

## Abstracts

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### Abnormalities in lymphocyte populations in infants with neural crest cardiac defects can predict nonsurvivability

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Children with neural crest congenital heart defects and manifestations of the DiGeorge spectrum of anomalies are known to have a high incidence of mortality despite improvements in management of congenital heart disease. Embryologically, neural crest tissue is known to contribute to development of aortic arch and conotruncal regions of the heart, as well as parts of connective tissue of facial structures, thymus, thyroid and parathyroid glands. Previous studies demonstrate various significant immune deficiencies in patients with classic DiGeorge syndrome; however, the cardiac defects are not inclusive of those associated with abnormal neural crest development. We hypothesize that children with neural crest congenital heart disease defects (truncus arteriosus, interrupted aortic arch, double outlet right ventricle, tetralogy of Fallot, single ventricle), but not manifesting classic DiGeorge syndrome, have subtle immune deficiencies which are clinically significant. Nonsurvivors had normal white blood cell counts. Fifty-seven percent of these had a lower number of total lymphocytes and B lymphocytes. T cell subsets were also decreased in nonsurvivors (CD3 66%, CD4 78% and CD8 89%). The significance of these data is that abnormalities in lymphocyte populations and subsets are strong predictors of nonsurvivability in infants with neural crest cardiovascular defects. In spite of being treated as if immune deficient (administration of irradiated and filtered blood products, infection control measures), these infants were nonsurvivors. Additional studies are ongoing to compare nonsurvivors with survivors as well as with an infant control group.

### Closed loop temperature controlled radiofrequency ablation prevents impedance rises

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We performed nine ablation sessions in the first nine consecutive patients using a new temperature controlled radiofrequency catheter ablation (RFCA) system. Using a thermocouple in the 4 mm tip of a 7 French omnidirectional steerable catheter and a control system which used up to 50 watts in attempts to reach 70 °C, we made 107 RFCA lesions. Twenty-six lesions reached 70 °C and the output was automatically decreased to a mean of 24 watts. During 81 lesions 50 watts were used for the entire 30-60 seconds without reaching 70 °C. No impedance rises occurred. Beginning impedances averaged 100 and peak impedance were never over 150 Q. No patient had an inducible dysrhythmia during a one hour isoproterenol and programmed stimulation session immediately after RFCA. In comparison in nine patients matched for dysrhythmias using standard catheters and a standard radiofrequency generator, 3.2 impedance rises occurred per patient range 0-13. The initial impedance for this system averaged 122 Q. Twenty to 48 watts, mean 33 watts were used. One hundred thirty-three lesions were made in this group of patients. Each patient had no inducible dysrhythmia. The fluoroscopy time averaged 37 minutes in the standard group versus 28 minutes in the temperature controlled group. Closed loop temperature control prevents impedance rises and shortens fluoroscopy time of RFCA.

### Results of the arterial switch (Jatene) procedure

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The arterial switch (Jatene) procedure has been the preferred procedure for correction of selected patients with transposition of the great arteries (TGA) at our institution since 1985. We reviewed the results of this procedure performed consecutively on fifty children. All underwent diagnostic cardiac catheterization and balloon atrial septostomy. Age at the time of surgery ranged from five days to 29 months with 39/50 (78%) newborns. 34/50 (68%) had TGA/intact ventricular septum, 9/50 (18%) had TGA/VSD, and 2/50 (4%) had other cardiac anomalies. Coronary artery anatomy was "normal" in 46/50 (92%); in 4/50 (8%), the circumflex coronary artery arose from the right coronary artery. No perioperative deaths occurred. There were three late deaths (6%); two not directly related to cardiac disease and one due to a left ventricular cardiomyopathy but with normal coronary arteries at autopsy. At latest follow-up, all of the survivors are asymptomatic. 49/50 (98%) have experienced normal growth to date. 21/50 (42%) have mild stenosis at the pulmonary valve or pulmonary anastomosis. 4/50 (8%) underwent reoperation or angioplasty for severe pulmonary stenosis; one required reoperation for stenosis at the aortic anastomosis. 7/50 (14%) have other minor residual hemodynamic abnormalities (trivial neo-aortic insufficiency or stenosis, small VSD, up to moderate pulmonary insufficiency, or mildly decreased contractility). One patient developed persistent complete heart block after VSD closure necessitating pacing. Excellent functional results can be obtained with the arterial switch procedure in selected patients with TGA. No perioperative mortality occurred and late mortality was rare. Significant residual cardiac abnormalities are uncommon. While minor residual hemodynamic abnormalities appear well tolerated over the intermediate period, continued follow-up is warranted.

### Intracranial hemodynamics in recurrent syncope

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The etiologic mechanisms for recurrent syncope in early adolescence are not well defined. The objective of this study was to determine the relationship between changes in blood pressure (BP) and indices of cerebral hemodynamics in symptomatic children. Seven boys and one girl (ages 9-16 years) were referred for recurrent symptomatology. During head-up tilt testing (75°), surface ECG, BP, heart rate (HR) and blood flow velocities (BFV) of the middle cerebral artery (MCA) by transcranial Doppler (TCD) were recorded every 1-3 minutes. All data were analyzed using repeated measures analysis of variance. Symptomatic children showed: 1) A significant drop in systolic, diastolic and mean BP's (p<0.05). There was no significant variation in HR. 2) A significant reduction in mean and end diastolic BFV with increase in pulsatility indices of the MCA by TCD (p<0.05). 3) A compensatory overshoot of mean BFV on normalizing the patient position to supine. We conclude that the vasodepressor responses noted were associated with significantly abnormal cerebral hemodynamics. It appears that although the vasoreactivity of the cerebral vessels is intact, the syncopal patients demonstrate a vascular fatigue or response to abnormal stimulation manifested as a sudden reduction in cerebral BFV's in the erect posture.

