127 RESULTS OF REPAIR OF TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION IN INFANCY USING DEEP HYPOTHERMIC CIRCULATORY ARREST.

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In order to determine the results of repair of total anomalous pulmonary venous connection (TAPVC) utilizing deep hypothermic circulatory arrest and to observe the hemodynamic changes following relief of pulmonary venous obstruction, we obtained early and/or late postoperative (postop) data on 13 of 15 infants who underwent correction in infancy. Seven had supracardiac drainage (SC), 4 had cardiac drainage (C) and in 4 the drainage was infracardiac (IC). Additional lesions included PDA (8), VSD (1), hypoplastic LV (1). Pulmonary artery hypertension (PAH) was present in 13. Median age was 27 days (1.5d-8mo) and mean body weight was 3.68Kg (2.22-5.25Kg) at surgery. Three infants died (20%) within the initial hospitalization (2 SC, 1 IC); there were 2 late deaths, 3 and 10 mo postop, both with IC. Combined mortality was 33%. Systolic pulmonary artery pressure (PAp) was measured within 24 hours postop in 6, all with SC or C, and had decreased from an average of 65 mmHg down to 30 mmHg. Among the 10 long-term survivors (mean 20mo postop) 3 have not been restudied, but are well clinically. Six of the 7 infants restudied (average 15 mo postop) had normal PAp (average 22mmHg). One with IC had residual PAH and we suspect this infant has pulmonary vascular obstructive disease. None had residual shunts. We believe, on the basis of these observations, that physiological correction may be accomplished at an acceptable risk in infants with TAPVC utilizing deep hypothermic circulatory arrest.

128 LYMPHOCYTE CYTOTOXICITY TO CARDIAC FIBROBLASTS IN RHEUMATIC CARDITIS.

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The role of cell-mediated immunity in the pathogenesis of acute rheumatic carditis was investigated by measuring in vitro cytotoxicity of peripheral lymphocytes to myocardial fibroblasts. The microcytotoxicity assay was adapted using as target cells, ^{51}Cr-labelled fibroblasts derived from explants of left atrial appendage. Lymphocytes and plasma were separated from peripheral blood of patients and incubated with target cells in Eagle's minimal essential medium containing 10% fetal calf serum. An effector:target cell ratio of 200:1 was utilized. Cytotoxicity was quantitated as % release of ^{51}Cr activity. Studies to-date on lymphocytes from four non-rheumatic individuals showed a mean cytotoxicity of 9% (0-20%). Lymphocytes from two patients with acute rheumatic fever without carditis, showed only 12% and 18% cytotoxicity. In contrast, lymphocytes from two patients with acute rheumatic carditis released 81% and 90% of the radioactivity. Three patients with chronic rheumatic carditis showed cytotoxicity of 10%, 11% and 34% respectively. No significant cytotoxicity was obtained with fresh plasma alone and no enhancement of cytotoxicity occurred following addition of homologous plasma to lymphocytes. Lymphocytes from the above patients produced similar low levels of cytotoxicity (less than 20%) when skin fibroblasts were used as target cells. These data suggest that lymphocyte-mediated cytotoxicity to myocardial tissue is present and may play a role in the pathogenesis of rheumatic carditis.

129 SPECTRUM OF EXERCISE RESPONSES IN CHILDREN WITH AORTIC STENOSIS

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Significant abnormalities in cardiovascular dynamics may occur during exercise (Ex) in patients (P) with aortic stenosis (AS). This study was designed to record systolic pressure (ESP), electrocardiogram (ECG) and working capacity (WC) during upright bicycle Ex in 50 preoperative P (ages 6 to 23 years) with valvar or discrete subvalvar AS. Ninety-six normal children (ages 5 to 21 years) were the controls (C). Resting aortic gradients (AG) in the P ranged up to 234 mmHg at cardiac catheterization. Mean (±standard error) Ex data were subdivided by BSA and/or sex.

Males ($\geq 1.2M^2$)	(NO-number)	(HR-heart rate)		Females $\geq 1.2M^2$			
NO	ESP	WC	HR	NO	ESP	WC	HR
P(29)	145±6**	102±10**	190±3	P(6)	136±7**	50±9**	181±7*
C(42)	178±5	160±12	195±2	C(27)	169±4	115±11	202±2

p <.01*; <.005**

In M and F < $1.2M^2$, ESP were 115±4 in P and 133±4 in N, p < .01. WC's were 77±11 in P and 88±8 in N, p > .1. ESP was below resting value in 2 and increased up to 10 mmHg in 8 P with AG > 60 mm Hg. A positive Ex ECG occurred in 15/17 (88%) P with AG ≥60, 16/30 (53%) with AG of 10 to 59, 0/3 with AG < 10 and 7/96 (7%) N. ST depression (ST ↓) occurred in 16/30 P with normal resting ECG and 15/20 with left ventricular hypertrophy. We concluded that increased frequency of ST ↓, decreased ESP and WC are typical changes which may reflect severity or progression of AS in children.

