85 DETECTION OF ATRIOVENTRICULAR VALVE ABNORMALITIES BY ECHOCARDIOGRAPHY IN CHROMOSOMAL ANOMALIES Alfonso Casta, Karen Haslund, Kirk Aleck, Janet Williams, Chalermlarp Mongkalsmai, Amarjit Singh, Harold Katkov, (Spon. by Ben Brouhard) Univ. of Texas, Galveston, Southern Illinois Univ., Springfield, Minneapolis Children's Health Center

Cardiac malformations are not uncommon in chromosomal anomalies and may contribute to early death. In this study, two-dimensional echocardiography demonstrated unusual atrioventricular valve abnormalities in 4 neonates without overt structural heart defects (except for 1) and chromosomal anomalies. Three of 4 presented with signs of congestive heart failure: one had partial trisomy 11p, redundant mitral and tricuspid valves and an aneurysm of the interatrial septum; one had Teschler/Nicola-Killian syndrome, a redundant mitral valve and a small atrial septal defect; and one had trisomy 18, a large ventricular septal defect and redundant mitral and tricuspid valves. Cardiac angiography was done in 2 of these 3 patients and documented protrussion of the redundant mitral valve into left ventricular outflow tract during diastole. The other patient without heart failure had trisomy 13 and a redundant tricuspid valve.

Unexpected atrioventricular valve abnormalities were demonstrated in these 4 patients by echocardiography. Serial echo exams will be necessary in order to determine the natural history of these abnormalities. We recommend echocardiography evaluation in infants with chromosomal anomalies even if heart disease is not suspected.

86 ELECTROPHYSIOLOGICAL STUDIES BEFORE AND AFTER CARDIAC ANGIOGRAPHY IN CHILDREN: A COMPARATIVE EVALUATION. Alfonso Casta, David W. Sapire, (Spon. by David K. Rassin), Dept. of Pediatrics, University of Texas Medical Branch, Galveston, Texas

Electrophysiological (EP) evaluation in children is performed before or after angiography in several centers. Since contrast medium is known to depress contractility and to prolong the QT interval, a prospective study was designed to compare the effects of angiography on EP studies. Eleven children with various congenital heart defects, ages 2-18 yrs, and free of arrhythmias were evaluated by programmed atrial stimulation. EP evaluation consisted of determining SNRT, Wenckebach periodicity and refractory periods before and after angiography at comparable cycle lengths. In 5/11 (45%) Wenckebach periodicity was observed at longer cycle lengths after angiography. In 6/11 (55%) the atrial effective refractory period shortened after angiography, prolonged in 1/11 (9%) and did not change in 4/11 (36%). The atrial functional refractory period shortened after angiography in 5/11 (45%), lengthened in 4/11 (36%) and did not change in 2/11 (18%). Atrioventricular nodal refractory period determination was limited by atrial refractoriness. No significant change was noted in SNRT. Cardiac angiography depresses atrioventricular nodal function as evaluated by Wenckebach periodicity, had no effect on sinus node function and shortened atrial refractory periods. Therefore, electrophysiological evaluation in children should be performed prior to cardiac angiography.

87 LEFT VENTRICULAR FALSE TENDONS (BANDS): ECHOCARDIOGRAPHIC AND ANGIOCARDIOGRAPHIC DELINEATION Alfonso Casta, Wendy J. Wolf (Spon. by Ben H. Brouhard) Department of Pediatrics, University of Texas Medical Branch, Galveston, Texas

Left ventricular false tendons have been demonstrated by two-dimensional echocardiography in adults (5%) and children (61%). After identifying 3 children with left ventricular false tendons and mitral stenosis, a prospective study was designed. Over a 4 month period, 62 children underwent complete two-dimensional echocardiographic exam for either suspected heart disease or pericardial effusion. Ages ranged between 1 day and 18 years. In 7 children, a single left ventricular false tendon was identified in the apical 4 chamber view. The diagnoses included congenital heart disease (5), cardiomyopathy (1) and functional murmur (1). The left ventricular false tendon usually extended from the left ventricular apical free-wall to the mid portion of the interventricular septum. Left ventricular angiography was performed in 6 children (including the 3 prior to the prospective study). In 5 of 6, a fine radiolucent line was seen towards the apex of the left ventricle after careful review, confirming the echocardiographic finding. No left ventricular outflow tract obstruction was detected by pressure recordings in these 6 patients.

Left ventricular false tendons were identified in 11% of children by two-dimensional echocardiography and were successfully detected during left ventricular angiography. Left ventricular false tendons appear to have a benign course and should not be mistaken for structures that can produce left ventricular outflow tract obstruction.

88 FUNCTIONAL THYROTOXIC CARDIOMYOPATHY IN CHILDREN. Anita Cavallo, Alfonso Casta, H. Daniel Fawcett, Martin L. Nusynowitz, and Wendy J. Wolf (Spon. by Walter J. Meyer), Departments of Pediatrics and Radiology, University of Texas Medical Branch, Galveston, Texas.