#### Factors influencing survival of patients with congenital heart disease, 1981-1989

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Between 1981 and 1989, 4390 cases of congenital heart disease (CHD) were registered by the BWIS, a case-control study of environmental risk factors in CHD. Virtually all cases of CHD born in the entire state of Maryland, the District of Columbia and six counties of northern Virginia were identified. To be included in the study, a case must have been born in an area hospital and have had the diagnosis established by echocardiography, cardiac catheterization, surgery or autopsy. Only exclusions to entrance into the study were patent ductus arteriosus (PDA) in premature infants, arrhythmia and birth in a military hospital. Cases were updated at one year to account for changes in cardiac diagnosis which occurred in 9% of cases. The all cause mortality at one year was 18%. Since 28% of cases had associated non-cardiac malformations, it is difficult from an epidemiologic standpoint to ascertain whether death is causally related to the cardiac or to the non-cardiac malformation. We, therefore, have chosen to discuss only "all cause" mortality which is a reflection of the entire clinical situation faced by the pediatric cardiologist. There was a marked excess of infants with low birth weight (less than 2,500 grams) in all case groups except complete transposition (TGA) and bicuspid aortic valve (BAV). Although the rate of prematurity (gestational age less than 37 weeks) was elevated over the population average of 6% in our controls, birth weight was reduced proportionately more than gestational age. In all groups except TGA, single ventricle and BAV, up to 30% of infants were small for gestational age (birth weight below tenth percentile). The lower the birth weight, the higher the mortality. The case fatality at one year of infants with birth weight under 1,500 grams was four times that of infants whose birth weights were over 4,000 grams (35% vs. 8%). The lower the age of diagnosis, the higher the mortality. CHD diagnosed less than one week had a 39% one year mortality and 1/4 did not survive four weeks. Diagnosis greater than 12 weeks had only a 4.9% one year mortality. There were trends in mortality in specific lesions. Mortality from hypoplastic left heart and total anomalous pulmonary venous return showed a marked decrease which may have been related to improved diagnosis, advances in surgical technique or to termination of pregnancy in severely involved cases. Mortality from VSD showed a marked decline due to increased detection of small muscular defects by color flow Doppler echocardiography. Surgical mortality was not addressed in this study, but age of surgery was a factor in one year survival. Birth weight, age of diagnosis, need for early surgery and associated non-cardiac malformations are causally related to one year survival in cases with CHD and compound the decision process relative to care of cases with CHD.

#### Medical management of cardioinhibitory syncope in the pediatric population

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Pacemaker therapy has been advocated for patients (pts) with neurocardiogenic syncope (NCS) who manifest the cardioinhibitory (CI) response (asystole > 5 sec). We postulate that CI syncope can be managed medically. From 1/91 to 1/93, six pediatric pts (8-17 yrs, m=12 yrs) were diagnosed with CI syncope (asystole of 5-34 sec duration, m=15 sec) at head-up tilt (HUT) testing. Asystole occurred from 1-16 min (m=6 min) after onset of HUT. 4/6 pts were treated with alpha-adrenergic agonist, acutely during HUT with IV phenylephrine therapy and as outpatients with oral pseudoephedrine. Acute IV phenylephrine therapy with repeat HUT in 3/4 pts yielded no syncope and a mixed (combined vasodepressor and cardioinhibitory) response without asystole in one pt. Oral pseudoephedrine therapy was successful in 3/4 while additional Florinef therapy was required in one who had a mixed response (no asystole) at follow-up HUT. Oral betablocker was initial therapy in 2/6 pts and was successful in one. Additional Florinef therapy was required in one pt who had a mixed response (no asystole) at follow-up HUT. Pt follow-up has been from 3-22 months (m=13 months). All pts are improved with no further syncope, fewer presyncopal episodes, and no side effects. In conclusion, these six pts with CI syncope were treated medically without complications. We speculate that vigorous medical therapy prevents the initiation of NCS and prevents CI syncope. Since NCS may be a self-limiting disease, medical management has significant benefits over pacemaker therapy.

#### Scimitar syndrome—natural history and indications for surgery

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Scimitar Syndrome (SS) is characterized by 1) hypoplasia of the right lung, 2) systemic arterial supply to a portion of the right lung, and 3) venous drainage of all or part of the right lung via an anomalous vein to the IVC. Nineteen patients, 10 men and nine women, with SS (mean age 29.4±22.5 years) have been treated at our institution. Complete follow-up is available on 18 patients (94.7%), with a mean follow-up of 11.1±8.5 years (range 0.1 to 30.1 years). Venous drainage of the entire right lung to the IVC was found in nine patients, a portion of the right lung in five patients, and no evaluated in five patients. Systemic arteries supplying the right lung were present in eight patients, absent in six patients, and not evaluated in five patients. An ASD was present in three patients. Mean PA systolic and diastolic pressures were 30.6±11.3 mm Hg, and 13.3±7.8 mm Hg, respectively (n=13). Mean Qp/Qs was 1.8±0.5 (n=12). Mean FVC and FEV1 were 76.8±18.7 and 72.2±26.0% of predicted respectively (n=9). Thirteen patients had hypoplasia of the right lung and 10 patients had abnormal right bronchial anatomy. Twelve patients were treated medically and seven patients were treated surgically. Surgical procedures included 1) ligation of systemic arteries (n=1), 2) closure of ASD (n=1), 3) baffling of venous return to the LA via an existing or surgically created ASD (n=2), 4) reimplantation of the anomalous vein (n=2), and 5) pneumonectomy after reimplantation of the anomalous vein at another institution (n=1). Two patients in the nonsurgical group have died (one in infancy and one of pancreatic cancer at age 71). Four patients have died in the surgical group (two patients died after reimplantation of the anomalous vein, one patient died of hepatitis at age 35 and one died of emphysema at age 71. All patients undergoing reimplantation of the anomalous vein either died or required pneumonectomy. No patients treated medically have required surgery. Previous reports (n=124) of SS describe a total of 502 patients. Survival data are available for 229 of these patients. Meta-analysis shows a Kaplan-Meier survival estimate of 72.1±0.1% at 55 years. Age at diagnosis <1 year (p<0.001) is a significant risk factor for death. Patients presenting at an age <1 year are more likely to be symptomatic (p<0.001), with associated anomalies (p<0.001), a systemic arterial supply to the right lung (p=0.012), pulmonary hypertension (p<0.002), and a Qp/Qs >2.0 (p<0.003). SS is a benign condition in most adult patients. In contrast, patients presenting in infancy are critically ill with a high mortality. Attempts at reimplantation of the anomalous vein may be hazardous and therapy should be directed towards decreasing the magnitude of the left-to-right shunt. Because of the frequency of anatomic abnormalities of the right lung, pulmonary resection may be the preferred operation in adults with significant symptoms or pulmonary hypertension.

#### Initial experience with multiplane transesophageal echocardiography in patients with congenital heart disease

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The advent of a 180° rotatable multiplane annular phased array TEE probe has permitted visualization of intracardiac anatomy in virtually any plane. We have commenced multiplane intraoperative TEE evaluation of suitable size patients to facilitate assessment of immediate surgical results. The spherical probe (15 mm diameter) affixed to a 10.8 mm diameter endoscope shaft was passed without difficulty in patients weighing over 30 kg, however, this could not be easily achieved in an intubated 15 kg patient. Unique views of the mitral valve were obtained in two patients with atrioventricular septal defect by transgastric imaging. True short-axis images, unobtainable with biplanar imaging, were produced with 35° to 55° rotation from the horizontal plane (0°), which delineated detailed anatomy of the cleft mitral valves. Novel long-axial views of the right and left ventricular outflow tracts were easily obtained by scanning from flexed transgastric position with 0° to 135° rotation, and perfect alignment for PW and CW Doppler evaluation was obtained. The entire extent of the atrial septum was reliably interrogated. Excellent short and long-axial visualization of the aortic valve was achieved. Color flow Doppler provided precise localization of AV and aortic valvar insufficiencies. Our continuing evaluation and use of multiplane TEE reveals a unique and valuable application in patients with congenital heart disease.

