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Abstracts

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Optimal management of all patients with tetralogy of Fallot

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Controversy still exists as to whether patients with tetralogy of Fallot (TOF) should have initial primary repair or a systemic-to-pulmonary-artery shunt followed later by staged repair. From May 1, 1984 to May 1, 1991, we have utilized a selective policy in 42 consecutive patients with TOF, the management option being dictated by the patient's preoperative condition and the presence or absence of incremental risk factors for hospital death or poor result.

These included pulmonary atresia, small pulmonary valve ring and/or small main, right or left pulmonary arteries and arborization abnormalities. Age at repair ranged from 4.8 to 216.3 months (mean=34.9 months). There were three deaths (7%; 70% CL-3 to14%). For the group of 30 selected for primary repair there was one death. There were two deaths in 12 patients having staged repair. Postrepair right-to-left ventricular pressure ratios for each subgroup were 0.44 and 0.56, respectively. Of the three deaths, two were in the group of five with pulmonary atresia. Ten operations were done using the classical approach through the right ventricle. Twenty-eight were performed without a ventriculotomy (if a transannular patch was required [no.=27] it was kept small and usually less than 10 mm.). More recently,complete repair has been accomplished working only through the right atrium in four patients. The postrepair right-to-left ventricular pressure ratios were 0.47, 0.46 and 0.53, respectively. As our experience has increased we have tended to earlier repair but still follow a selective policy. We believe these data support continuation of this protocol.

Outcome after repair of tetralogy of Fallot correlated with intraoperative epicardial echo

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Fifty-one patients (pts) (27 pts <1 y/o) undergoing complete correction of tetralogy of Fallot (TOF) had pre- and post-CPB intraoperative echo with Doppler color flow imaging (IE-DCFI). Intraoperative data were compared to long-term outcome and to follow-up transthoracic echocardiograms (TTE). The series includes eight patients with TOF and pulmonary atresia. Follow-up continues except for two perioperative deaths and three patients who have died late following the hospitalization period (two SBE, one aspiration). IE-DCFI had an immediate impact on the intraoperative outcome for 13 patients (25%) by modifying the operative plan in six patients and encouraging revisions prior to leaving the OR in seven patients (five residual VSD, two RVOT reconstruction). Eight patients have had late reoperations (three VSD, three pulmonary artery plasty, one PVR, one conduit change). The reoperation in two patients with VSD and one with pulmonary artery plasty were predictable by IE-DCFI but occurred in patients who were operated on early in the series before we had adequate appreciation as to how to interpret IE-DCFI data. Therefore, IE-DCFI had potential to impact on 16 patients (31%). This technology is well known to be exquisitely sensitive, and 31 patients had small or moderate VSDs seen on immediate post-CPB IE-DCFI; however, 36% (11) were improved or undetectable on follow-up TTE,and 48% (15/31) remained unchanged. Residual RVOT turbulence usually remained stable or gradually increased during follow-up. IE-DCFI can be useful during repair of TOF, and follow-up studies reveal that many small residual defects in these patients may actually improve with time.

Combined transesophageal and epicardial imaging provide optimal intraoperative echocardiographic evaluation during ventricular septal defect surgery

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Intraoperative transesophageal echocardiography (TEE) was used in combination with epicardial imaging (EPI) following ventricular septal defect (VSD) repair in 18 children, ages seven to 137 (median 32) months and weights 6.3 to 49.1 (median 10.8) kgs. VSD types were seven malignant, six perimembranous, three supracristal, one mid-muscular, and one inlet. In two EPI studies and one TEE study, full VSD patch interrogation was not feasible. Seven residual VSDs were documented by color flow Doppler. TEE visualized six of the seven defects as did EPI. The missed defect was different with each technique. All residual defects were small in size by color flow jet analysis and by intraoperative oximetry (no Qp/Qs was greater than 1.50). All residual defects were located at the margins of the VSD patch; therefore, signal "masking" was not limiting. All residual defects were confirmed by postoperative surface echocardiography. In conclusion, combination intraoperative TEE and EPI was more effective in detecting residual VSDs than either technique alone.

