

Shuler CO, Wiles HB, Gillette PC

Medical University of South Carolina, Charleston

The purpose of this study was to compare the findings of magnetic resonance imaging (MRI) of aortic roots of children with Marfan's syndrome and those with other causes of aortic root dilation. The cardiac MRI scans of 11 children were retrospectively reviewed. The patient population included children with Marfan's syndrome (6), bicuspid aortic valve (BAV) with coarctation of the aorta (2), coarctation alone (1), Takayasu's arteritis (1), and aortic valve stenosis and insufficiency (1). Measurements were made at the levels of the aortic valve hinge point, sinuses of Valsalva, ascending aorta and the descending aorta. In all the children, the measurement at the level of sinuses of Valsalva were greater than the 95th percentile for published 2D echocardiographic normals. The patients with Marfan's syndrome had the largest measurements at the level at the sinuses of Valsalva in contrast to all other patients who had the largest measurement in the ascending aorta. In conclusion, MRI is a useful tool in assessing aortic root dilation in children and shows different features in those patients with Marfan's syndrome as compared to other causes of aortic root dilation.

Radiofrequency catheter ablation of incessant, medically resistant supraventricular tachycardia in infants and small children

Case CL, Gillette PL, Knuck BJ, Oslizlok PC, Blair HL

Medical University of South Carolina, Charleston

This study evaluates the efficacy and safety of radiofrequency catheter ablation (RF) of incessant supraventricular tachycardia (SVT) in a group of 6 infants (<1 yr) and two small children (<14 kgs). In this group of eight patients, the average age was nine months (range 1-27 mos) and the average weight was 7 kgs (range 4-13.5 kgs). All patients had failed amiodarone or IC agents alone or in combination prior to ablation. Diagnoses included WPW and SVT (4), SVT via a unidirectional retrograde accessory connection (AC) (3), and atrial ectopic tachycardia (AET) (1). In the seven patients with AC, four had single left-sided AC, one patient had two left-sided AC, and two patients had right and left AC; the AET was right sided. The 7 patients had a total of 10 RF procedures. RF alone was successful in tachycardia ablation in six patients, including four patients who were <3 mos. RF in combination with surgery was successful in one patient with dual AC; in the remaining patient, RF failed, yet surgery eliminated the SVT. There were no complications, and all patients are tachycardia free off medications. We conclude that RF of medically resistant SVT in selected infants and small children is a feasible nonpharmacologic alternative to surgical ablation.

The diagnosis and management of vascular rings

Redmond CR, Culpepper WR III, Pridjian AK, Ochsner JL
The Ochsner Clinic, New Orleans

Understanding the embryology and anatomy of tracheoesophageal compression syndromes is key to their management. Barium swallow, bronchoscopy, bronchography, esophagoscopy, esophagography, two-dimensional echocardiography, and angiography have all been used for preoperative diagnosis. This study was performed to determine which diagnostic modality best predicts the surgical findings. Twenty-four patients with symptomatic tracheoesophageal compression syndromes were treated surgically between January 1962 and November 1992 at the Ochsner Clinic. The mean age at the time of surgery was 5.0 years (range four days to 43 years). Fourteen patients were female and 10 were male. All patients were symptomatic prior to surgery. Using the Mayo Clinic classification of vascular rings there were ten patients with double aortic arch (Type IA); one with a left aortic arch and a ductus from the right pulmonary artery to the descending aorta (Type IIA); four with left aortic arch and a retroesophageal left subclavian artery with a ductus to the right pulmonary artery (Type IIB); six with right aortic arch, mirror image branching, and a left ductus (Type IIIA); and three with a right aortic arch, mirror image branching, and a retroesophageal left subclavian artery with a left ductus (Type IIIB). Barium swallow was performed in 18 patients and in 17 the diagnosis of vascular ring was made. However, the exact anatomy was correct in only 6 (33%). There was one false negative exam. Angiography correctly defined the anatomy in 19 of 20 patients (95%). In one patient (5%) the precise anatomy of the vascular ring was misinterpreted. Angiography provided additional useful information in 6 (25%) patients with associated congenital heart defects and was helpful in planning the operative approach in three patients. All but three patients were operated upon through the left chest. Two of the patients were operated on through a right thoracotomy, and in one patient the vascular ring was divided via a median sternotomy at the time of VSD closure. There were no operative deaths. 11 patients were symptom-free immediately after their operation. On long-term follow-up two patients have evidence of persistent tracheomalacia. There was one late death at four months secondary to complications with associated congenital heart disease. All vascular rings can be approached through the left chest but occasionally a right thoracotomy or a median sternotomy is more appropriate. Plain films, esophagoscopy, esophagography, bronchoscopy, bronchography, and two-dimensional echocardiography were not reliable diagnostic tests for vascular rings in our series. Our experience indicates angiography can precisely diagnose vascular rings and provide the surgeon with the correct information to optimally manage tracheoesophageal compression syndromes.