**The Fontan fenestration—open or closed?**

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Creation of an adjustable fenestration in the intraatrial baffle has contributed to a marked reduction in morbidity and mortality following Fontan repair. Prior to the advent of the fenestration, cardiac output following Fontan was limited by the amount of blood which could be driven through the pulmonary circulation by a venous pressure head. The Fontan fenestration allows shunting of venous blood directly into the systemic circuit to support cardiac output in the perioperative period. While the issue of suitability for closure of the fenestration remains to be resolved, we have chosen a CVP greater than 20 mm Hg or clinical signs of venous engorgement, such as prolonged pleural effusion or ascites, as contraindications to fenestration closure. We reviewed the cardiac catheterization data and medical records of patients who underwent the fenestrated Fontan procedure at the Ochsner Hospital to determine whether there were any preoperative factors which might predict the need to leave fenestrations open. Between February 1992 and July 1993, eleven patients received the fenestrated Fontan procedure. There was one perioperative death secondary to junctional tachycardia, and this patient was excluded from analysis. Diagnoses were tricuspid atresia in five patients, double inlet left ventricle in three, and complex forms of univentricular heart in three. There were five patients in each group. Age was  $67 \pm 10$  months in the group with fenestrations closed (CF) and  $72 \pm 20$  months in patients with open fenestrations (OF). Fenestrations were left open for CVP greater than 20 mm Hg in three patients, for prolonged chylous pleural effusion in one, and for ascites in the remaining patient. All CF patients and 3/5 OF patients had received previous Blalock-Taussig shunts. In both groups, three of five patients had received intermediate bidirectional Glenn procedures. Pulmonary vascular resistance was  $1.1 \pm 0.2$  Wood units in patients with CF and  $1.8 \pm 0.4$  Wood units in OF patients. The mean pulmonary artery pressure in CF patients was  $20 \pm 2$  mm Hg and  $17 \pm 3$  mm Hg in OF patients. McGoon indices for patients in the CF group were  $2.4 \pm 0.4$  and were  $1.2 \pm 0.1$  in the OF group. Nakata indices were  $347 \pm 77$  versus  $128 \pm 14$  in CF versus OF patients. All patients with Nakata indices greater than 200 were candidates for closure of the fenestration, while all patients with indices less than 200 were not. In addition, a McGoon index greater than two accurately predicted successful closure of the fenestration. Using McGoon and Nakata indices derived from preoperative cardiac catheterization, it may be possible to predict which patients will tolerate closure of their fenestrations following Fontan repair.

**MRI imaging of the arterial switch operation for D-TGA**

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The arterial switch operation is now considered the operation of choice for patients with DTGA. The long-term results of this procedure are yet to be confirmed. The purpose of this report is to describe the MRI findings in children who are one to two years postoperation. Patients were imaged on a Phillips 0.5 Tesla Magnet. Ten patients were imaged and compared to echocardiography and angiography within one week. Attention was placed on the pulmonary and aortic anastomosis as well as distal pulmonary artery anatomy. All ten patients were adequately imaged with MRI. Five patients had narrowing of the distal pulmonary artery as it coursed around the centrally placed aorta (Four on the left, one on both left and right). Echocardiography could not visualize any distal narrowing, but did show a left pulmonary artery gradient of 20 mm Hg. Angiography showed three of the five with narrowing. However, no significant hemodynamic stenosis was noted by direct pullback. MRI is superior at seeing distal pulmonary arteries after the Jatene procedure due to its unlimited plains of imaging. Fifty percent of the children after the Jatene procedure showed narrowing of one or the other of the distal pulmonary arteries. The narrowing has not, however, been hemodynamically significant at this time.

**QTc interval prolongation during exercise—a nonspecific finding or indicative of long QT syndrome**

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Exercise testing has been recommended in the evaluation of long QT syndrome (LQTS), yet normal and abnormal responses of the QTc interval with exercise are not well established. We reviewed exercise treadmill studies in 32 young patients aged 6.8-17.2 yrs (mean  $12.7 \pm 2.9$  yrs) without structural heart disease or documented Torsades de Pointes. In fifteen patients, the indication for study included possible LQTS on the basis of unexplained syncope (11 pts), family history of LQTS (three pts), or QTc  $> 0.45$  sec on a resting ECG (one pt). Seventeen pts underwent evaluation for other indications and were not suspected to have LQTS (NQT) prior to the exercise study. All patients displayed normal exercise tolerance using age-related norms for the Standard Bruce Protocol, and exercise was terminated due to fatigue in all patients. Mean QTc were calculated using the Bazett formula under at rest, during peak exercise, during early recovery ( $< 3.5$  min post-exercise), and during late recovery (5-10 min post-exercise). Data are summarized below for all pts and separately for those patients in whom possible LQTS was or was not considered among the indications for exercise. The numbers of pts demonstrating QTc  $> 0.45$  sec at any point during the test are also indicated for each group.

	n	QTc rest	QTc peak	QTc early	QTc late	QTc $> 0.45$ sec
All pts	32	$0.42 \pm 0.03$	$0.42 \pm 0.03$	$0.43 \pm 0.04$	$0.44 \pm 0.03$	23/32
No LQTS	17	$0.41 \pm 0.03$	$0.42 \pm 0.03$	$0.43 \pm 0.04$	$0.44 \pm 0.02$	11/17
Pos LQTS	15	$0.42 \pm 0.02$	$> 0.43 \pm 0.03$	$0.42 \pm 0.03$	$0.45 \pm 0.03$	12/15

No pts developed syncope, Torsade de Pointes, or bizarre T waves. Five pts (one possible LQTS, four non-LQTS) developed isolated VPBs during recovery. Two pts (one possible LQTS, one non-LQTS) each had single QTc measurements  $> 0.50$  sec during the study, but normal QTc during other phases of the study. In no cases were these findings alone considered sufficient to confirm a diagnosis of LQTS. Alternative diagnoses were eventually established in six syncope patients who also underwent tilt testing. We conclude that QTc prolongation (QTc  $> 0.45$  sec measured from a standard ECG) is a common finding during normal exercise and recovery in young patients. QTc prolongation occurred with equal frequency in this population regardless of the apparent likelihood of LQTS. While such QTc prolongation during exercise may warrant further evaluation when LQTS is suspected, it should not be considered diagnostic in the absence of Torsade de Pointes, characteristic T wave changes, or family history of long QT syndrome.

**Failure of echo-Doppler analysis of cardiac diastolic function to predict rejection in pediatric heart transplant patients**

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Echo-Doppler (ED) indices of cardiac diastolic function (CDF) have been proposed as a means to noninvasively diagnose cardiac allograft rejection (R). As the interpatient variability in measurement of ED indices is high, comparison of data from different patients is often confounded. To test the usefulness of ED indices of CDF in predicting R, a blinded comparison of several ED indices of CDF was performed within 24 hours of biopsy proven R and non-rejection (N). Each patient served as his/her own control. Measurements included: left ventricular isovolumic relaxation time, mitral and tricuspid peak E and A wave velocities, E wave deceleration time, and fraction of filling in the first one-third of diastole and during atrial contraction. A total of 22 pairs of R [ISHLT grade IB (5), II (8), IIIA (8) and IV (1)] and N (separated by  $6 \pm 2$  weeks) were identified in 10 patients. No differences were found between R and N for any measured mitral or tricuspid ED index of CDF (paired t test,  $p > 0.05$ ). We conclude that, after controlling for interpatient variability, ED analysis of CDF does not reliably identify R. More reliable and reproducible noninvasive means of diagnosing R should be identified.