Clinical spectrum of venous thrombi in the Fontan patient

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Venous thrombus formation is a known complication after Fontan-type procedures. We retrospectively reviewed the records of seven patients who developed thrombi following modifications of the Fontan procedure. Patients were aged six months to 18 years. Right-sided thrombi were detected from two days to four years after operation. Diagnostic modalities included angiography and surface

and transesophageal echocardiography. Three patients presented late (median five years), four patients presented early (median 16 days) following the Fontan. Additional "risk" factors for venous thrombosis were present in six of seven patients. These were central venous lines (four of seven), low cardiac output (three of seven), hypoteinemia (three of seven), intracardiac pacing wires (two of seven), atrial baffle obstruction (one of seven), and protein C deficiency (one of seven). Treatments included thrombectomy with revision or takedown of the Fontan (four of seven), coumadin (three of seven), streptokinase (three of seven), heparin (two of seven). "Late" presenters had resolution of their clots during coumadin therapy. "Early" presenters were critically ill at time of diagnosis and universally had poor outcome. In conclusion, 1) the clinical spectrum of thrombosis in Fontan patients is diverse, 2) risk factors for venous thrombi can be identified, and 3) "early" presentation is associated with high mortality.

TWO UNUSUAL CASES OF ANOMALOUS PULMONARY VENOUS RETURN TO THE RIGHT ATRIUM: CASE PRESENTATIONS AND INTRAOPERATIVE ECHOCARDIOGRAPHIC FINDINGS

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Two cases of anomalous pulmonary venous return (TAPVR) to the right atrium (RA) are presented. The first case is a five-month-old male infant who presented with cyanosis at birth and progressive congestive heart failure. The second case is a one-month-old male infant who presented with congestive heart failure. The first patient was initially thought to have a primum atrial septal defect and the other a cor triatriatum with polysplenia syndrome. The definitive diagnoses were only made at the time of surgery. The pulmonary veins appeared to be draining normally. In each case, a secundum ASD was present, but the septum primum was positioned in such a way to have three out of four pulmonary veins in case one, and all pulmonary veins in case two, drain to the RA. Surgical correction requires excision of part of the septum primum to enlarge the ASD followed by patch closure to direct the pulmonary veins to the left atrium. The prebypass echocardiogram in both cases revealed a markedly dilated right atrium and ventricle with a diminutive left ventricle. The atrial septum was abnormally positioned in both long axis and short axis views. Postbypass echocardiograms revealed that the left ventricle became normal in size and exhibited excellent function, demonstrating that it could quickly adapt to an increase in volume load without the need for a residual ASD. The findings in these two cases also suggest that the embryological defect in this subset of cases of TAPVR is related to an abnormality of attachment of the septum primum rather than failure of development of the common pulmonary vein.

A NEW TECHNIQUE FOR REPAIR OF AORTICO-LEFT VENTRICULAR TUNNEL

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Aortico-left-ventricular tunnel (ALVT) is a rare form of congenital heart disease that usually occurs in association with the right coronary sinus. It occurs in less than 0.1% of congenital heart defects. We report a case associated with the left coronary sinus in a three-year-old male patient. After repair, a disturbing number of patients develop severe aortic valvular insufficiency in the late follow-up period requiring valve replacement. This is believed to be secondary to the inherent lack of substantive tissue between the aortic wall and the defect. We describe the cardiac catheterization technique for determining the diameter of the ALVT and simultaneously assessing the competency of the aortic valve. Of the surgical techniques proposed, the main consideration has always been obliteration of the aortic end of the abnormal communication. We used a new patch-plug technique for ALVT repair which involves a measured diameter of plug to effect a snug fit in the ALVT with the patch end of the prosthesis sutured circumferentially around the aortic side of the defect. The patient had immediate narrowing of the pulse pressure and has remained asymptomatic with no aortic insufficiency for the 24-month follow-up period. A review of the world literature revealed patients who receive only medical management have 100% fatality. The surgical mortality of the reported cases approximates 20%. The surgical technique described herein offers new hope in avoiding postoperative aortic insufficiency.

LEFT VENTRICULAR ANEURYSM DIAGNOSED BY FETAL ECHOCARDIOGRAPHY

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A 32-week fetus presented with an irregular heart rhythm and an abnormal appearance of the left ventricle by ultrasound. Fetal echocardiography revealed an aneurysm of the apical portion of the left ventricle with decreased contractility but with concentric motion. Fetal arrhythmia was also diagnosed with frequent PVC's in a bigeminal pattern and short runs of ventricular tachycardia. The ventricular function remained good without signs of congestive failure.

