

- **115** ECHOCARDIOGRAPHIC ASSESSMENT OF VENTRICULAR FUNCTION DURING EXERCISE IN HYPERTENSIVE CHILDREN. Robert C. Boerth, James A. Breitwieser, Dolores Petty and Jessica T. Weinberger and Judith L. Devlin. Dept of Peds., Vanderbilt Univ., Nashville, Tennessee 37232.

Little is known about the hemodynamic response to exercise in the early development of essential hypertension (EH). Graded, supine bicycle exercise was performed in 13 children with EH (14.6±0.7 yr) and 13 normotensives (N) (12.3±1.8 yr). All children were taken to maximum voluntary exercise (PEAK). Left ventricular function was monitored echocardiographically by measurements of the left ventricular shortening fraction (LVSF). The responses to peak exercise in the two groups are shown below.

	HR (bpm)		SYSTOLIC BP		LVSF (%)		PEAK EXERCISE (Kg·M/M ²)
	REST	PEAK	REST	PEAK	REST	PEAK	
N	77±5	171±5	108±2	156±8	38±2	46±1	3935±610
EH	82±5	176±5	133±3*	204±6*	32±2*	46±1	4838±577

(* = different than N at p < 0.01)

These results show that young people with EH compared to N have a greater increase in systolic BP in response to peak exercise, and children with EH have significantly lower LVSF at rest. There is no difference of LVSF at peak exercise between EH and N showing that early EH does not impair the left ventricular response to the increased demands imposed by exercise. The reduced LVSF at rest is somewhat surprising and may indicate an early deleterious effect of essential hypertension on the heart which has not been recognized previously.

- 116** DETECTION OF PRECLINICAL LEFT VENTRICULAR DYSFUNCTION USING METHOXAMINE AFTERLOAD CHALLENGE IN PATIENTS WITH THALASSEMIA MAJOR. Kenneth M. Borow, Richard Propper, Frederick Z. Bierman (Spon. by Roberta G. Williams), Harvard Medical School, Children's Hospital Medical Center, Departments of Cardiology and Hematology, Boston.

Cardiomyopathy is the major cause of death in patients (pt) with Thalassaemia Major (TM). Standard tests of left ventricular (LV) function have not dependably identified pts with TM and cardiac dysfunction. Methoxamine afterload challenge with assessment of end systolic pressure (ESP)-dimension (ESD) relationships has been used to identify subclinical cardiac dysfunction in pts with structural heart disease. In an effort to detect early abnormalities in LV contractile reserve, 8 pts with TM, ages 6-18 years, underwent echocardiography (echo) during Methoxamine infusion. ESD was measured using standard methods. ESP was estimated from the dicrotic notch of a calibrated carotid pulse recording. All pts were asymptomatic with normal resting systolic time intervals, LV percent fractional shortening by echo and treadmill exercise capacity. ESP-D values were determined for each pt at various levels of afterload. The slope of the ESP-D curve was abnormal in 3/8 pts as defined by values >2SD below the mean for 17 control pts. These 3 pts were the oldest subjects in the TM group. The use of Methoxamine to increase afterload in combination with readily available noninvasive techniques, offers a sensitive method of unmasking LV dysfunction not evident on standard resting or dynamic exercise tests. Serial studies using this technique may identify pts at risk for the development of cardiomyopathy.

- 117** GENETIC VARIATION IN APOLIPOPROTEIN E. A CLUE TO ATHEROSCLEROSIS SUSCEPTIBILITY? Jan L. Breslow and Vassilis I. Zannis. Harvard Med. School, Child. Hosp. Med. Ctr., Dept. of Pediatrics, Boston, MA

