## 97 CARDIAC HYPERTROPHY IN YOUNG SPONTANEOUSLY HYPERTENSIVE RATS BEFORE THE ONSET OF HYPERTENSION.

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Myocardial hypertrophy (MH) is usually considered the result of increased afterload in the hypertensive patient. However, we found MH in the prehypertensive stage of the disease (prior to 3 wks of age) and in older spontaneously hypertensive rats (SHR) (4-24 wks old) when hypertension was prevented by treatment during the first week of life with nerve growth factor antiserum (NGFAS). Left (LV) and right ventricular (RV) to body weight (BW) ratios were significantly elevated in SHR compared to those of normotensive Kyoto-Wistar rats (WKY) at 1d and 2 wks of age. (LV: 1d-3.11±0.09 and 2 wks-3.62±0.13 vs 2.76±0.09 and 2.95±0.07, $p < 0.05$ and $< 0.001$; RV: 1d-1.18±0.06 and 2 wks-1.0±0.006 vs 0.94±0.06 and 0.74±0.03, $p < 0.005$). Systolic blood pressure in NGFAS treated SHR remained normal up to 6 mos of age when compared to untreated SHR, 129±13 vs 190±9 mmHg, $p < 0.001$. (WKY BP=107±4). Despite the normal BP, LV/BW ratios in NGFAS treated SHR were elevated compared to WKY, 2.6±0.06 vs 2.14±0.05, $p < 0.001$. Various parameters of ventricular performances (cardiac index, peak flow velocity and flow acceleration) were decreased in both treated and untreated SHR compared to WKY. These data suggest that myocardial hypertrophy in SHR is independent of increased afterload and may reflect an underlying myocardial abnormality in familial hypertension.

## 98 PRECIPITOUS CIRCULATORY COLLAPSE IN THE IMMATURE NEWBORN FROM SHUNTING ACROSS THE DUCTUS ARTERIOSUS.

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Sudden, unexplained circulatory collapse occurring after successful initial resuscitation of the extremely immature newborn has been attributed to massive intracranial hemorrhage. Our failure to find pathologic confirmation in three such infants led to a prospective attempt to define alternative mechanisms. Ten consecutive infants between 24 and 26 weeks gestation were resuscitated, placed on ventilators and followed serially with aortograms and differential blood gases and blood pressures.

Four could not be resuscitated. After initial stabilization of the six remaining infants, leg pressures were higher than arm pressures and aortograms revealed no left to right shunt. Coincidental with rising $P_aO_2$ and radiographic evidence of improved lung aeration, leg blood pressures fell to levels equal to or lower than arm pressures in 5/6 infants. Aortograms revealed massive left to right shunts with minimal distal aortic flow. Only 1/5 had a murmur. Intractable acidemia and cardiovascular collapse occurred within 1 to 6 hours of the fall in leg pressures. 4/5 infants died. Only two had significant intracranial hemorrhage. Sudden massive ductal shunting in the immature newborn can cause precipitous circulatory collapse in the absence of intracranial hemorrhage. The ability of the extremely immature newborn to survive in the extrauterine environment may depend on early closure of this low resistance shunt.

## 99 ECHOCARDIOGRAPHIC MANIFESTATIONS OF THE MUCOPOLYSACCHARIDOSES AND MUCOLIPIDOSES

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Cardiac involvement in the mucopolysaccharidoses (MPS) and mucolipidoses (ML) was assessed by echocardiography (E) in 32 patients (pts). Standard echocardiograms were examined for asymmetric septal hypertrophy (ASH), mitral and aortic valve abnormalities, and left ventricular (LV) function. 25 pts had MPS and 7 pts had ML. The median age of the group was 7 years (range 15 months - 25 years).

ASH (septal:posterior wall thickness ratio ≥ 1.3:1) was found in 20/32 pts (63%) of the total group. Mitral valve abnormalities included a diminished EF slope (<50mm/sec) in 11/32 (34%); decreased CE excursions (<1.5cm) in 23/32 (72%); abnormally thickened chordae in 22/32 (69%); marked flutter in 24/32 (75%) thickened valve in 16/32 (50%). Aortic valve thickening was noted in 9/25 (32%) of the pts with MPS and in none of the pts with ML. LV function as determined by ejection fraction, VCF, AD, and systolic time intervals was normal in all but 2/32 (6.2%) pts.