A healthy looking male infant was delivered at 38 weeks. Frequent PVC's and short runs of ventricular tachycardia were present. Postnatal echocardiography, MRI and angiography confirmed the presence of an LV aneurysm without evidence of thrombus and ruled out other cardiac anomalies. The infant remains healthy at four months of age and is now free of significant arrhythmias in the absence of medical therapy. The contribution of fetal echocardiography in prenatal diagnosis of this rare anomaly and its sequelae is discussed.

EFFICACY AND SAFETY OF INTRAVENOUS AND ORAL NADOLOL FOR SUPRAVENTRICULAR TACHYCARDIA IN CHILDREN

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Nadolol is a long-acting, nonselective, beta-adrenoceptor-blocking agent. The efficacy and safety of nadolol in supraventricular tachycardia (SVT) was evaluated prospectively in 27 children (median age 5.5 years). Intravenous nadolol was tried in seven patients during EPS; five had no induced SVT, two had 25% decrease in SVT rate and six out of six these patients had a similar response to oral nadolol. Twelve patients received both propranolol and nadolol. Of six patients, intravenous propranolol was successful in four and unsuccessful in two patients; all six had similar response to oral nadolol. On oral propranolol, SVT was well controlled in four and failed in two patients; five out of five had similar response to oral nadolol. Twenty-six patients were treated with oral nadolol; 23 were well-controlled, two had recurrent SVT and one had SVT at 25% slower rate. During follow-up (three-36 months), compliance was good with adverse effects necessitating change of therapy in only five patients (20%). Five patients had adverse effects as follows: 1) a three-month-old had abdominal colic or nightmares, 2) two had wheezing, 3) one 14-year-old had sleep and personality changes, and 4) one 14-year-old female patient had frequent headaches. The effective dose of nadolol in 23 SVT-free children in this study ranged between 0.5 and 2.5 mg/kg/day with a median effective dose of 1 mg/kg/day. Nine SVT-free patients received 0.5 to 0.7 mg/kg/day nadolol, whereas three children failed despite doses of 1.5 to 2 mg/kg/day of nadolol. In conclusion, nadolol, once-a-day, is a safe and effective agent in children with SVT. Its long-term efficacy can be predicted by the acute response to intravenous nadolol or propranolol during EPS. The safe and effective starting oral dose of nadolol is 0.5 to 1 mg/kg/day. It is a well-tolerated drug with no significant acute or chronic adverse effects.

CARDIOINHIBITORY SYNCOPE IN INFANTS AND CHILDREN—A POTENTIALLY MALIGNANT DISORDER?

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Neurally mediated syncope is being recognized with increased frequency in children. It is generally regarded as a benign disorder and is treated medically. Most studies document a predominant vasodepressor response, a dominant cardioinhibitory response being uncommon in children. This study reports on the prevalence, clinical features, management, and outcome of patients presenting to our syncope service with dominant cardioinhibitory syncope. Of 197 consecutive patients (pts) investigated for over a five-year period for recurrent unexplained syncope, six patients (3%), four female, two male, had a dominant cardioinhibitory response. Mean age at onset of symptoms was 7.2 yrs (0.2-15 yrs). Mean age at diagnosis was 10.1 yrs (1.5-21 yrs). All but one patient (S/P Senning for TGA, VSD) had structurally normal hearts and normal ECGs. Previous diagnoses included breath holding (two pts), seizure disorder (three pts), unknown cardiac etiology (one pt). Three out of six patients had been treated with anticonvulsants. Three out of six patients had received CPR by parents or EMS, and five out of six patients had seizures during their syncopal episodes.

Autonomic evaluation was performed in five out of six patients. On orthostatic testing, each pt had sudden onset of profound bradycardia and asystole (range 8 to to 20 secs), without a prior vasodepressor component. Syncope followed in each and was associated with a brief seizure in four out of five patients. The sixth patient (aged 18 months) had sudden onset of asystole for 20 secs on Holter monitor, associated with syncope. Five out of six patients underwent pacemaker implantation (four DDD, one VVI). One patient was treated medically. Two out of five pacemaker patients had recurrence of syncope, and both became asymptomatic on fludrocortisone and salt. One of these patients (S/P Senning repair) died suddenly one year after discontinuing fludrocortisone, with normal pacemaker function. In conclusion, 1) isolated cardioinhibitory syncope is an unusual cause of recurrent syncope in infants and children and may account for some patients with apparent breath-holding, 2) brief seizure activity is commonly seen in this disorder, and 3) pacemaker implantation with or without medical therapy is effective where indicated.