130 POSITIVE T WAVES IN RIGHT CHEST LEAD (V1) AS AN INDEX OF RIGHT VENTRICULAR HYPERTENSION IN VENTRICULAR SEPTAL DEFECT IN THE YOUNG.

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It is believed that right ventricular pressure (RVP) at or near systemic levels is associated with negative T waves in V1 (TV1), while positive TV1 represents mild RV hypertension. This study was undertaken to assess TV1 morphology as a predictor of RVP in 35 patients (pts.) with isolated ventricular septal defects (VSD) and 28 pts. with VSD and pulmonary stenosis (PS), aged 0.5-36 mos. who had electrocardiogram and cardiac catheterization within 24 hours. TV1 morphology was classified as positive (TV1+), isoelectric (TV1±), or negative (TV1-). Significant RV hypertension was defined as a systolic RVP ≥60mmHg. or an RVP/LVP ≥0.66. Results follow:

	VSD RVP≥60mmHg.	VSD RVP≤60mmHg.	VSD,PS RVP≥60mmHg.	VSD,PS RVP≤60mmHg.
TV1+	9/9 (100%)	0/9 (0%)	11/12 (92%)	1/12 (8%)
TV1±	6/10 (60%)	4/10 (40%)	8/8 (100%)	0/8 (0%)
TV1-	3/16 (19%)	13/16 (81%)	7/8 (88%)	1/8 (12%)

Thus, TV1+ appears to be an excellent predictor of significant RV hypertension in pts. with isolated VSD, while TV1- suggests RVP ≤60mmHg. On the contrary, regardless of TV1 morphology, the majority of patients with VSD and PS had significant RV hypertension.

131 ECHOCARDIOGRAPHIC OBSERVATIONS IN PATIENTS WITH RIGHT VENTRICULAR CONDUCTION DELAY.

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The echocardiographic (E) pattern of an early systolic anterior septal notch (ESASN) in post-op patients (pts) with complete right bundle branch block has been recently reported. This study evaluates the E findings in 35 pts with a right ventricular (RV) conduction delay (CD) on ECG (rsR' pattern in V1 with QRS duration ≤.08secs). 24 pre-op pts with catheterization diagnoses of secundum atrial septal defect (2°ASD) (17), normal heart (2), mild pulmonic stenosis (2), obstructed pulmonary veins (1), ventricular septal defect (1) and IHSS (1) were evaluated. In each pt, ESASN was recorded, regardless of RV dimensions or type of septal motion. The onset of ESASN after the QRS, its amplitude and duration were 84±9msec, 2.3±55mm and 114±11msec (mean±S.D.) respectively. Time intervals were corrected for heart rate using the square root of the RR interval. 11 pts were evaluated 2-7(M=3.5) years after surgical closure of a 2°ASD. In 8 of 11 pts with, and 2 of 3 pts without a residual RVCD, an identical ESASN was noted. In 20 normal pts without an RVCD, no ESASN was observed. Our observations indicate: 1) an ESASN is a characteristic pattern of septal motion associated with RVCD on ECG; 2) the occurrence of ESASN in pts with normal hearts and both systolic and diastolic overload, regardless of RV size or type of septal motion suggests a direct relationship to the ECG pattern of RVCD; 3) the presence of ESASN in pts with normal ECG after 2°ASD repair suggests that the echocardiogram may be a more sensitive indicator than the ECG for detection of residual RVCD.

132 SURGICAL LIGATION OF THE PDA IN PREMATURE INFANTS WITH SEVERE CARDIOPULMONARY DISEASE.

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Fifteen premature infants (wt 765-2863 gms) underwent surgical ligation of the PDA. The infants had a mean birth weight of 1211 ± 559 gms and a gestation of 29 ± 3 weeks. The murmur was first noted on day 6 ± 3. Heart failure occurred in 12/15 with an onset on day 12 ± 4. Age at surgery was 22 ± 8 days. Indications for surgery were CHF (3/15), inability to wean from the ventilator (3/15) and worsening cardiopulmonary function (9/15). Associated clinical findings were RDS (13/15), pneumonia (9/15), pneumothorax (5/15) and NEC (4/15). 10/15 improved postoperatively, 3/15 were unchanged and one became worse. There were 6 deaths: 1 operative, 1 aspiration, 1 ICH and hydrocephalus, 1 massive air embolus and 2 BPD. 4/5 infants with pre-op BPD expired. Significant differences between survivors and nonsurvivors were birth weight (1376 vs 963 gms) and BPD prior to surgery. Data suggests that late surgical ligation of the PDA in patients with BPD does not improve survival.