Significant reduction in the medical care costs associated with radiofrequency catheter ablation in children

Case CL, Knick BJ, Gillette PC, Crawford FA, Buckles DS
Medical University of South Carolina, Charleston

The total costs of definitive nonpharmacologic therapy in 10 children who underwent radiofrequency catheter ablation (RF) were compared to those of 10 children who had surgical ablation (SA) for medically refractory supraventricular tachycardia. The two groups did not differ according to mean age at procedures (6.2 yrs RF vs 6.8 yrs SA), weight (25 kgs RF vs 23 kgs SA), duration of symptoms (25 mo RF vs 23 mo SA), drugs used (2.1 RF vs 2 SA), or number of pathways (1.1 RF vs 1.1 SA). SA was performed in 1990 and 1991 and all RF procedures were performed in 1991. All pathways were ablated successfully with both procedures. Cost analysis revealed that the average total cost of therapy in the RF group was \$15,136±3,124 vs \$31,705±6,068 in the SA group ($p<0.005$). The mean total duration of hospitalization was 3.7±1 day in the RF group and 9.4±3 days in the SA group ($p<0.005$). We conclude that when compared to SA, RF therapy in a pediatric population results in a significant reduction in the cost of medical care as well as shortening the length of hospitalization. These reductions offer a major advantage of RF over SA therapy.

Early experience with total cavopulmonary connection and the Fontan fenestration

Adams CV, Pridjian AK, Williams LC, Pearce BF, Ochsner JL, Culpepper WS
The Ochsner Clinic, New Orleans

Total cavopulmonary connection and the adjustable Fontan fenestration are recent modifications of the Fontan procedure which have greatly reduced the morbidity and mortality for patients with functional single ventricle. Total cavopulmonary connection involves anastomosis of the superior vena cava to the undivided pulmonary artery with placement of an intraatrial baffle to channel inferior vena caval blood to the undersurface of the pulmonary artery. For Fontan fenestration, a five to 15 mm defect is created in the baffle which can be closed with a purse-string suture and snare. Between November 1991 and November 1992, eleven patients underwent the Fontan procedure. Patients were divided into two groups: those receiving total cavopulmonary connection alone (CPC) ($n=5$), and those receiving total cavopulmonary connection with fenestration (FF) ($n=6$). Since February 1992 all patients have received FF. Diagnoses were tricuspid atresia in five (one type I, three type II, and one type III), double inlet left ventricle in three, and complex univentricular heart in three. Mean age was 67 months in the CPC group and 80 months in the FF group. Four of five CPC patients and 4/6 FF patients had previously received Blalock-Taussig shunts. Intermediate bidirectional Glenn had been performed in seven patients (4/5 CPC and 3/6 FF). A pulmonary artery band had been placed in one patient. Mean preoperative pulmonary artery pressure was 16.4 mm Hg in CPC patients and 17.5 mm Hg in FF patients. Mean pulmonary vascular resistance was 2.0 ± 0.4 Wood units in the FF group. Additional procedures were performed in four patients, including DamusKaye-Stansel connection in one and pulmonary artery angioplasty in three patients. The fenestration was enlarged in one patient with elevated CVP and low blood pressure following cardiopulmonary bypass. Postoperative junctional tachycardia developed in two patients after CPC and in no patient following FF. Two patients had greater than 10 days of chylous drainage in the CPC group and three in the FF group. One patient in the FF group with persistent pleural effusion required pleurectomy and thoracic duct ligation. There were no hospital deaths. Snares have been closed in three of six FF patients. Two of the closures were performed at six and eight days following surgery in the cardiac catheterization laboratory. Oxygen saturations rose a mean of 4% and SvO₂ fell 8% upon closure. One snare was closed at the patient's original procedure and three have been left open because of suboptimal hemodynamics or prolonged chylous drainage. Mean arterial oxygen saturation on discharge in FF patients with an open fenestration was 85.3% compared to 97.3% in patients whose fenestrations were closed. We have been pleased with our experience with the CPC and FF procedures. Many patients with functional univentricular heart may receive FF without intermediate bidirectional Glenn. To ensure adequate cardiac output the FF is a safe and easily-performed adjunct to the CPC and is currently our procedure of choice in patients who are candidates for orthoterminal correction.

Radiofrequency catheter ablation via the atrial approach minimizes post-procedure valvular dysfunction in children

Lau YR, Case CL, Gillette PC, Shuler CO, Fyfe DA, Knick BJ, Buckles DS
Medical University of South Carolina, Charleston

A previous study in children has shown a 30% increase in aortic regurgitation and a 12% increase in mitral regurgitation (MR) after radiofrequency (RF) ablation via the retrograde ventricular approach. To examine if RF ablation via the atrial approach also causes valvular dysfunction, we performed 2-D and color Doppler echocardiography before and after each RF procedure on 65 consecutive patients (ages 1-324 months; median 158), for a total of 74 RF procedures. Diagnoses included accessory pathways (66 patients), ectopic atrial foci (four patients), and atrioventricular node reentrant tachycardia (four patients). The mean procedure time was 318 minutes (range 43-529) and the mean number of lesions was 21.1 (range 2-99). Our overall success rate was 84%. No patient developed aortic or tricuspid regurgitation. Only one patient (1.3%) developed MR. This patient had two left-sided pathways and required 37 lesions to ablate both pathways. We conclude that, compared with the ventricular approach, RF ablation of SVT via the atrial approach is associated with less post-procedural valvular dysfunction.