#### Improved survival following heart surgery with extracorporeal life support—impact of surgical procedure

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The impact of extracorporeal life support (ECLS) on mortality for congenital heart surgery is controversial and uncertain. In infants  $\leq 1$  yr, operative mortality following cardiopulmonary bypass surgery at Arkansas Children's Hospital for the four years before ECLS was available was 15.5% (33/213). Since 1989 (through 1/93), the use of ECLS has reduced mortality to 9.8% (25/256), a difference mainly attributable to ECLS. Survival was significantly improved by ECLS in infants less than one month of age especially following definitive repair versus palliation. To further evaluate the effect of the type surgical procedure on long-term outcome, the 56 patients who received cardiac ECLS were reviewed. Of those, 38 patients (age one day to 5.5 yr) had received single support ECLS for circulatory collapse following cardiac surgery, with 28 patients weaned from support, 21 extubated and discharged home, and 18 remaining long-term survivors. While the time following surgery and cardiopulmonary bypass as indicated by the site of the initiation of ECLS (ICU versus OR) had no effect on the number of patients weaning from ECLS, long-term survival was improved in patients when ECLS was initiated in the ICU versus OR, ( $p < 0.05$ ) and in patients who had complete repair versus palliative surgery ( $p < 0.05$ ). The effect of the type of operative procedure was especially pronounced in the patients where ECLS was necessary in the OR following a palliative surgical procedure (no long-term survivors in nine patients). ECLS in patients greater than one year of age also tended towards better outcomes. Deaths while on ECLS were secondary to the patient's primary cardiac lesion and failure to respond to ECLS. No deaths occurred secondary to ECLS complications. We conclude 1) ECLS can be successfully initiated following cardiopulmonary bypass surgery for congenital heart defects with a decrease in long-term surgical mortality, 2) successful ECLS can be initiated in the OR, however 3) ECLS following palliative procedures has a higher mortality, especially when required immediately following cardiopulmonary bypass indicating 4) the cardiac defect and type of surgery are the main predictors for poor outcome of post surgical cardiac ECLS.

#### Does intraoperative transesophageal echocardiography predict severity of late mitral regurgitation after atrioventricular septal defect repair

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Intraoperative epicardial (EPI) and transesophageal echocardiography (TEE) have been shown to be useful in the evaluation of mitral valve function following surgery for complex congenital heart defects. To evaluate the long-term significance of intraoperative echocardiographic findings, we compared TEE and EPI detected mitral regurgitation (MR) with follow-up transthoracic echo in 31 consecutive surviving patients after repair of atrioventricular septal defect (AVSD). TEE: Of seventeen patients [operative dates 5/90-1/92, age at operation 2-17 (median 6) months, follow-up 3-15 (median 8) months], nine had isolated mild MR, and four had isolated moderate or severe MR. In these patients, there was no change in the severity of MR at follow-up. In two of the 17 cases, moderate MR by TEE was associated with poor left ventricular function in the early post bypass period. This improved to mild MR as left ventricular function normalized postoperatively. MR with mitral stenosis by TEE in two patients with unbalanced AVSD was associated with progressive mitral dysfunction at follow-up. EPI: Of 14 patients [operative dates 4/89-6/92, age at operation 2.5-42 (median 7) months, follow-up 3-42 (median 12) months], seven had mild intraoperative MR, two of whom had moderate or severe insufficiency at follow-up. The remaining seven had moderate MR at operation. Four of these had only mild insufficiency at follow-up. We conclude that, 1) transesophageal intraoperative echocardiography accurately predicted the severity of mitral valve insufficiency in the follow-up period after repair of atrioventricular septal defect; 2) epicardial examination provided less predictive information about long term mitral valve function; and 3) mitral stenosis identified by intraoperative echo predicted continued severe mitral valve dysfunction.

#### Asymptomatic prosthetic valve dysfunction

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Because of two deaths caused by sudden prosthetic valve dysfunction, we began a program of periodic surveillance by echocardiography/Doppler examination and fluoroscopy in 1989. All patients studied, as well as the two patients who died, had mitral valve replacement (MVR) with a St. Jude's prosthesis. Since 1989, eighteen patients with MVR have been regularly followed with echocardiography/Doppler and fluoroscopy. These studies were to be performed at six month intervals and patient compliance was generally good. The patients ranged in age from seven months to sixteen years at the time of MVR. The primary cardiac defects in these patients were partial atrioventricular canal defect (6), complete atrioventricular canal defect (1), complete atrioventricular canal defect with tetralogy of Fallot (1), congenital mitral stenosis (1), congenital mitral regurgitation (5), anomalous left coronary artery with mitral regurgitation (1), arterial switch operation with mitral regurgitation (1), and ventricular inversion with left atrioventricular valve regurgitation (2). The echocardiography/Doppler study included estimation of mitral valve area by the pressure half-time method. Fluoroscopy was performed with the patient in a supine position and with zero craniocaudal angulation. The opening angle of the two leaflets was estimated by viewing each leaflet on end and noting the degree of non-parallelism. Normally, the maximum extent of opening produces an angle of approximately 10 degrees. During the study period, three patients developed an opening angle of greater than 25 degrees. All of these patients returned to the "normal range" spontaneously and without any specific intervention other than careful control of Warfarin therapy. All patients returned to the "normal range" within a period of three and a half months. No patient was symptomatic with the apparent prosthetic valve dysfunction. Some factors associated with apparent prosthetic valve dysfunction in these patients were analyzed. Prosthetic valve dysfunction with impaired opening occurs in asymptomatic patients. The clinical relevance of this finding is unclear.

#### Repeat radiofrequency catheter ablation procedures in the pediatric population—"if at first you don't succeed..."

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This study examines our experience with repeat radiofrequency catheter ablation procedures in pediatric patients whose initial radiofrequency procedure failed to control their tachycardia symptoms. Since initiation of radiofrequency catheter ablation in March of 1991 there have been a total of 175 procedures done on 149 pediatric patients. A total of 22 pediatric patients needed 48 total repeat procedures after recurrence of their initial clinical tachycardia. In this patient group, five patients had atrioventricular node reentry tachycardia, two patients had ventricular tachycardia, and 15 patients had supraventricular tachycardia secondary to an atrioventricular bypass tract. The average patient age was 12 years (range five months to 23 years). The average patient weight was 45 kilograms (range seven kilograms to 90 kilograms). In this group of 22 patients, 17 patients (78%) had elimination of their tachycardia circuit during the second procedure, and 4/5 of these failures were successfully treated by a third procedure, making the overall success rate of repeat procedures 95%. The average length of the repeat procedures was 80% of the average initial procedures and the average fluoroscopy time of the repeat procedures was 60% of the initial procedures (56 minutes vs. 32 minutes). In conclusion, this study demonstrates that the success rate for repeat radiofrequency catheter ablation procedures in the pediatric population approaches 95%, and that failure of tachycardia control at the initial or subsequent procedure does not preclude the possibility of ultimate success.

