121 MANAGEMENT OF INFANTS WITH UNIVENTRICULAR HEART

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With definitive repair for the univentricular heart now available, review of the management of infants with this defect is important. Twenty-eight infants were seen at less than 1y and followed for 8m-17y (mean=73m). All had cardiac cath. Eight who died had autopsy; 18 had single left ventricle with an outlet chamber, and 10 had other types; 13 had pulmonary stenosis (PS) and 7 pulmonary atresia (PAtr.); 8 had no PS; 4 had coarctation; and 1 subaortic stenosis. Nine patients (pts.) with severe cyanosis had 13 systemic-pulmonary artery (SA-PA) shunts (8 Blalock-Taussig, 5 Ao-PA with Impira graft); the initial shunt was done at 1d-6m, and the second at 3m-14y; there was no operative death, but 4 died later. Two pts. with PAtr. and 1 with coarctation died without surgery. Of 6 pts. with pulmonary hypertension (PH) and PA banding (PAB) at 2-20m, 4 survived, 2 died. Three pts. had no surgery and are doing well. Of the total group of 28 pts., 20 survived beyond 1y of age. Ten pts. subsequently had more surgery (7 Glenn shunt, 2 Fontan, and 1 ventricular septation & Rastelli procedure) and 3 pts. died. The overall mortality was 39.3% (11/28); 8 of them had PS. The prognosis is better if pts. survive beyond infancy. Most surviving pts. had clinical improvement. Pts. with severe PS need SA-PA shunts, and pts. with PH need PAB at an early age. If palliative procedures are successful, the pt. could undergo Glenn procedure and Fontan procedures at an older age.

122 HEART BLOCK COMPLICATING TRICUSPID VALVE REPLACEMENT IN CHILDREN

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Evaluation of pediatric patients following tricuspid valve replacement (TVR) with Hancock porcine heterograft valves indicated a high incidence of complete heart block (CHB). Eight patients who received Hancock valves in the tricuspid position were identified. Their ages ranged from one and one half to 18 years. All patients had insufficient tricuspid valves: three had Ebstein's anomaly, three with enlarged right ventricles underwent TVR three to eight years after repair of tetralogy of Fallot, one had congenital tricuspid insufficiency associated with ventricular septal defect and pulmonic stenosis, and one had tricuspid insufficiency associated with d-transposition and mitral atresia. All patients developed at least transient CHB after surgery, five required cardiac pacing, and the two youngest received permanent pacemakers. Four of eight patients died. One died from massive bleeding in the immediate postoperative period and another from disruption of the prosthetic valve by bacterial endocarditis one and one half months after surgery. One patient with a permanent pacemaker died suddenly three months after surgery, and another who did not require cardiac pacing died suddenly 22 months after surgery. The nature of the injury to the conduction system is unknown. It is clear that CHB can be expected to complicate TVR in pediatric patients, that temporary or permanent cardiac pacing may be required after surgery, and that the long term prognosis despite pacing is only fair.

123 VALIDATION OF A PRACTICAL CARDIAC OUTPUT DETERMINATION IN CHILDREN WITH SHUNTS AND VALVULAR REGURGITATION: Wesley Covitz and Casimir Eubig (Spon by

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A first pass cardiac output determination was tailored to meet the needs of children with congenital heart disease. Acquisition was complete 3.5 minutes after a single peripheral injection of TC-99m albumin. The anterior projection was used to enable shunt determination from time activity histograms over the right lung. Cardiac output was measured over the right atrium in 31 children (6.2-61.0 Kg, mean 22.4) with a wide range of cardiac diagnoses. Cardiac output was measured over the left ventricle from the same bolus in 24 of these children and validated by cardiac catheterization (Fick method) in 15. In the absence of a shunt or valvular regurgitation cardiac output over right atrium and left ventricle were equivalent ($r=.83$, $n=17$, $p<.001$). In the presence of a left to right shunt the cardiac output at one site was equivalent to the output at the volume overloaded site divided by the pulmonary to systemic flow ratio ($r=.96$, $n=5$, $p<.001$). In two children with left heart valvular regurgitation, the time activity curves over their left ventricles were broadened giving spuriously low cardiac outputs, but their right atrial outputs were nearly identical to those at catheterization. The catheterization cardiac outputs correlated well with right atrial outputs ($r=.79$, $n=15$, $p<.001$). Cardiac output can be determined from a single injection and in a short time for children with the full spectrum of cardiac diseases.