THE ATRIAL APPROACH TO RADIOFREQUENCY CATHETER ABLATION OF DYSRHYTHMIAS IN CHILDREN

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Cardiac surgical ablation has been the mainstay of definitive surgery for dysrhythmias in children. Recently radiofrequency catheter ablation has been reported in adults and children. These reports have focused on a ventricular approach.

Because of the concern that producing a lesion in the ventricle might lead to ventricular dysrhythmias, we attempted radiofrequency ablation of the atrium in 10 pediatric patients, age range two months to 28 years. Their dysrythmias were six WPW, concealed one WPW, one His bundle reentry, one atrial automatic focus, and one A-V node reentry. Three of four left-sided pathways were successfully ablated, two of four right-sided, one of one His bundle, one of one right atrial automatic, and zero of one A-V node reentry. No complications occurred only the planned complete A-V block occurred. Fluoroscopy times ranged from 18-77 minutes and procedure times from four to seven hours. We believe this initial experience supports continued application of atrial radiofrequency ablation to pediatric supraventricular dysrhythmias. Intense follow-up is also indicated.

USE OF ECHOCARDIOGRAPHY TO LOCALIZE CANNULA POSITION IN NEONATES ON EXTRACORPOREAL MEMBRANE OXYGENATION

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In 1988, we began to use 2-D echo-Doppler to visualize intravascular cannulae and their relationships to cardiac structures in selected patients undergoing treatment with extracorporeal membrane oxygenation (ECMO). This series includes one additional patient from 1987 who underwent multiple follow-up 2-D echo-Doppler studies while on ECMO during the post-operative period after a Jatene procedure. The study population included 60 patients, 58 of whom had records available for review. Of these 58, we attempted to demonstrate the cannula position(s) in 39. In 38 cases we were able to locate both cannulae and demonstrate their relationships to cardiac structures. In the first two patients in the series, the arterial cannulae were positioned less than 0.5 cm from the aortic valve and were not retracted. One patient went on to develop aortic valve endocarditis and severe aortic insufficiency and eventually required homograft aortic root replacement for intractable congestive heart failure. The other patient (post-operative Jatene) developed progressive aortic insufficiency. Subsequently, we confirmed the position of the arterial cannula tip to be at least 1.5 cm above the aortic valve in 29 patients and repositioned the cannula in seven others in whom the cannula was less than 1.5 cm from the aortic valve. Two-dimensional echo-Doppler follow-up was performed after decannulation in 13 patients, and all are free of aortic insufficiency. Clinical follow up was performed in 17 and all are free of diastolic murmur. One patient died subsequent to decannulation, and autopsy study demonstrated no abnormality of the aortic valve leaflets. Five patients died while on ECMO and no autopsy or follow-up data are available.

We conclude: 1) 2-D echo-Doppler provides excellent and unambiguous data regarding cannula position and its relationship to cardiac structures, 2) aortic valve leaflet damage is unlikely when the arterial cannula tip is greater than 1.5 cm above the aortic annulus, and 3) arterial cannulae may damage the aortic valve, especially when the tip is positioned less than 0.5 cm from the valve annulus.

PARAMETRIC IMAGING OF PULMONARY BLOOD FLOW

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The presence of pulmonary vascular disease may preclude the correction of congenital heart defects. However, evaluation for pulmonary vascular disease can be imprecise. We hypothesized parametric or time imaging of pulmonary blood flow would aid in diagnosing pulmonary vascular disease. Parametric images were created and displayed from digital pulmonary angiograms (Phillips DCI-S) in four children on a Sun microsystem 4/390 computer. For each point, or pixel, in the frame of the digital angiograms, density time curves representing the change in contrast over time for that pixel were calculated. The mean ascending time for each curve was identified. The mean ascending times of all pixels were displayed as gray values then false colored, forming a physiologic picture of the time that low reached individual areas of the lungs. Patient one (five yrs) had chronic lung disease after palliation of tetralogy of Fallot with an associated AV septal defect. Pulmonary artery pressure was low-normal. However, parametric imaging revealed delayed left lung flow and transit time and decreased left lung volume. Patient two (one yr) had a VSD, lung disease and VH on ECG. Hemodynamics, hypoxic response and parametric imaging were normal. Patient three (15 mo) had an ASD and lung disease. Hemodynamics were normal on oxygen but were unmeasurable in room air because of profound hypoxemia. Parametric imaging revealed no differences in flow. Patient four (ten wks) had a complete AV septal defect. In room air, pulmonary arterial resistance was elevated. On

oxygen, pulmonary arterial pressures were unchanged but flow increased. Parametric imaging confirmed the decrease in resistance as a faster transit time on oxygen. Parametric imaging in each patient assisted in clinical decisions. We conclude parametric imaging displays the physiologic state of the lung circulation dramatizing regional and whole lung flow differences thus enhancing assessment of pulmonary hemodynamics.

