● 145 THE EFFECT OF IONOTROPIC AGENTS ON THE ECHO IMAGED LEFT VENTRICULAR POSTERIOR WALL

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The purpose of this investigation was to determine the influence of ionotropic agents upon systolic thickening and diastolic thinning of the left ventricular posterior wall. Two populations were investigated: #I--14 children with congenital cardiac disease who had echoes recorded at catheterization before and during isoproterenol infusion, and #II--8 boys with Duchenne's muscular dystrophy who participated in a double-blind, placebo controlled, cross-over study of the effects of digoxin. All echoes were analyzed by digitizing left ventricular posterior wall endo- and epicardium and evaluating % systolic time and % diastolic time at standardized time intervals. No significant differences occurred when control and ionotropic group values were compared for either group at any percentage of systolic or diastolic time. Thus, the sequence of contraction and relaxation was unaltered by ionotropic agents. However, when wall thickness values at standardized time intervals were compared before and during administration of ionotropic agents, significantly greater ($p<.01$) systolic wall thickening was present at each time interval. A 15% mean additional systolic thickness occurred for isoproterenol and 10% mean increase occurred for digoxin. This investigation shows that ionotropic agents do not alter the shape of the systolic time or diastolic time curves but significantly change systolic contraction magnitude.

146 LONG TERM LEFT VENTRICULAR POSTERIOR WALL ECHOCARDIOGRAPHIC CHANGES IN DUCHENNE MUSCULAR DYSTROPHY

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Cardiac pathology in Duchenne's muscular dystrophy (DMD) is principally left ventricular posterior wall (LVPW) fibrosis. The aim of this study was to track long term LVPW echocardiographic (echo) changes in DMD. Echoes were recorded each 12 months from 19 boys with known DMD over 3 years and compared to controls. LVPW endo- and epicardium were digitized and % thickening and % thinning at standardized time intervals were determined; 15/19 had two-dimensional echoes. As previously reported, impaired diastolic relaxation was an early finding. The ratio of peak systolic LVPW to end diastolic LVPW was 2.06 (controls) to 1.83 (boys with DMD) ($p<.01$), but a given boy's ratio was not predictive. All 19 had LVPW thickness, adjusted for body size, below the control mean ($p<.01$). The major finding was that two wall pattern groups emerged: Group I-LVPW thickness increased normally with time and body surface area but at a low percentile curve. Group II-LVPW decreased progressively and in 4, almost no systolic LVPW thickening was found. Two-dimensional echoes were studied for change in cavity size during contraction. All Group I patients had normal two-dimensional echoes. All Group II patients had abnormally decreased LV free wall contraction. Sequential abnormal findings in DMD patients are impaired LVPW relaxation, decreased LVPW thickness in systole and end diastole, and a contraction impairment imaged with two-dimensional echo.

147 EFFECTIVE TREATMENT OF NEONATAL SUPRAVENTRICULAR TACHYCARDIA WITH AMIODARONE

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We evaluated Amiodarone, a relatively new antiarrhythmic agent, in the management of supraventricular tachycardia in 5 infants. The arrhythmias were associated with Ebstein anomaly in one case and W-P-W Syndrome in two others. The infants ages were $\bar{x}=38\pm2$(SD) weeks. Mean heart rate on admission was 298 ± 1.7(SEM). 3 infants responded initially to digoxin (0.06mg/Kg) half the dose given intravenously during the crisis. The other two babies failed to respond to such treatment, to application of a cold face cloth or to cardioversion. They received 5mg/Kg/dose of amiodarone intravenously under electronic monitoring. Reversal to sinus rhythm occurred in 65 and 73 seconds respectively. Four of the infants have received maintenance oral Amiodarone for 6 months and one for 1 year. All have remained symptom-free. No abnormalities in neurological development or physical growth have been found. Corneal microdeposits, found in adults, were not seen in these infants. The drug is a benzofuran derivate resembling thyroxine. No abnormal thyroid function (T3-T4-TSH by RIA) was found in our patients. Oral Amiodarone appears to have a half-life of 30 to 120 days ($\bar{x}=41\pm5.2$[SD]) and seems to prolong the action potential in both atrial and ventricular muscle. It may become a useful alternative for the treatment of supraventricular arrhythmia in the neonate.

● 148 LUNG FLUID IN HYPOXIC LAMBS.