Myocardial performance during thyrotoxicosis (T) was evaluated by multigated radionuclide angiocardiography during graded exercise in 8 children (8-17 ys) with Graves' disease. With exercise, heart rate (HR) and systolic blood pressure (SBP) increased in all patients; left ventricular ejection fraction (LVEF) increased normally by 7 to 10% in 4 (2 male, 2 female), but did not increase significantly in 1 (male) and decreased by 3 to 8% in 3 (female). There was no significant difference in the duration of T, resting and exercise HR and SBP, or resting LVEF, between patients with normal and abnormal responses. The change in LVEF was inversely correlated with serum T_4 ($r= -.82$, $p<.02$) and T_3 ($r= -.88$, $p<.05$). Three abnormal and 2 normal responders were reassessed after return to euthyroid state. Resting HR and SBP were considerably lower than during T; resting LVEF was 64% $\pm$ 6. With exercise, HR and SBP rose to levels attained during the first study; LVEF increased in all patients by 7 to 23%. Our study suggests that T causes diminished LV reserve (compensated functional cardiomyopathy) which appears to be reversible. Severity of T (measured by T_4 and T_3 levels) may be a determining factor in the development of the functional cardiomyopathy.

89 RHYTHM DISTURBANCES AFTER FONTAN PROCEDURE Su-chiung Chen, Leonard Fagan, Soraya Nouri, D. Glenn Pennington (Sponsored by Jen Yih Chu) St. Louis University School of Medicine, Department of Pediatrics, St. Louis, MO.

Persistance of sinus rhythm is common after Fontan type operation (op) and the reported arrhythmias are infrequent. We reviewed 18 patients (pt) who survived Fontan op with regard to the occurrence of rhythm disturbances detected by ECG, Holter monitoring and stress ECG. All underwent op between 1975 and 1984 at age 3-18 years (median 11 years). Anatomical lesions were tricuspid atresia 11, univentricular heart 4, pulmonary atresia 2, transposition and pulmonary stenosis 1. Duration of follow-up was 6 months-9 years (mean 3.2 years) and all had serial ECGs. Preop rhythm disturbances were present in 8 pt (2 had short P-R intervals, 3 had ectopic atrial pacemaker or junctional rhythm and 3 had 1° A-V block). Post-op transient atrial dysrhythmias were seen in 9. Late post-op 1° A-V block developed in 1, sinus bradycardia in 2, PVCs in 3. 24 hour Holter monitor recordings were done in 12: 8 had rare PACs,, wandering pacemaker or junctional beats, 1 had short runs of supraventricular tachycardia. 8 had PVS: 6 uniform, 2 multiform. Bradycardia with junctional escape were seen in 4. Stress tests were done in 10 pt; 2 had ventricular tachycardia, and 1 also had couplets. Conclusion: Transient atrial dysrhythmias were common in immediate post-op period but prolonged atrial tachycardia was not seen. Significant ventricular dysrhythmias occurred but were rare. Sinus bradycardia developed in some. Stress test and 24 hour Holter monitoring were helpful in evaluating and detecting rhythm disturbances in post-op Fontan patients.

●90 CORRELATION OF CARDIAC HEMODYNAMICS AND BIOENERGETICS USING PHOSPHORUS NMR SPECTROSCOPY. Bernard J. Clark, Mark Hilberman, Hari Subramanian, Shoko Nioka, Mitchell Schnall, George Holland, John Egan, Mary Osbakken, Britton Chance, and William J. Rashkind. Univ. of Pennsylvania, Sch. of Medicine, Departments of Pediatrics and Biochemistry-Biophysics, Phila., PA.

A model for the study of cardiac phosphate metabolism in vivo is presented which provides simultaneous measurement of hemodynamic parameters and myocardial phosphate metabolites including phosphocreatine(PCr), ATP and inorganic phosphate(Pi). Anesthetized beagles were instrumented to provide: aortic, left ventricular, pulmonary artery, and pulmonary artery wedge pressures and allow cardiac output determinations by thermal dilution. Pacing wires provided control of heart rate. Spectroscopy was performed using a 1.9 Tesla large bore magnet with a one inch surface coil placed directly on the left ventricular free wall via a left thoracotomy. Cardiac gaiting provided spectrographic data at any point in the cardiac cycle. Ten animals had resting gaited studies and four had gaited studies during periods of increased myocardial demand produced with isoproterenol (2) or hypoxemia, PaO_2 35-40 mmHg (2). No significant change in the ratio of PCr/Pi or PCr/ATP occurred at any point during the cardiac cycle at rest or during stress. During graded hypoxia PCr/Pi and PCr/ATP levels remained stable until a PaO_2 of 20-25 mmHg at which time cardiovascular shock occurred. These preliminary data demonstrate the wide range over which the normal heart can meet its metabolic demands and suggest that similar models may provide an in vivo method for the evaluation of high energy phosphate metabolism in acquired and congenital states of cardiac dysfunction.