Bidirectional cavopulmonary anastomosis in the surgical management of high risk single ventricle

Fontana GP, Greeley WJ, Kanter RJ, Kern FH, Schulman SR, Armstrong BE, Garson AC, Ungerleider RM

From the Departments of Surgery, Pediatrics and Anesthesiology, Duke University Medical Center, Durham

Since the introduction of the Fontan procedure in 1968 for the treatment of tricuspid atresia, its use has been extended to many complex forms of single ventricle. Unfortunately, these complex forms have been associated with increased mortality and morbidity rates. There appear to be a number of anatomic and physiologic risk factors predictive of poor outcome with the Fontan procedure. Anatomically, hypoplastic left heart syndrome, mitral atresia, double inlet ventricles, Epstein's anomaly with an inadequate right ventricle, anomalous pulmonary venous drainage, aortic outflow obstruction, pulmonary artery distortion, and heterotaxy syndrome carry an increased risk. Physiologically, elevated pulmonary artery resistance (>2 Wood units), atrioventricular valve regurgitation, and elevated ventricular end diastolic pressures (>12 mm Hg) are associated with elevated risk. In an attempt to reduce mortality and morbidity in the high risk single ventricle patient, staging towards a Fontan procedure was performed utilizing a bidirectional cavopulmonary anastomosis in 16 patients who met at least one of the above high risk criteria. Four patients have undergone bilateral bidirectional cavopulmonary shunts. All shunts were performed on cardiopulmonary bypass with a 0% mortality, and the surgical technique will be described. All patients had an uncomplicated postoperative course and were discharged, on average, six days after surgery. Subsequently, six patients have come to a Fontan procedure. Of the remaining 12 patients, four may never be candidates for Fontan completion due to persistence of high risk factors. All patients were symptomatically improved and arterial oxygen saturations exceeded 80%. There are several advantages to this approach including early reduction of ventricular volume loading, the option of performing concomitant pulmonary artery reconstruction (to enable enlargement of small pulmonary arteries), and providing for future access to pulmonary arteries for balloon dilatation, if necessary. The disadvantages are few and include potential development of pulmonary to systemic AV fistulae, development of systemic veno-venous collaterals (reduced with azygous vein ligation), and need for a future operation (if a candidate) to complete the Fontan. The Fontan procedure is now considered for many patients with high risk single ventricles. Improved survival may be allowed by a transition (or staging) to a Fontan-type physiology with procedures such as the bidirectional cavopulmonary anastomosis which allows stepwise adaptation.

Are fetal supraventricular extrasystoles always benign?

Ferrer PL, Gelband H

Department of Pediatrics, University of Miami/Jackson Memorial Medical Center, Miami

Supraventricular extrasystoles (SVE) is a frequent cardiac arrhythmia in newborns and infants. We have observed, as many other investigators, that SVE may precede supraventricular tachyarrhythmias (SVT) in the young. Until recently, fetal SVE have been considered by obstetricians a form of benign arrhythmia which resolves spontaneously. The aim of this investigation was to study the frequency of SVE in a population referred for prenatal cardiac evaluation by ultrasound, its form of presentation, its natural history, its relationship with SVT, and its outcome. We evaluated 144 patients (pts) with fetal cardiac arrhythmia by echocardiography; 89/144 pts (62%) had extrasystoles, the majority of which (84/89) were supraventricular (SVE). Its form of presentation was: in 59/84 pts (70%) isolated; in 25/84 pts (30%) they were frequent (bigeminy, trigeminy); in 18/84 pts (21%), some SVE were blocked, decreasing the ventricular rate. Spontaneous disappearance occurred in 80/84 pts (95%), mostly during pregnancy. Four/84 pts (5%) developed SVT (three supraventricular tachycardia, one atrial flutter), three prenatally (21-38 wks of gestation) and one perinatally. Conversion to sinus rhythm was achieved by pharmacologic intervention, prenatally in two pts and postnatally in two pts. Two pts had multiple rhabdomyomas of the heart diagnosed prenatally (each at 32 wks of gestation) and WPW syndrome diagnosed postnatally. All patients survived. In summary, SVE is the most frequent form of fetal cardiac arrhythmias (58%). Its natural history is benign; however, 5% of them are at risk of developing SVT, a potentially lethal cardiac arrhythmia that may produce hydrops fetalis. Thus, fetuses with SVE should have frequent monitoring of their heart rate.

Transaortic resection of subaortic membrane as treatment for subvalvar aortic stenosis

Jacobs JP, Palatianos GM, Welch EM, Chandar JS, Kaiser GA

University of Miami/Jackson Memorial Medical Center, Miami

We reviewed 19 consecutive patients operated on from 1973 to 1984 to evaluate the efficacy of transaortic resection of subaortic membrane as treatment for congenital subvalvar aortic stenosis. The patients' mean age at surgery was 8.8 years with a range of eight months to 18 years. Eight patients had symptoms of congestive heart failure; four patients had symptoms of angina pectoris; and the remainder were asymptomatic. All patients had systolic ejection murmurs, ten with palpable thrills. All patients underwent preoperative cardiac catheterization which revealed the presence of subaortic stenosis. Valvar aortic stenosis was also present in three patients and supravalvar stenosis in one. Systolic pressure gradient across the subaortic ring was 74.53 ± 31.49 mm Hg. Eight patients also demonstrated a wide range of associated cardiac anomalies including three with ventricular septal defects, three with pulmonic stenosis, and two with a single main coronary artery. Transaortic resection of subaortic membrane was performed in all patients, aortic valvuloplasty performed in two patients, and additional operative procedures performed in four patients. Operative mortality was one patient, an eight-month-old with multiple cardiac anomalies including a ventricular septal defect, an atrial septal defect, a single main coronary artery, and cor triatriatum. Follow-up extended to 16 years with a mean of 6.9 years. No patients demonstrated post operative heart block or surgically induced mitral valve dysfunction. In five symptomatic patients, repeat cardiac catheterization revealed systolic pressure gradients across the subaortic membrane to be 33.0 ± 31.94 mm Hg 24.7 months (mean) after surgery. Two of these patients required reoperation for recurrent subaortic stenosis 23 and 34 months after their initial operations. One of these patients also had a tubular fibrous subvalvar tunnel as well as supravalvar and subvalvar mitral stenosis. This patient died 34 months after the second mitral valve replacement. Of the remaining thirteen asymptomatic patients, twelve underwent two dimensional and Doppler echocardiograms. Four of these patients were found to have recurrent subaortic stenosis with systolic pressure gradients between 55 and 85 mm Hg. Of these four patients, one underwent successful reoperation and the remainder are under observation. We conclude that transaortic resection of subaortic membrane is an acceptable treatment for subvalvar aortic stenosis in children but is associated with a high incidence of recurrence. Echocardiography appears to be an important method to serially follow patients after transaortic resection of subaortic membrane. Mortality from this problem is often related to associated anomalies.