### Can heart rate variability analysis predict response to orthostatic testing in children?

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Heart rate variability (HRV), as an indicator of autonomic tone, may differ in children with the various subtypes of neurally mediated syncope. To elucidate this, HRV on 24 hour Holter recordings were compared between 20 orthostatic test positive (OT+) adolescents and 20 age- and sex-matched controls. Time domain (TD), [mean RR, standard deviation about the mean (s.d. RR), standard deviation of five minute RR intervals (SDANN), mean of all SDANNs (SD), proportion of adjacent RRs more than 50 msec different (pNN50), root mean square of difference of successive RRs (rMSSD)], and frequency domain (FD), [low frequency (0.04-0.15 Hz), high frequency (0.15-0.40 Hz), all frequencies (0.01-1.00 Hz), and ratios] variables for each patient were compared statistically to determine which factor, or combination of factors, best separated the groups. OT+ patients were further subclassified by type of syncope, [vasodepressor (VDS), cardioinhibitory (CIS), or mixed]. Stepwise discriminant analysis revealed no significant variable discriminating between OT+ and controls. However, comparing OT+ subtype and controls the TD variable of mean RR interval successfully separated the mixed syncope group with 70% accuracy ( $p < 0.005$ ). Mean RR for mixed type was  $654 \pm 57$  ms, for VDS was  $705 \pm 66$  ms, for CIS was  $881 \pm 191$  ms, and for controls was  $695 \pm 90$  ms. In conclusion, HRV did not identify children prone to syncope. However, TD analysis helped to subclassify the response to orthostatic testing. Therefore, HRV may be an useful adjunct to the non-invasive evaluation of syncope.

### Reversible long QT interval in a child with triple A (Allgrove) syndrome

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Long QT interval syndrome (LQTS) is a rare but dangerous entity, which may lead to syncope, seizures or even sudden death. The triple A (Allgrove) syndrome is a rare familial disorder characterized by adrenal insufficiency, alacrima and esophageal dysmotility (achalasia). We report the simultaneous occurrence of T wave abnormalities frequently seen in patients with LQTS in a patient with Allgrove syndrome. RH presented at age four years after her mother found her unarousable and unresponsive at approximately 6 AM. She had not eaten well in the previous 36 hours. She was taken to an emergency room and junctional bradycardia was noted. The initial 12-lead ECG showed sinus rhythm with a heart rate of 108 and a QT and QTc of 0.36 and 0.50. Electrolytes were within normal limits, and the blood glucose was 93. Repeat laboratories after transfer to our hospital were also normal except for a glucose of 40. ECG showed a prolonged QT interval (QT=0.36, QTc=0.50). The ECG continued to show a prolonged QT interval of 0.41 and QTc of 0.49. Holter monitoring showed no ectopy, but persistent prolongation of the QT and QTc intervals as well as changing polarity of the T waves. Morning serum cortisol was 5.1 mg/dl, (nl 6-18 mg/dl), and no significant rise in cortisol occurred after two ACTH stimulation tests. Further endocrinologic evaluation resulted in the diagnosis of ACTH nonresponsiveness. In four months of cortisol therapy, the EKG returned to normal, including a QT and QTc interval of 0.34 and 0.43 seconds. This case emphasizes the need for metabolic evaluation in some patients with long QT intervals. It also raises questions about the relationship of the neural adrenal axis to ECG abnormalities found in patients with LQTS.

### The influence of the learning curve on the efficacy of radiofrequency catheter ablation in the pediatric age group

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In the adult population, success rates of radiofrequency catheter ablation (RCA) have been shown to be dependant on operator experience. To evaluate the effects of the "learning curve" on the success rate in children, we studied the outcomes of the first 148 consecutive pediatric and young adult patients who had RCA for a single tachycardia. The age range was one month to 23.4 years (mean  $11.7 \pm 5.4$  yrs). The weights ranged from 3.4 to 128 kgs (mean  $48.8 \pm 25.9$  kgs). There were 82 males and 66 females. The patients were divided into three groups by chronological order according to their catheterization dates. Time I included the first 48 patients seen over the first 10 months, Time II the second 50 patients seen over the following 8.5 months, and Time III with the last 50 patients seen over the last 8.5 months. The diagnoses of these patients were divided into groups: 1) SVT due to concealed pathways (SVT), 2) SVT due to overt Wolff-Parkinson-White (WPW), and 3) non-pathway tachycardias including AVNRT, AET, VT, and AF (NPT). Overall success rates were as follows: Time I—58%, Time II—78%, and Time III—76%. The improvement in efficacy from Time I to Time II was paralleled by improved success rate in the SVT group (75% to 85%), and the WPW group (48% to 86%). Although the overall success rate for Time II and Time III were similar, the absolute number of procedures performed on patients with the more difficult tachycardias (NPT) doubled. We conclude that operator experience influences the efficacy rate of first time RCA procedures in the pediatric population. While the efficacy rate of first time procedures seems to plateau, the difficulty of cases which are considered for RCA increases, illustrating another dimension of the learning curve.

### The utility of a dedicated magnetic resonance imaging unit for pediatric cardiology practice

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Magnetic resonance imaging (MRI) is a noninvasive test which does not require injection of contrast agents. Our center uses a Phillips 0.5 tesla Gyroscan MRI unit for the sole purpose of diagnosing congenital and acquired heart disease in children. This study was performed to evaluate the utility of a dedicated MRI unit in a pediatric cardiology patient. In the first year of service we performed 309 scans in 291 patients. Patient age ranged one day to 34 years with 24 (8%) <1 month old and 49 (17%) <1 year old. Weight ranged from two to 181 kg. The average scan time was 80 minutes. Seven patients were intubated, 12 had intravenous drips, 187 were sedated and 54 had cyanotic heart disease. The only complication was mild apnea in two infants. Our standard medical management and practice routine was altered in 40% of patients scanned. The number of diagnostic catheterizations performed the same year decreased by 36%. MRI images were most useful when evaluating pulmonary artery, pulmonary vein, aortic and complex intracardiac anatomy. Our experience shows that pediatric cardiac MRI is both safe and practical. It replaces catheterization in as many as one-third of patients. MRI should be a standard imaging modality for diagnosis of children with heart disease.