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In newborn lambs, alveolar hypoxia for 3-6 h increases filtration pressure in the pulmonary microcirculation and drives fluid into the lungs: pulmonary lymph flow increases and the concentration of protein in lymph decreases (Circ Res 46:111, 1980). To be certain that the increased lymph flow was not simply the result of an acute release of fluid from the lungs, we assessed lung fluid balance in 4 unanesthetized lambs kept hypoxic for 12 h. We used radioactive albumin tracers to see if sustained hypoxia altered microvascular permeability to protein in the lungs. After a 2-4 h control period in air, the lambs breathed 10% O_2 and 5% CO_2. We measured pressures in the pulmonary artery (Ppa) and left atrium (Pla); lung blood flow ($\dot{Q}b$) and lymph flow ($\dot{Q}l$); and concentrations of protein in lymph (Lp) and plasma (Pp). During control and experimental periods, we injected ^{125}I-albumin intravenously and determined the time at which specific activity in lymph reached 1/2 that in plasma ($T_{1/2}$). Results ($\bar{X}\pm s_{\bar{X}}$; *$p<.05$):

	Ppa torr	Pla torr	Lp g/dl	Pp g/dl	$\dot{Q}b$ l/min	$\dot{Q}l$ ml/h	$T_{1/2}$ min
Air	16 ± 1	2 ± 1	3.7 ± .1	5.7 ± .1	2.3 ± .2	1.7 ± .4	126 ± 5
Hypoxia	34 ± 1*	2 ± 1	3.0 ± .2*	5.8 ± .1	3.4 ± .1*	3.4 ± .6*	131 ± 16

In all lambs, Ppa, $\dot{Q}b$, and $\dot{Q}l$ remained up for the duration of hypoxia; pulmonary vascular resistance increased 62 ± 4%; Lp was persistently low; and $T_{1/2}$ did not change. We conclude that prolonged alveolar hypoxia causes a sustained increase in transvascular filtration of fluid into the lungs of lambs; hypoxia does not influence microvascular permeability to protein in the lungs.

149 LIGATION OF ANOMALOUS LEFT CORONARY (ALC)

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The purpose of this study was to evaluate 3 patients 8-9 years after the ligation of an ALC at its insertion into the main pulmonary artery during the first year of life. The patients at 2, 4 and 8 months of age were in growth failure and profound congestive heart failure. Chest x-ray showed marked cardiomegaly and electrocardiogram (ECG) showed left ventricular enlargement (LVE) and deep Q waves in leads L_1, AVL and V_6 with flat to inverted T waves in the V leads. Cardiac catheterization (CC) and angiocardiography (A) showed the ALC (2, L-R; 1, bidirectional shunt), a dilated poorly contracting left ventricle (LV) in 3, and mitral insufficiency (MI) in 2 patients. All 3 patients had the ALC ligated and were asymptomatic and showed normal growth and development 8-9 years after operation. Two patients had normal heart size by x-ray and 1 showed only slight LVE. The ECG was normal in 1 and showed slight LVE by voltage in 2. Bicycle exercise stress test was normal in all 3, as was a Thallium 201 myocardial imaging scan. CC and A showed normal intercardiac pressures, slight dysfunction of the posterior wall of the LV in 2, slight MI in 2 and well developed collateral vessels from the right to the left coronary artery in 3. Although the present day popular treatment is to implant the ALC into the aorta either directly or with a graft, ligation may also give excellent results and preserve the developed collateral circulation.

● 150 EFFECTS OF EXCHANGE TRANSFUSION(ExTx) ON THE PULMONARY AND SYSTEMIC CIRCULATIONS IN LEFT-TO-RIGHT SHUNTS(Q_{LR})

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We studied the effect of increasing hematocrit(Hct) on the pulmonary and systemic circulations, and systemic O_2 transport(SOT) in the presence of a large Q_{LR}. Isovolemic ExTx designed to raise Hct but keep blood volume constant was performed during cardiac catheterization on 8 infants(ave age=7 wk, wt=3.8 kg) with large Q_{LR} at the ventricular level. Hct, vascular pressures, O_2 uptake, and O_2 saturations were measured prior to and after ExTx. Blood flows(L $min^{-1}M^{-2}$) and vascular resistances(Wood units) were calculated. With ExTx, Hct increased with no change in mean atrial pressures. As shown in the table, both pulmonary and systemic blood flows(Qp,Qs) fell, however SOT(ml O_2 $min^{-1}M^{-2}$) increased despite the decline in Qs. After ExTx pulmonary(Rp) and systemic(Rs) vascular resistances increased, but Rp more than Rs as shown by the rise in Rp/Rs. These resistance changes are consistent with the expected increase in vicosity, but the disparity in effect may be due to concommittant alterations in the cross sectional area of the respective vascular beds. Thus an increase in Hct may provide an important adjunct to the acute management of a large Q_{LR} by reducing the shunt flow while maintaining or improving SOT.

	Hct	Qp	Qs	Rp	Rp/Rs	Q_{LR}	SOT
pre	30±5	11.2±4.1	2.3±0.6	2.5±1.1	0.11±.05	8.9±3.9	287±94
post*	44±4	7.6±3.3	1.8±0.6	5.8±3.1	0.19±.13	5.7±3.4	337±87

* all changes significant by paired t, $p<.05$