Balloon aortic valvuloplasty in infants and children—results of early, intermediate and late follow-up

Fontenot EE, Gessner IH, Victorica BE

University of Florida, Gainesville

From May 1986, 37 pts have undergone balloon aortic valvuloplasty (BAV) for congenital aortic valve stenosis (AS). Mean age at BAV was seven yrs (range three days-23 yrs) with nine pts <2 mos old at the time of BAV. Mean systolic pressure difference (-P) across the aortic valve was 63 ± 27 prior to BAV and 29 ± 19 mm Hg immediately after the procedure. At early (<1 yr, n=25), intermediate (1-2 yrs, n=20), and late (>2 yrs, n=9) follow-up, DP was 51 ± 24 , 43 ± 17 , and 39 ± 19 mm Hg respectively. Two deaths occurred (mortality 5.7%) and both pts were <2 mos of age at BAV. Morbidity (27%) was largely related to femoral artery injury. Four of the nine pts <2 mos old required repeat intervention, either surgery (n=2) or repeat BAV (n=2). Of the 26 pts >2 mos of age, eight required either surgery (n=6) or repeat BAV (n=2). All but one of the 12 repeat therapies were for relief of residual DP >60 mm Hg. BAV is an effective alternative to surgery in pediatric pts with AS. BAV in infants <2 mos old is considered palliative; 22% mortality in our small series at this age should be reducible.

Effects of amrinone on the pulmonary and systemic vasculature in children with left-to-right shunts

Robinson BW, Mas MS, Gelband H
University of Miami, Department of Pediatrics, Miami

Treatment of heart failure in children with left-to-right shunts is controversial. Amrinone is a bipyridine derivative that possesses both potent inotropic and vasodilator effects. The drug has not been systematically evaluated in the patients with congenital heart defects with increased pulmonary blood flow secondary to left-to-right shunts. The present study was performed to determine if there is a beneficial effect from amrinone on the systemic and pulmonary vasculature in infants and children with cardiac left-to-right shunts. Nineteen patients (ages two months to 8.3 years) with cardiac left-to-right shunts were evaluated during cardiac catheterization with direct hemodynamic measurements made before and 10 minutes after a 3 mg/kg bolus of amrinone (peak effect). The Fick method was used to calculate pulmonary and systemic blood flow and resistances were then calculated. In patients with normal pulmonary artery pressure and resistance (n=5, Group A), amrinone significantly reduced mean pulmonary artery pressure by 19%, mean left atrial pressure by 39% and systemic vascular resistance by 17%. In patients with pulmonary artery hypertension (mean pulmonary artery pressure >20 mm Hg), and normal pulmonary vascular resistance (total pulmonary resistance <3 Woods units·m², n=7, Group B), amrinone significantly reduced the mean pulmonary artery pressure by 27%, mean aortic pressure by 12%, pulmonary arteriolar resistance by 36% and total pulmonary vascular resistance by 26%. In patients with pulmonary artery hypertension (mean pulmonary artery pressure >20 mm Hg) and elevated pulmonary vascular resistance (total pulmonary resistance >3 Woods units·m², n=7, Group C), amrinone significantly increased the left-to-right shunt by 94%, reduced the mean pulmonary artery pressure by 22%, pulmonary arteriolar resistance by 49% and total pulmonary resistance by 47%. In Group C, systemic vascular resistance did not change significantly. We conclude that in children with left-to-right shunts, amrinone: (1) has selective vasodilatory effects on the systemic and pulmonary vasculature depending on the degree of pulmonary artery pressure and resistance, (2) may have a beneficial hemodynamic effect in children with normal pulmonary artery pressure and resistance by lowering systemic vascular resistance and left atrial pressure and (3) may have an important therapeutic role in patients with pulmonary artery hypertension from elevated pulmonary vascular resistance without causing significant systemic hypotension.

Interventional catheterization palliation of "cor triatriatum" in a 1200 gram five-month-old sick infant

Raviele A, Sharma S, Vincent R
Emory University School of Medicine, Atlanta

A 1200gram five-month-old was transferred to our institute with the echocardiographic diagnosis of cor triatriatum. This diagnosis had initially been made when the infant's weight gain stalled at 1100 grams at four months of age. Her birth weight had been 450 grams. She had remained extubated until approximately one week before transfer to our institute, but was intubated because of possibly worsened congestive heart failure versus sepsis. At the family's insistence, the infant's diagnosis was discussed in our group and cardiac catheterization to substantiate the diagnosis and surgical repair at high risk were offered to the family. The possibility of palliation at catheterization was considered but doubted possible. The echo findings will be presented. Cath data and subsequent catheter intervention performed will be discussed. Follow-up clinical and echo data will also be presented. This case is being presented because of the patient's unusual form of "cor triatriatum" and the technical aspects of intervention in a 1200 gram sick infant.