**MRI myocardial tagging for localization of accessory pathways in Wolff-Parkinson-White**

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MRI with myocardial tagging (MT) using spatially modulated magnetization (SPAMM) can be used to assess myocardial wall motion. We theorized that MT with SPAMM could identify earliest ventricular contraction corresponding to WPW preexcitation at the insertion of the accessory pathway (AP). Seventeen patients (2-12 yrs), with WPW scheduled for radiofrequency ablation (RFA), were studied using MT with fast gradient echo-cine, gated to sinus rhythm delta wave EKG on a 1.5 T MRI system. Short axis 8 mm thick tagged images were acquired along the ventricular aspect of the A-V grooves. Cine sequences were visually analyzed to identify early contraction, blind to RFA. Nineteen AP's were localized by RFA. MT correctly located 1/3 right (rt) anterior, 1/1 rt posterior, 1/1 rt anterior septal, 1/2 rt post lateral, 1/2 rt posterior septal, 0/2 rt anterior lateral, 4/5 left (lt) lateral, 1/1 lt anterior lateral and 0/1 lt posterior septal AP's. For four additional AP's, MT correctly localized early contraction to the A-V groove but differed from RFA by a 45-60° sector. These included one rt posterior lateral, one rt posterior septal and two rt anterior lateral AP's. Two patients were found at RFA to have two separate AP's. These were not correctly identified by MRI. There was no MT/RFA concordance for the remaining two rt and two lt AP's studied. One MT study reported two AP's where only one was found at RFA. Interference from atrial appendage and ventricular outflow tracts, tangential AP's, limits of EKG gating, and MRI resolution may contribute to inaccurate MT/RFA matching. These findings suggest that MT with SPAMM, with further refinement, can be an accurate and reliable noninvasive method for AP localization.

**Partial versus complete neural crest ablation in a chick embryo model of heart defects**

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Ablation of premigratory cardiac neural crest produces structural heart defects such as truncus arteriosus and aortic arch anomalies. Cardiac neural crest ablations also produce abnormal hemodynamic variables in early cardiovascular development. We hypothesize that ablation of the complete cardiac neural crest (destined for arches 3, 4 and 6) will cause more severely depressed indices of contractility than the partial cardiac neural crest (destined for arches 3 and 4). Using 100 frames/sec cinephotographic film, end-diastolic and end-systolic dimensions of the primitive ventricle were digitized in chick embryos with a looped cardiac tube at stages 16 and 18. There is a significant effect by stage across all variables and a significant interaction between stage and group for shortening fraction and ejection fraction. If the complete or partial cardiac neural crest is ablated in experimental embryos, it produces depressed contractility indices at stage 18 of incubation. At stage 16, only the complete neural crest-ablated embryos have decreased indices of contractility. Thus, it is necessary to have 6<sup>th</sup> arch neural crest for normal ventricular contractility. Ventricular dysfunction and enlargement may cause incomplete looping of the cardiac tube. This early developmental abnormality correlates with more malalignment defects such as a double outlet right ventricle or double inlet left ventricle seen in embryos after complete compared to partial cardiac neural crest ablation. In conclusion, these early hemodynamic alterations provide an explanation for the occurrence of a wide spectrum of outflow tract, inflow tract, and aortic arch cardiovascular defects after neural crest ablation.

**Pediatric heart, heart-lung, and lung transplantation**

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From 1984 to 1993 almost 2,000 infants and children from 28 countries around the world have had heart, heart-lung, and lung transplantation as reported to the International Society for Heart and Lung Transplantation. Sixty-three institutions have reported heart transplantation in 491 infants under one year of age and in 1,147 children one to 18 years of age. The number of procedures increased each year until 1992. Of the transplantations in infants, most were in neonates. The most common indications have been congenital heart disease in infants and cardiomyopathy in children. The 30-day perioperative mortality rate for pediatric heart transplantation in 1992 is 15% as compared to 10% in adults. The 36-month survival rate is 66% in infants and 71% in children. The most common cause of death is infection in infants and coronary artery disease in children. Nonspecific graft failure and acute rejection are common causes in both infants and children. The overall 12-month survival rate of heart retransplantation is 50% in infants and children, similar to the rate of 54% in adults. The registry has received reports of children who have had 202 heart-lung transplantations in 27 institutions and 112 lung transplantations in 22 institutions. Indications have been congenital heart disease, primary pulmonary hypertension, cystic fibrosis, and congenital and acquired pulmonary disease. The 5-year survival rate for pediatric heart-lung transplantation is 40% to 44%. The 2-year survival rate for pediatric double-lung transplantation is 57%. Infection is the most common cause of death for all heart-lung and lung transplantations in children. Heart transplantation provides significant short- and long-term benefits to infants and children offering an otherwise unavailable solution in the treatment of certain complex cardiovascular diseases. Heart-lung and lung transplantation provides good short- and mid-term results, but currently the long-term benefits are limited by the almost invariable development of obliterative bronchiolitis.

**Intraoperative measurement of left ventricular performance in children with congenital heart disease**

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To define a safe and reproducible means of assessing intraoperative left ventricular (LV) performance in children with congenital heart disease, eight children (age 21±8 months, weight 8.8±1.8 kg) underwent intraoperative measurement of load-independent indices of LV function. Epicardial ultrasonic transducers were placed to measure the LV anterior to posterior minor axis diameter and a micromanometer was placed in the LV. After separation from cardiopulmonary bypass, pressure and dimension data were acquired at baseline and during transient partial inferior vena caval occlusions (VCO). Baseline heart rate was 151±9 beats/min, mean ejection pressure 71.6±6.2 mm Hg, end diastolic pressure 9.7±1.1 mm Hg and end-diastolic diameter 34.5±9.6 mm. Ventricular performance was assessed by the stroke work-end-diastolic length (SW-EDL) and end-systolic pressure-length (ESP-L) relations. Both the SW-EDL and ESP-L relations were linear,  $r=0.99±0.01$  and  $0.78±0.1$ , respectively. The mean slope of the SW-EDL relation was  $81.8±8.6$  kerg/cm<sup>3</sup> with an x-intercept of  $31.6±27$  mm. The mean slope of the ESP-L relation was  $12.4±8.0$  mm Hg with an x-intercept of  $26.2±2.9$  mm. Two VCO's were performed in each patient with no change in heart rate, mean ejection pressure, end diastolic pressure or end diastolic diameter ( $p>0.1$ ). Neither the slope nor the intercept changed between VCO's for either the SW-EDL or the ESP-L relations, ( $p>0.1$ ). No complications occurred. This technique allows safe, reproducible, intraoperative measurements of load-independent indices of LV function in children and provides a basis for studies of the physiology of cardiopulmonary bypass and myocardial protection.