THE UTILITY OF COLOR FLOW DOPPLER IN THE DIAGNOSIS OF SYSTEMIC VASCULAR MALFORMATIONS

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Systemic vascular malformations (SVM) are a rare cause of congestive heart failure in infants. However, clinical diagnosis of an SVM is exceedingly difficult. We prospectively evaluated five infants with an SVM to assess the usefulness of color flow Doppler (CFD) in the diagnosis of these lesions. Five infants (1588-3714 gms) from birth to three months were studied. All had congestive heart failure and only one patient had a bruit (cranial) at initial presentation. All patients were evaluated using cross-sectional imaging with pulse wave and CFD. Two patients had a cerebral arteriovenous fistula, two had a hepatic vascular malformation, and one had a pulmonary sequestration. CFD findings were confirmed by either CT scan or angiography. Pulsed Doppler interrogation of the transverse and abdominal aorta demonstrated diastolic runoff in four of five infants. Routine ultrasound imaging visualized a pulsating echo-free space (venous varix) within the brain substance of two infants, discrete vascular plexi (nidus) within the liver of one infant, and no unusual findings in two other infants. CFD diagnosed SVM in all. CFD features included 1) identification of all feeding vessels, nidi, venous drainage, 2) flow of uniform color within the venous varices, and 3) flow of mosaic character within the feeding vessels and nidi. Pulse wave Doppler interrogation of feeding vessels identified continuous turbulent flow with systolic velocities ranging from 1.2 to 2.5 m/sec. CFD is a valuable diagnostic tool in the evaluation of SVMs and provides information concerning anatomic detail such as size, location, feeding and draining vessels.

TRAUMATIC VENTRICULAR SEPTAL DEFECT AND ARDS IN A CHILD FOLLOWING BLUNT CHEST INJURY: TREATMENT WITH ECMO AND SURGICAL REPAIR. A CASE REPORT

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A case is presented of a three-and-a-half-year-old male who sustained blunt chest trauma in a motor vehicle accident. Initial evaluation disclosed the presence of a new cardiac murmur; 2-D Echo/ Doppler study identified a large defect in the muscular ventricular septum. Deteriorating respiratory status required mechanical ventilation and subsequent transfer to our institution for ECMO, which was initiated shortly after his arrival. Due to the combined effects of ARDS and the additional load imposed by the left-to-right shunt, the VSD was closed surgically, and the patient was weaned promptly from cardiopulmonary bypass. Postoperative course was complicated by a pericardial effusion which required drainage; cultures subsequently yielded Candida sp., and treatment was begun with Amphotericin B. Cardiovascular recovery was uneventful.

ECHOCARDIOGRAPHIC AND ANATOMIC CORRELATION OF VENTRICULAR SEPTAL DEFECT MORPHOLOGY IN NEWBORN YUCATAN MICROPIGS

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Using a Yucatan micropig strain which has a high incidence of ventricular septal defects (VSDs) in its off spring, we correlated two-dimensional and Doppler echocardiography of VSD morphology and size with autopsy examination. A spectrum of perimembranous (P), doubly-committed subarterial (DS) and muscular outlet (MO) VSDs was found. Two-dimensional echo/color Doppler examination was performed on 23 piglets weighing 1.2-3.6 (mean 2.3) kgs and studied between four and 18 days of life. Echo diagnosed 13 VSDs in 13/23 subjects; morphology was P=9, DS=2, and MO=2. At autopsy, the presence and location of VSDs was confirmed in 13/13 cases. No additional VSDs were found. Diameters of VSDs by echo and color Doppler flow jet width were 1.0-5.0 mm (mean 4 .2) and by autopsy measurement were 1.0 - 6. 0 mm (mean 2.9). Using aortic annulus size as an internal control for tissue shrinkage during fixation, echo/color Doppler VSD sizing overestimated VSD diameter by 17%. In conclusion echo/Doppler accurately identified the morphology and size of VSDs in newborn Yucatan micropigs. Echocardiographic classification of VSD morphology in vivo will facilitate future research on specific VSD types.

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