Effect of medicines and substance abuse during pregnancy on heart disease in the offspring

Krovetz LJ
Hollywood

Mothers of patients seen from July 1, 1979 to October 31, 1992 were questioned regarding their use of medicines, alcohol or illegal drugs during pregnancy. If the mother was not available for questioning, the data were not counted. The only exception to this was if a positive drug screen was found or a history of drug abuse was obtained from relatives or the police. A total of 6,131 patients were seen and usable information was obtained in 4,150 instances. Memory of events during pregnancy was generally surprisingly good, with most mothers able to recall illness and medicines taken even years later. However, it was also obvious that most mothers did not consider over-the-counter medications in the same light as prescription drugs. Patients were divided into three categories—no heart disease (1835 pts), acquired heart disease (722) and cardiac anomalies (1593). The use of medicines and drugs was higher for mothers with offspring with cardiac anomalies (56%) compared to normals (49%) and acquired heart disease (47%) (p<0.05). Fourteen drugs were used by 20 or more mothers. Sex hormones (including birth control pills), antibiotics, antihistamines, apresoline, Bendectin, decongestants and diuretics were used by essentially the same proportion of each group. Maternal drug abuse was significantly greater in both cardiac disease groups. For acquired it was 10.4% and for the congenital 6.6%, compared to 3.5% in the normals. This was especially common for diseases associated with prematurity, e.g. PDA and RDS. Interestingly enough, marijuana usage was equal for all three groups. Insulin usage was more common in the acquired group (5.0%), compared to 1.4% of the other patients; these were mainly cardiomyopathies. There was a significant association of the use of Tylenol with certain cardiac anomalies. There was a higher use in ventricular septal defects (38%), tetralogy of Fallot (30%) and transposition of the great arteries (31%), compared to 24% for the normal and 18% for the acquired group. A total of over 100 medicines and drugs were used by the mothers in this study. At our present state of knowledge, it seems prudent to avoid any use of medicine, whether prescription drugs or over-the-counter, unless it is absolutely necessary for the health of the mother, during at least the first trimester of pregnancy.

High risk fetal cardiac arrhythmias

Ferrer PL, Gelband H
Department of Pediatrics, University of Miami/Jackson Memorial Medical Center, Miami

This investigation was aimed to evaluate the diagnosis, natural history and outcome of high risk or potentially lethal cardiac arrhythmias diagnosed prenatally by ultrasound. From an experience of 340 patients/551 fetal echocardiograms performed in our institution (1983-1992): 144 pts (42%) had cardiac arrhythmias of which 26 were considered high risk; four other pts from our perinatal experience were included. GI: 21 pts had supraventricular tachyarrhythmias (SVT), (11 supraventricular tachycardias, 10 atrial flutter). Atrial extrasystoles preceded SVT in 4/21 pts (19%); in 15/21 pts (71%) the SVT was sustained; 10/21 pts (48%) had associated congestive heart failure (CHF) (hydrops); 3/21 pts (14%) had associated structural heart disease (two rhabdomyomas, one oncocytic cardiomyopathy); 3/21 pts demonstrated WPW syndrome postnatally. Eleven pts with SVT had pharmacologic intervention in utero, eight of them (73%) with successful conversion to sinus rhythm. Others had obstetrical intervention and postnatal treatment. 18/21 pts (86%) survived. GII: Nine pts had complete heart block with eight requiring postnatal pacemakers; five had associated structural heart disease (four CHF in utero) and died, four without heart disease survived. In summary, high risk arrhythmias occurred in 18% of arrhythmias diagnosed prenatally; 8/30 pts (27%) had associated structural heart disease; 14/30 pts (47%) had CHF in utero. Aggressive therapy resulted in survival of 22/30 pts (73%).

Magnetic resonance imaging of complex congenital heart disease

Lau YR, Wiles HB, Denslow S, McKellar L, Gillette PC
Medical University of South Carolina, Charleston

In order to elucidate the strengths and weaknesses of magnetic resonance imaging (MRI) of congenital heart disease, we retrospectively examined eight patients with complex congenital heart malformations and compared MRI to echocardiography and angiography. These eight patients were chosen because they represented the most complex that we have imaged. Below is a table that lists the diagnoses of each patient.

Pt.	No.	Diagnoses	Summary of diagnoses
1	5		DTGA with aortic stenosis
2	6		Cor triatriatum with TAPVR to right atrium
3	12		Situs inversus totalis with ventricular inversion & VSD
4	8		AV discordance with VA concordance
5	8		VSD with double orifice mitral valve and aortic stenosis at two levels
6	5		Aortopulmonary window with interrupted aortic arch
7	6		LTGA with "criss-cross" heart and ventricular rotation and subpulmonic stenosis
8	8		Mesocardia with common AV valve and common atrium

The following table identifies the number of diagnoses imaged by each modality.