**Cardiovascular effects of cocaine in the neonate after prenatal exposure**

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This prospective study compares cardiac function, heart rate, rhythm and conduction disturbances in term neonates chronically exposed to cocaine in utero with those unexposed. Three groups were studied in first 24 hours of life: Group A (n=32) consisted of neonates with positive maternal cocaine ingestion (MCI+) and positive neonatal urine cocaine (NUC+). Group B (n=23) consisted of (MCI+) but (NUC-) neonates. Group C (n=32) consisted of (MCI-) and (NUC-) neonates as control. Mean Apgar scores and gestational ages were similar in all groups. Group C had a higher mean birth-weight than Group B ( $p<0.05$ ) and Group A ( $p<0.05$ ). Echo data revealed no statistical difference between groups A, B and C ( $p>0.05$ ) for left ventricular ejection fraction  $0.67\pm0.08$  vs  $0.68\pm0.08$  vs  $0.69\pm0.08$ . One patient in Group C had a small ventricular septal defect. No other significant heart defect was seen. Holter monitor for 24 hours likewise revealed no statistical difference ( $p>0.05$ ) for the following measured parameters: minimum heart rate  $96\pm14$  beats/min vs  $99\pm18$  beats/min vs  $97\pm13$  beats/min, maximum heart rate  $188\pm13$  beats/min vs  $196\pm10$  beats/min vs  $191\pm12$  beats/min and average heart rate  $134\pm10$  beats/min vs  $136\pm11$  beats/min vs  $131\pm23$  beats/min. Supraventricular ectopic beats  $>12$ /hour were noted in one group B, two group A and two group C patients. No patient had supraventricular tachycardia. Ventricular ectopic beats  $>10$ /hour were not present in any of the groups. One group C patient had a four beat run of ventricular tachycardia. No significant atrioventricular or intraventricular conduction disturbances were noted in any group. In contrast to previous reports, this study did not find any significant structural, functional or electrocardiographic abnormalities in neonates chronically prenatally exposed to cocaine compared to those not exposed.

**The effect of low dose aspirin (ASA) on the cardiac function (CF) of fetuses and newborns**

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A multicenter preeclampsia prevention study evaluated the utility of ASA (60 mg) administered to women during the 2nd and 3rd trimester of pregnancy. ASA was compared to placebo in a double blind protocol. During this study 146 two-dimensional-Doppler examinations were performed on 63 fetuses from 15 to 40 weeks gestational age (GA) to assess the effects of ASA on CF. Left ventricular output (LVO) and diastolic area (LVDA), right ventricular output (RVO) and diastolic area (RVDA), ductal peak velocity (DPV), mitral peak (M) E and A velocity, as well as peak tricuspid (T) velocity, were measured noninvasively. Shortening fraction (SF), cardiac output (CO), ductal patency (PDA), and frequency of tricuspid regurgitation (TR), were assessed in 73 newborns. In the fetuses DPV, RVO, RVDA, LVO, LVDA increased with GA. There was no difference in these parameters between ASA and placebo exposed fetuses. M and T flow velocities were also similar in both groups. In newborns, there was no difference in FS, CO, frequency of PDA or TR between the groups. Fetal exposure to low dose ASA during gestation does not adversely effect the circulation before or after birth.

**Temporary dual-chamber pacing in pediatric patients with hypertrophic obstructive cardiomyopathy**

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Dual chamber permanent pacing with right ventricular preexcitation in adult patients with hypertrophic obstructive cardiomyopathy (HOCM) and pharmacotherapy failure has provided excellent gradient reduction and relief of symptoms. In an effort to evaluate this technique in pediatric patients with HOCM, 5 patients (age 8-17 years, m=13) underwent cardiac catheterization and temporary dual chamber pacing. Each patient had a maximum left ventricular outflow tract (LVOT) gradient greater than or equal to 40 mm Hg by Doppler echocardiography and had failed pharmacotherapy for gradient reduction. Comprehensive hemodynamic data were obtained in patient's baseline rhythm and with atrioventricular sequential pacing at rates of 100-200 bpm with short A-V intervals (75-125 ms). Comparing baseline rhythm to paced rhythm in 4/5 patients who responded, significant differences were noted for LVOT gradient ( $54\pm16$  vs.  $8\pm3$  mm Hg,  $p<0.05$ ), left ventricular systolic pressure ( $147\pm19$  vs.  $107\pm12$  mm Hg,  $p<0.05$ ) and left ventricular end diastolic pressure ( $18\pm3$  vs.  $11\pm1$  mm Hg,  $p<0.05$ ). No significant difference was noted for aortic pressure ( $93\pm9$  vs.  $99\pm11$  mm Hg,  $p>0.1$ ). In the patient with no response to this technique, echocardiography had revealed a HOCM variant with fixed mid-cavitary obstruction of the left ventricle by an anomalous papillary muscle. These findings suggest that dual chamber pacing may be an effective therapy in pediatric patients with HOCM. We propose that institution of chronic pacing therapy early in the disease course may prevent later compensatory hypertrophy, progression of LVOT gradient, diastolic dysfunction, and symptom onset.

**Linkage analysis of the elastin gene region in familial supravalvar aortic stenosis**

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Supravalvar aortic stenosis (SVAS) is an inherited defect causing narrowing of ascending aorta±pulmonary or other systemic arteries. SVAS may occur sporadically, segregate as an autosomal dominant trait, or as part of Williams syndrome (WS). A gene for SVAS was recently localized to the elastin gene region (7q) by linkage analysis in two families. The elastin gene may itself be a SVAS gene, since a balanced translocation interrupting this gene was reported to cosegregate with SVAS in another family. We are testing another family with SVAS for linkage to the elastin gene. This four-generation family has five affected members [by clinical examination (CE) and echocardiography (E) or cardiac catheterization] and nine at risk individuals who do not have signs of disease (by CE and E). Currently we are genotyping this family with an elastin gene intragenic two-allele polymorphism as well as other nearby microsatellite markers. Preliminary data suggests that one microsatellite marker may not be linked in this family; however, we need further genotyping data from other markers before we can rule out linkage to this region. If this family does not show evidence of linkage to elastin, this will imply that another locus for SVAS resides elsewhere in the genome.

**Prenatal experience in patients with cardiopulmonary syndrome**

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Patients (pts) with cardiopulmonary syndrome present some of the most complex forms of congenital heart diseases (CHD). This investigation aimed to evaluate prenatal diagnosis, forms of presentation, natural history, and prognosis. A total of 778 fetal echocardiograms were performed in 475 pts (1983-1993); 104 pts had heart disease, 15 of them (14%) with CHD compatible with cardiopulmonary syndrome were selected for presentation (14 pts polysplenia, one pt asplenia). The patients were referred for abnormal obstetrical scan (nine pts), fetal bradycardia (four pts) and hydrops fetalis (two pts); the first study was at  $23 \pm 5$  wks of gestation. Of the 14 pts with polysplenia, 10 had the heart in levocardia, two in mesocardia, and two in dextrocardia position. Twelve of them (86%) were complex forms of AV canal and two pts were single ventricles; seven cases (50%) were associated with double outlet right ventricle. Ten pts (71%) had normally related great arteries (GA), and four pts (29%) had malposition of the GA. Pulmonic stenosis and anomalous pulmonary venous return were present in three pts (21%), respectively. Four of the 15 pts (27%) were fetuses of diabetic mothers; 7/15 pts (47%) had congestive heart failure (CHF) in utero; 12/15 pts (80%) had severe bradycardia, in seven of them due to complete AV block. Of the 15 pts, three died during pregnancy (two terminations, one fetal demise); 10 died during the neonatal period in spite of all therapeutic efforts; only two pts (13%) survived. Complex CHD and splenic syndromes are a frequent and early cause of referral for prenatal cardiac evaluation. It may occur in diabetic mothers. Its association with complete AV block or severe bradycardia, increases the high risk of CHF in utero; its prognosis is somber. The prenatal diagnosis and management of these CHD are one of the most challenging in pediatric cardiology.