124 IMMEDIATE EFFECTS OF CARDIAC SURGERY ON MYOCARDIAL AND RENAL FUNCTION IN CHILDREN: Wesley Covitz,

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The purpose of this study was to assess the extent of intraoperative myocardial compromise during cardiopulmonary bypass. A radionuclide ejection fraction (EF), cardiac output (CO), and left ventricular volume (V) determination, and inulin clearance were performed on 20 children (1-19 years old) 2-3 hours after total surgical repair of their congenital heart defects. Three measurements of EF, CO, and V were made at hourly intervals in 7/20 patients post-op. In spite of changes in CO and V, EF and heart rate did not change significantly. The duration of aortic cross clamp time (CT) reflects the potential for intraoperative myocardial insult. Immediately after surgery EF correlated with CT ($r=-.77$, $p<.001$) but not with pre-op EF. Late post-op (1 month-1 year) EF correlated with pre-op EF ($r=.71$, $p<.05$). Glomerular filtration rate correlated with EF ($r=.76$, $p<.01$). EF in the immediate post-op period reflected intraoperative myocardial compromise. Late post-op EF showed recovery from immediate post-op lows and was most influenced by pre-op EF. Low immediate post-op EF was associated with reduced renal function independent of the effect of altered cardiac output.

125 SYSTOLIC PRESSURE-VOLUME RELATIONSHIP DURING EXERCISE

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The purpose of this study was to noninvasively measure left ventricular (LV) contractility independent of pre-load and after-load during exercise (Ex) to unmask LV dysfunction. The peak systolic blood pressure/end-systolic LV volume (P/V) index was measured in 12 children (9-19 years) with heart disease during sitting Ex. Pressures were obtained by arm cuff. (Aortic stenosis excluded). Cardiac output, ejection fraction, and LV volume were measured serially by radionuclide technique. Resting P/V index for children with EF .55 was $1.46 \pm .36$ (mLSD, $n=5$). P/V index during maximum Ex for children with an abnormal EF response to Ex (decrease from resting value) was 2.19 ± 1.0 ($n=2$). P/V index exceeded 95% confidence levels for the abnormal contractility group at rest in 4 children ($2.3 - 4.0$ mm/mL/M²) and during Ex in 6 ($4.2 - 8.8$). Contractility improved with Ex in 2 post open heart surgery patients without residual abnormality. Their resting EF's were normal. One of three patients with moderate to severe LV volume overload had abnormal contractility based on P/V index but would not have been identified by EF response alone. P/V index obtained noninvasively during Ex provides the means to unmask LV dysfunction.

126 LEFT VENTRICULAR CONTRACTION-RELAXATION SEQUENCES IN THALASSEMIA MAJOR Lilliam M. Valdes-Cruz, Cynthia

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Echo evaluation of patients (pts) with Thalassemia Major (TM) has shown thick left ventricular posterior walls (LVPW) and, terminally, decreased LV function. This study assessed utility of a relatively new echo technique for detecting possible early myopathy in asymptomatic children with TM hypertransfused since diagnosis. High quality LVPW echoes were recorded at chordal level on 26 pts with TM (mean age=11.4 $\pm$ 1.0(SE)years) and 22 controls (mean age=11.6 $\pm$ 0.9years). Incremental LVPW thickness at normalized times in contraction and relaxation were measured by computer. TM pts had thinner LVPW at all times in contraction and relaxation ($p<.02$); at 100% contraction time, LVPW was 8.7 ± 0.32 (SE) mm vs 11.7 ± 5.5 mm control ($p<.001$); at 100% relaxation time, LVPW= 4.3 ± 1.8 mm vs 5.9 ± 3.4 mm control ($p<.001$). Contraction was linear in controls, but in TM it was faster, completing 63 $\pm$ 1.5% total contraction by 50% systolic time ($p<.01$). Relaxation by 40% diastolic time was nearly total in controls but in TM, it was slower, completing only 82 $\pm$ 1.7% of total ($p<.005$). In conclusion, contrary to older TM pts, hypertransfused children with TM do not have thick LVPW. Contraction in these children is more consistent with chronic anemia and increased LV volume; however, slower relaxation suggests early myopathic changes. This echo technique is sensitive for detection of early myopathy prior to cardiac symptoms and should be useful in assessing myocardial response to chelation therapy in transfusion-dependent anemias.