Pt.	MRI	Echo	Angio
1	4/5	3/5	2/3
2	5/6	5/6	6/6
3	12/12	4/12	10/10
4	5/8	7/8	5/8
5	6/9	7/9	
6	4/5	4/5	4/5
7	7/7	6/7	7/7
8	7/7	5/7	6/7
Total	50/59	41/59	30/36

We conclude that MRI is excellent at imaging complex congenital heart disease. Its advantages over echocardiography are (1) spatial relationship of cardiac structures as well as surrounding organs, (2) morphology of pulmonary arteries distally. The advantages over angiography are (1) no invasion of blood vessels, (2) no risk of angiographic dye, (3) spatial relationship of cardiac structures as well as surrounding organs. The limitations of MRI are its inability to estimate pressures and image valves, and the need for sedation.

Percutaneous transcatheter occlusion of coronary artery fistulas using detachable balloons

Skimming JW, Quisling RG, Fontenot EE, Victorica BE, Mickle JP, Gessner IH
University of Florida College of Medicine, Gainesville

Three pediatric patients, aged 13 months to 17 years, underwent successful transcatheter occlusion of their coronary artery fistulas using a detachable latex balloon system. This system offers several advantages over other occlusion techniques. First, the system permits controlled delivery and release of the balloon. Second, "test-occlusions" can be performed allowing for simultaneous balloon inflation, coronary cineangiography, and electrocardiographic monitoring. Third, since the balloons are flow directed, they are easily positioned in properly chosen locations. Finally, the balloons can be constructed to suit the size and shape of the fistula. In this study, two of the patients required only one balloon; in the other patient, two balloons were placed in the same fistula. All fistulas drained into the right atrium or ventricle and were successfully occluded. During a follow-up period of up to two years, no local or systemic reactions to the balloons were recognized. Fluid gradually leaks out of the balloons over a period of up to eight months. We conclude that, detachable balloon occlusion of coronary artery fistulas is a safe and effective alternative to surgical ligation in selected pediatric patients.

Syncopal children—the role of autonomic testing

Balaji S, Oslizlok PC, Allen MC, Case CL, Gillette PC
Medical University of South Carolina, Charleston

Autonomic tests were done in 180 children with syncope or presyncope between November 1986 and August 1992. All had a normal heart, no arrhythmias or long QT syndrome. The autonomic testing protocol consisted of tilt testing, carotid sinus massage, diving reflex, Valsalva maneuver, and response to graded intravenous boluses of isoproterenol and phenylephrine. Tilt test reproduced symptoms of syncope or presyncope in 120 (67%). Type of syncope was classified into vasodepressor (hypotension and bradycardia) in 41 (34.5%); cardioinhibitory (asystole > 5 sec) in 7 (5.5%), and mixed (hypotension and bradycardia) in 72 (60%). Carotid hypersensitivity was seen in four. Of the 120 tilt-positive patients, 116 were treated with fludrocortisone and salt with no further symptoms in 82 (72%), partial improvement in 15 (13.5%) and no change in 19 (16.5%) at follow-up of 1-65 months (mean 25). Of the 19 who were not improved on fludrocortisone and salt, 17 were treated with beta-blockers with symptom resolution in 14. Side effects (weight gain, headache) prompted stopping of fludrocortisone and salt in five (0.04%). Of the seven cardioinhibitory patients, four had pacemaker implantation, the other three opted for drug therapy. One patient had presyncope despite pacemaker and needed additional medical therapy. We conclude that tilt testing and carotid massage were the only useful maneuvers during autonomic testing. Fludrocortisone and salt therapy was effective in controlling syncope in most patients and beta-blockers helped the rest. Tilt testing does indeed help guide the management of children with syncope.

Extracorporeal membrane oxygenation (ECMO) for support of pediatric cardiac patients

Fasules JW, Frazier EA, Kiel E, Erickson CC, Moss MM, Norton JB,
Van Devanter S

The David M. Clark Cardiovascular Center, Arkansas Children's Hospital, Little Rock

Since July 1989, 41 infants and children underwent 49 ECMO supports at Arkansas Children's Hospital for circulatory failure secondary to myocardial dysfunction, dysrhythmias, or pulmonary hypertension postcardiac surgery. Six patients required seven ECMO supports prior to or unrelated to operative intervention. Thirty-five patients underwent 42 ECMO supports for cardiac dysfunction following cardiac surgery. Of these, ECMO was initiated in the operating room 25 times in 19 patients with eight long-term survivors (42%). ECMO was initiated in the ICU following surgery 17 times in 16 patients with 11 long term survivors (69%). ECMO was required twice in eight patients. All were weaned from the second support but only three survived (38%). Twenty of the 41 patients were long-term survivors. Causes of death included multi-organ failure with or without sepsis (11), failure of myocardial recovery (4), and renal failure (3). An analysis of all patients undergoing cardiac surgery with cardiopulmonary by-pass since the initiation of ECMO revealed a significant reduction in mortality since the application of ECMO. This reduction occurred predominantly in children with separate pulmonary and systemic circulations following surgery. We conclude: (1) ECMO offers improved survival to children having difficulty following complex cardiopulmonary bypass operations, (2) successful ECMO support can be initiated in the operating room, and (3) mortality may be increased in those patients requiring ECMO support prior to cardiac surgery.