**Pulmonary arterial and venous resistance changes in response to PGE<sub>1</sub> in an isolated canine lung preparation**

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Previous studies have shown an increase in pulmonary edema during prostaglandin E<sub>1</sub> (PGE<sub>1</sub>) infusion in spite of reduction in total pulmonary vascular resistance (PVR). This observation could result from direct effects on the pulmonary vascular permeability or from increased venous resistance (R<sub>v</sub>). Our hypothesis was that venous resistance remains constant or increases while arterial resistance (R<sub>a</sub>) diminishes during PGE<sub>1</sub> infusion. The right upper lobe from mongrel dogs was removed and perfused at constant flow and maintained on 5 cm H<sub>2</sub>O CPAP. Arterial (P<sub>a</sub>) and venous (P<sub>v</sub>) pressures, and lobe weight were recorded continuously. Occlusion of arterial and venous cannulas were performed to obtain a double occlusion pressure (P<sub>do</sub>) which represents average capillary pressure. R<sub>a</sub> was determined from (P<sub>a</sub>-P<sub>do</sub>)/Q; R<sub>v</sub> from (P<sub>do</sub>-P<sub>v</sub>)/Q. Baseline pressures (P<sub>a</sub>, P<sub>v</sub> and P<sub>do</sub>) were obtained followed by pretreatment with 0.1 mg/gm lobe weight meclofenamate for cyclooxygenase inhibition (COI). In group I, 5-hydroxytryptamine (5-HT) was infused continuously to maintain P<sub>a</sub> twice baseline. In group II, U46619, a thromboxane-A<sub>2</sub> analog which predominantly increases venous resistance, was infused continuously to maintain P<sub>a</sub> 40% above baseline. In both groups PGE<sub>1</sub> was infused continuously starting at 0.01 µg/min and sequentially increased to 0.20 µg/min. Group I (n=5) demonstrated reduction in R<sub>a</sub> (p<0.01) and PVR (NS) while R<sub>v</sub> increased (p<0.05) to PGE<sub>1</sub>, which did not change in doses greater than 0.02 µg/min. In group II (n=4) R<sub>a</sub> did not change while R<sub>v</sub> increased (p<0.05) to PGE<sub>1</sub>. This effect tended to increase with the increased doses of PGE<sub>1</sub>. These data suggest that the effects of PGE<sub>1</sub> in the constricted pulmonary vascular bed are arterial vasodilatation and venous constriction. This may explain the observation that patients treated with PGE<sub>1</sub> for ductal dependent cyanotic heart disease may have pulmonary edema. Further studies are planned to determine alterations in filtration coefficient (K<sub>f</sub>) resulting from PGE<sub>1</sub> infusion. The model also holds promise for testing other vasoactive drugs used in clinical practice.

**Determinants of success for balloon angioplasty of branch pulmonary artery stenosis**

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Balloon angioplasty for branch pulmonary artery stenosis (PAS) is often recommended as a therapeutic modality for residual cardiac lesions after surgery. We report our experience on balloon angioplasty for PAS in 28 consecutive procedures performed on 23 patients from January 1991 to May 1993. The patient's age range from four months to 20 years (13 M, 10 F). All patients had hemodynamically significant PAS following surgical repair for tetralogy of Fallot (nine pts), homograft repair for pulmonary atresia (5 pts), truncus arteriosus (two pts), d-TGA (one pt) and other complex anatomy (six pts). Short-term angiographic appearance and hemodynamic changes were evaluated. Success was defined as a 50% improvement in stenotic diameter along with improvement in angiographic appearance and hemodynamic parameters. Balloon diameter of at least three times the stenotic segment was used in 12 procedures (appropriate balloon size) while smaller balloon sizes were used in 16 procedures (inappropriate balloon size due to anatomical limitations). Balloon dilation of PAS was successful in 14 procedures (50%). Successful results were achieved in 67% when appropriate balloon size was used, but in only 38% when an inappropriate balloon was used. No difference was observed whether low or high atm pressure (> 6 atm) was used in either group. In five of 14 unsuccessful procedures (36%), failure to balloon dilation was attributed to marked "distensibility" of the branch pulmonary artery stenotic segment. In these five cases, the narrowed waist was removed during dilation but returned after deflation. Complications included blood transfusion in three, balloon rupture in one and transient bradycardia in one. There were no deaths, pulmonary artery perforations or acute aneurysm formations. In three patients who failed to respond to balloon dilation, Stents were successfully placed in two. In conclusion, balloon dilation of branch pulmonary artery stenosis is safe and effective procedure in many patients who would otherwise require cardiac surgery. Appropriate balloon size correlated to success. High pressure balloon dilation did not improve the success rate. Stents proved successful in "distensible" pulmonary artery stenotic lesions not responsive to balloon dilation.

**Radiofrequency catheter ablation of a verapamil sensitive ventricular tachycardia**

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A 14-year-old girl experienced several episodes of tachycardia in the past years which were associated with gymnastic activities. The tachycardia that was converted by verapamil had a wide-QRS pattern with RBBB, superior axis and VA dissociation. The echocardiographic study, right heart hemodynamic study and cardiac biopsy were normal. A treadmill exercise test was negative. The electrophysiologic study showed that the tachycardia could be induced, terminated and entrained by rapid atrial and ventricular pacing. Single ventricular extrastimulation could induce and terminate the tachycardia. Isoproterenol or epinephrine could not trigger the tachycardia. Esmolol or adenosine could not terminate the tachycardia. Verapamil IV slowed and terminated the tachycardia. She was discharged and treated with PO verapamil (Verelan 240 mg qd). However, frequent PVCs with ventricular couplets were noted on Holter monitor. The medicine was changed to diltiazem but she still had episodes of tachycardia that required IV verapamil conversion in the E.R. Radiofrequency catheter ablation was performed (30 Watts, 60 sec) with a 7 Fr Mansfield deflectable 4 mm-tip catheter at the midseptal region of the left ventricle where a local ventricular electrogram recorded a sharp spike preceding the ventricular electrogram (30 ms preceding the QRS). Pace mapping at this site produced a QRS pattern that was identical to the tachycardia. The tachycardia was not inducible for 29 min. A booster application (30 Watts, 30 sec) was delivered to the same site. However, the tachycardia reappeared 39 min later. The third ablation (30 Watts, 30 sec) was performed at the same site in a prolonged expiratory phase to lessen the movement of the ablation catheter. No tachycardia was inducible thereafter. A booster application (30 Watts, 30 sec) was then given to the same site. The post-ablation ECG showed no change from her preablation ECG. She was drug-free and tachycardia free at follow-up.