In summary, the spectrum of cardiac lesions in MPS and ML was demonstrated by E. In addition to previously described valvular involvement, we have demonstrated gross septal hypertrophy, presumably a result of myocardial deposition. Surprisingly, LV function was not depressed in our series. However, serial studies are indicated for signs of progressive myocardial dysfunction.

## 100 CARDIAC DYSRHYTHMIAS FOLLOWING CORRECTIVE MUSTARD OPERATION FOR TRANSPOSITION OF THE GREAT ARTERIES: THREE YEARS EXPERIENCE.

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To assess the frequency and type of cardiac dysrhythmias following initial corrective Mustard operations, we reviewed the clinical and electrocardiographic data, including Holter monitoring tapes, in 63 patients operated between November 1, 1972-Dec. 31, 1975. Early (in hospital) dysrhythmias (complete heart block 2, supraventricular tachycardia 2) were associated with mortality in 4 patients, all with complicated transposition. No known late deaths from dysrhythmia have occurred in the 53 survivors. Incidence of early non-fatal dysrhythmia was 50% (26/53), with the following distribution. Junctional escape rhythm(JR)14, atrioventricular dissociation(AVD) 6, junctional tachycardia JT 2, atrial flutter(AF) 2, sick sinus syndrome(SSS) 2. Late followup (1-3 years) disclosed sinus rhythm(SR) in 6/14 with JR and persistent JR in 3. Outcome is unknown in 5. AVD persisted in 1/6, returned to SR in 4, and is unknown in 1. AF converted to SR in both patients. JT returned to SR in one patient, and reverted to JR in the other. SSS returned to SR with normal sinus node recovery time in one patient, and required permanent pacemaker and antiarrhythmic medication for recurrent tachy-brady syndrome in the other.

We conclude that in uncomplicated transposition, the Mustard operation is associated with a high frequency of transient nonfatal dysrhythmias. Long term outcome remains unknown but supraventricular mechanisms appear to remain stable or improve for 1-3 years.

## 101 NON-INVASIVE ESTIMATION OF LEFT TO RIGHT SHUNTS USING ECHOCARDIOGRAMS AND RADIONUCLIDE ANGIOCARDIOGRAPHY

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Echocardiography (echo) has been used in children with ventricular septal defects (VSD) to estimate the magnitude of left to right shunt (Qp/Qs) and has been correlated with Qp/Qs measured by Fick method using assumed oxygen consumption ($VO_2$). The purpose of this study was to compare echo determined left atrial to aortic root ratio (LA/Ao) and radionuclide angiocardiography (RN) in assessing QP/Qs.

Twenty three patients were studied with echo. Nine also had RN and area-ratio analysis of the radioisotope pulmonary dilution curve. These were then compared to Fick Qp/Qs using measured $VO_2$.

We found the LA/Ao to be poorly correlated with the Qp/Qs regardless of shunt size (r=0.51). However, the RN area ratios significantly correlated to Fick Qp/Qs (r=0.93).

Thus we conclude that the LA/Ao is not accurate for estimation of magnitude of left to right shunt whereas RN aren ratios provide a better method for estimating Qp/Qs.

## 102 CARDIOVASCULAR EFFECTS OF MATERNAL METHADONE ADDICTION IN THE NEWBORN.

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Systemic hypertension has been observed in a number of infants born to methadone-addicted mothers. A study was designed to determine the characteristics of this hypertensive state. Fifty newborns of mothers participating in the methadone program were selected. Blood pressures were obtained in both a sleeping and in a quiet, wakeful state. In 24 patients (48%) an abnormal elevation in the systolic blood pressure was documented (range: 90 to 120 mmHg.). The majority were normotensive at birth; hypertension usually developed during the third and fourth days of life. The average duration of hypertension was two weeks (range: 3 to 60 days). The following observations concerning the hypertensive state were noted: (1) hypertension was demonstrated both in the presence and absence of withdrawal symptoms. Normal blood pressures were observed in several patients with obvious withdrawal, (2) elevated blood pressure persisted several weeks after withdrawal symptoms ceased and was unaffected by usual treatment for withdrawal, (3) hypertension was unrelated to serum electrolytes, serum catecholamines, or maternal blood pressure. Electrocardiograms of 18 patients (36%) displayed abnormalities of the ST segments, T waves, and/or QT intervals. Congenital heart disease was documented in three patients (6%). Transient hypertension seems the most common cardiovascular abnormality in infants born to methadone-addicted mothers.