Etiology and prognosis of heart disease diagnosed prenatally

Ferrer PL, Quetel TA, Salman F, Bezjian AA

From the Departments of Pediatrics and Obstetrics, University of Miami Jackson Medical Center, Miami

We studied the etiology and prognosis of heart disease (HD) diagnosed in utero. 551 fetal echocardiograms were performed in 340 patients (pts) (1983-1992); 75 pts with HD were selected for presentation. The most frequent cause of referral was an abnormal obstetrical scan (57%). Fetal/maternal factors were present in 32%; 25% of these had arrhythmias (A) some of them severe (AV block, supraventricular tachycardia); in 13% A was the cause of referral that permitted the diagnosis of HD. 24% had associated CHF (hydrops). Etiology included: (I) 12/75 pts (16%) had simple congenital heart disease (CHD): 1 ASD, 11 VSD (4 closed in utero); all survived. (II) 47/75 pts (63%) had complex CHD: 12 cardiopulmonary syndrome, seven atrioventricular defects, seven hypoplastic left heart, five single ventricle; three pulmonary atresia, three double outlet RV, three TV anomalies and seven pts with different CHD; only 13/47 pts (28%) survived. (III) 11/75 pts (15%) had myocardial disease: six cardiomyopathies, one myocarditis and four tumors; 7/11 pts (64%) survived. (IV) 5/75 pts (6%) had functional cardiac abnormalities: two twin-to-twin transfusion syndrome, three ductal constriction; 4/5 pts (80%) survived. Overall survival was 36/75 pts (48%); death occurred by obstetrical intervention (13%), fetal demise (23%), postnatal (64%). Prenatal evaluation of HD is a challenging diagnosis; its prognostic implications impose significant management and ethical dilemmas.

Pulse oximetry monitoring during cardiac magnetic resonance imaging in children with congenital heart disease

McKellar LF, Rexford TL, Wiles HB

Medical University of South Carolina, Charleston

Pediatric cardiac magnetic resonance imaging (MRI) requires careful safety monitoring since infants and young children require sedation and are often cyanotic. Use of pulse oximetry monitoring during MRI scans has been associated with burn injuries and interference with image quality. We safely monitored oxygen saturation in pediatric patients undergoing gated cardiac MRI without injury or sacrifice in image quality. Using a Nonin 8604D non-fiber optic pulse oximeter in a 0.5 Tesla Gyroscan we studied 193 patients with congenital or acquired heart disease. Patient age ranged from one day-31 yr.; weight ranged from 3-134 kg. The average scan time was 105 minutes. Of the 193 patients, 134 (69%) were sedated using a combination of chloral hydrate (50-100 mg/kg PO or PR), nembutal (2-5 mg/kg IM or IV) or Valium (1-10 mg PO). Moderate to severe cyanosis was present in 12 patients. A pediatric pulse sensor or an adult sensor clip was placed on the finger or toe. Extreme care was taken to ensure no loops or coils were in the oximeter cable. The oximeter unit always remained outside the 5 Gauss line marked on the magnet room floor. All 193 patients had excellent pulse correlations with saturations ranging from 62-100%. Direct observation of the sensor site prior to and immediately following the scan was performed on all 193 patients. A 24-hour follow-up was performed on 55 patients. There was no physical evidence of injury from the pulse oximeter. There was no effect on image quality or production of image artifact. Of the sedated patients, two scans were aborted due to mild apnea. One of these patients was receiving prostaglandin E₁. This study shows that the safety of pediatric cardiac MRI scans is increased by the effective use of pulse oximetry monitoring. Operator awareness and strict adherence to simple guidelines ensures that pulse oximetry monitoring causes no patient injury or loss of image quality. Cardiac MRI is safe for children, including cyanotic newborns.

Balloon dilatation of pulmonary artery valve in tetralogy of Fallot

Giroud JM, Martinez RM, Henry JG

All Children's Hospital and The University of South Florida, St. Petersburg and Tampa

Management of symptomatic infants with tetralogy of Fallot (TOF) still carries a significant morbidity and mortality even in the modern era. Recently there has been an interest in balloon dilatation (BD) of the pulmonary artery valve (PAV) as the first palliation in this condition. We wish to report our initial experience with this modality of treatment. Parents of infants with TOF and symptoms suggestive of hypercyanotic spells were presented with the option of balloon dilatation of the PAV as an alternative to surgical palliation. Patients were catheterized with the aid of an anesthesiologist. Careful measurement of the PAV annulus and diameters of the RPA, LPA and diaphragmatic aorta were carried out at the time of catheterization. If the calculated McGoon ratio number was ≥ 1.5 -1.8 with normal coronary architecture, BD was not carried out and the child considered for primary repair. Balloon dilatation was carried out with commercially available balloons using a diameter no greater than 100-140% of the measured annular diameter of the PAV. The initial patient (J.W.) was done as an emergency and the subsequent seven patients were offered BD of the PAV as their initial palliation. In conclusion, the preliminary data suggest that BD is an effective palliation in some infants with symptomatic TOF if the goal is a short term bridge to primary repair. A major advantage is that it can be performed using standard techniques and commercially available equipment, with acceptable morbidity and mortality.

Noninvasive assessment of total pulmonary vascular resistance in children with congenital heart disease

Ebeid MR, Ferrer PL, Gelband H

University of Miami School of Medicine, Department of Pediatrics, Miami

Noninvasive assessment of total and arteriolar pulmonary vascular resistance (TPR and APR) has not been well defined. In this study, 32 patients (pts) with congenital heart disease (age 10 days-28 yrs., mean 1.8 yr.) were studied. TPR and APR were measured in Wood units \cdot m² (W) in the cardiac catheterization laboratory using Fick's principle. Hemodynamic measurements were compared to Doppler echo parameters (DE) obtained in the main pulmonary artery and included pre-ejection period (PEP), ejection time (ET), its ratio PEP/ET and PEP divided by velocity time integral (PEP/VTI). The pts were further subdivided into four groups to determine the ability of PEP/VTI to select pts who have increased pulmonary artery pressure (PAP) on the basis of an increased TPR and/or increased pulmonary blood flow. Group (Gr) I (n=8) included pts with TPR < 4 W, normal PAP, Gr2 (n=13) pts with TPR < 4 W elevated PAP (systolic PAP > 30 mm Hg), Gr3 (n=5) pts with TPR between 4-10 W, Gr4 (n=6) pts with TPR > 10 W. PEP/VTI value of 0.4 sec/m or less recognized 21/21 of pts with TPR < 4 W, with and without elevated PAP, PEP/VTI of 0.4-0.6 sec/m recognized 4/5 of pts with TPR between 4-10 W, whereas > 0.6 sec/m recognized 5/6 of pts with TPR > 10 W. PEP/ET of 0.3 selected 15/21 pts of Gr 1 and 2, a value of 0.3-0.4 recognized 4/5 of Gr 3, and values > 0.4 recognized 4/6 of Gr 4. Overall accuracy of PEP/VTI was 94% and of PEP/ET was 73%. Furthermore, PEP/VTI was significantly different in the four groups. We conclude that PEP/VTI is an excellent parameter to assess the severity of pulmonary vascular resistance in the presence of pulmonary hypertension.

Efficacy of alpha-adrenergic agonist therapy for prevention of pediatric neurocardiogenic syncope

Strieper MJ, Campbell RM

From the Children's Heart Center, Egleston Children's Hospital at Emory University, Atlanta

Head-up tilt testing has recently proven effective for reproducing clinical symptoms of presyncope and syncope in pediatric patients suffering from neurocardiogenic syncope. Several studies have demonstrated that head-up tilt is relatively sensitive, specific and reproducible for this diagnosis. Recent reports suggest that withdrawal of alpha sympathetic stimulation is a possible mechanism responsible for neurocardiogenic syncope in adults. To test this hypothesis 16 pediatric patients (mean age 13.1 years, range 7 10/12 to 17 10/12 years) with a history of recurrent syncope were evaluated with head-up tilt (HUT). During baseline HUT, seven patients experienced vasodepressor syncope, seven patients had mixed vasodepressor-cardioinhibitory syncope and two had a pure cardioinhibitory response. All patients became symptomatic reproducing their clinical symptoms. Mean arterial pressure (MAP) (77 ± 14 vs 30 ± 14 mm Hg; $p < 0.0001$) and heart rate (HR) (105 ± 23 vs 87 ± 46 BPM; $p = \text{NS}$) decreased during baseline HUT. Phenylephrine infusion (average $1.69 \mu\text{g}/\text{kg}/\text{min}$; range $0.6\text{--}3.0 \mu\text{g}/\text{kg}/\text{min}$) was begun to achieve an increase in MAP of 10-15 mm Hg. Repeat 30 min HUT was performed. Fifteen patients remained asymptomatic without hemodynamic changes. One patient that had a pure cardioinhibitory response with baseline testing became presyncopal and had a blunted mixed response at 22 minutes on phenylephrine. During phenylephrine infusion the MAP (83 ± 19 vs 84 ± 18 mm Hg; $p = \text{NS}$) and HR (81 ± 17 vs 76 ± 16 BPM; $p = \text{NS}$) did not change. All patients received 60 mg oral pseudoephedrine twice daily as outpatients. Follow-up (mean 5.4 months, range 2 wks-14 months) revealed all patients to be subjectively improved without any reported alpha or beta side effects. In conclusion, (1) alpha-adrenergic withdrawal appears to play a significant role in neurocardiogenic syncope; (2) intravenous phenylephrine blocks the vasodepressor and cardioinhibitory response, even in the presence of reflex vagal bradycardia during phenylephrine infusion; and (3) oral pseudoephedrine alleviates symptoms in pediatric patients with neurocardiogenic syncope.

Effect of isoproterenol on the QT interval in children

Balaji S, Osluzlok PC, Gillette PC, Allen MC, Case CL

From the Medical University of South Carolina, Charleston

Catecholamines are known to increase the QT interval in patients with congenital long QT syndrome. They are also thought to reduce the QT interval in normals and patients with drug-induced long QT syndrome. The effect of a rapid injection of catecholamines in normal children has not been reported. We reviewed the effect of a rapid intravenous injection of isoproterenol (1-2 g) on the QT, RR, and QTc intervals of 19 children undergoing autonomic testing. Their ages ranged from four to 18 years (median 13). All presented with syncope but had a negative tilt test. Isoproterenol produced a reduction of QT in 14 patients, no change in two, and an increase in three. When analyzed by the paired t-test, the following results were obtained.

	QT ms	RR ms	QTc ms
Baseline	391 ± 57	891 ± 273	419 ± 24
Isoproterenol	341 ± 73	546 ± 190	459 ± 72
p value	< 0.025	< 0.00001	< 0.02

The QTc was increased spuriously due to a far greater effect of isoproterenol on heart rate than on QT interval. We conclude that a rapid injection of isoproterenol produces transient reduction of QT in most, but not all children. Use of the QTc can lead to misdiagnosis of long QT syndrome due to its overcorrecting effect. Isoproterenol given rapidly may not help in differentiating children with long QT syndrome from normals.