Apolipoprotein E plays a very important role in the metabolism of plasma lipoproteins enriched in cholesterol ester. We have studied the structure of this apoprotein in human plasma utilizing high resolution two-dimensional gel electrophoresis. Our studies have revealed a complex array of apo E isoproteins which vary from individual to individual. Six subclasses of apo E have been recognized and designated β II, β III, β IV, α II, α III and α IV. We have shown that the basis for this variation is genetic and can be explained by a single locus with three alleles, ϵ II, ϵ III and ϵ IV. Individuals with the β subclasses, β II, β III and β IV, are homozygous for the apo E alleles ϵ II, ϵ III and ϵ IV, respectively. Individuals with the α subclasses, α II, α III and α IV, are heterozygous for the apo E alleles and have the genotypes ϵ II ϵ III, ϵ III ϵ IV and ϵ II ϵ IV, respectively. In a study of individuals with type III hyperlipoproteinemia, almost all had the apo E subclass β IV, whereas in the general population, the apo E allele frequencies of ϵ II=11%, ϵ III=72% and ϵ IV=17% indicate that the apo E subclasses should occur with frequencies of β II=1%, β III=52%, β IV=3%, α II=16%, α III=25% and α IV=4%. Thus the apo E subclass β IV may serve as a molecular marker for type III hyperlipoproteinemia, a condition characterized by premature atherosclerosis. The 3% of the population with this apo E subclass may be ideal candidates for a targeted atherosclerosis prevention program.

- 118** NON-INVASIVE EVALUATION OF SUPERIOR VENA CAVA (SVC) AND RIGHT ARTERIAL (RA) THROMBOSIS COMPLICATING CENTRAL HYPERALIMENTATION (CHA) IN PREMATURE INFANTS. R.L. Bucciarelli and R. Jaffe (Spon. by L. Glasgow), Depts. of Pediatrics and Radiology, University of Utah and Primary Children's Medical Center, Salt Lake City, Utah.

Within the last 6 mos., we have encountered 3 infants who presented with signs of SVC obstruction complicating CHA. Signs occurred a mean of 30 days after placement of the central line (range 21-42 days) and included deterioration of pulmonary status with pleural effusions and edema of the upper trunk. In each case 2-D echocardiography (2DE) at a frequency of 5 mHz revealed a large, mobile echodense mass in the RA, with extension through the tricuspid valve into the right ventricular outflow tract. Two infants had the diagnosis confirmed by angiography and were treated with intravenous heparin for 10 days. The other infant did not receive heparin. Serial 2DE in the treated infants showed a decrease in size of the RA thrombosis in one and complete resolution in the other. Contrast echocardiography in one of the treated infants demonstrated patency of the inferior vena cava while the SVC appeared totally occluded. These findings were confirmed at autopsy. In the untreated patient the RA thrombosis remained stable in size and was operatively removed at age 6 mos. This series indicates that 2DE is a helpful non-invasive means of diagnosing SVC and RA thrombosis complicating CHA when the clinical signs suggest its presence. Further, serial 2DE is helpful in evaluating the status of the RA thrombosis during heparin therapy and in aiding the decision concerning operative removal.

- 119** PULMONARY FUNCTION AND EXERCISE RESPONSES IN CHILDREN WITH MITRAL VALVE PROLAPSE (MVP). Michael R. Bye, Michelle M. Cloutier, Frank J. Cerny, Daniel R. Pieroni, Robert L. Gingell. (Spon. by Gerd J. Cropp). SUNY at Buffalo, Children's Hospital, Dept. of Pediatrics, Buffalo, N.Y.

A recent report (Zuwallack et al. Chest 76:17, 1979) on 20 adults with MVP showed increased residual volume in 63%, decreased steady state diffusion capacity (DLCO) in 50%, and increased alveolar-arterial O₂ gradients in 42%. We evaluated 18 children (mean age 13.52 ± 3.3 SD years) with MVP and no history of lung disease. Lung volumes, forced expiratory flow rates, airway resistance, single breath DLCO, single breath oxygen closing volume and an incremental cycle ergometer exercise test with work loads increasing by 0.3 watts/kg every 3' were performed. During the last 1' of each load, heart rate (HR), end-tidal and mixed-expired PO₂ and PCO₂, minute ventilation, oxygen saturation and blood pressure (BP) were measured. EKG was continuously monitored. Pulmonary function results were normal before and after exercise. Peak work loads, HR, ventilation, oxygen consumption and BP response were all normal; no CO₂ retention or O₂ desaturation was noted. Four subjects had unifocal premature ventricular contractions at rest, which were abolished with exercise in three. We conclude that children with MVP have normal pulmonary functions and normal cardiorespiratory response to exercise and, therefore, should not be discouraged from exercise based solely on the MVP. The reason for the differences in lung function between children and adults remains unclear.

- 120** ELEVATED BLOOD PRESSURE IN INFANTS OF PRE-ECLAMPTIC MOTHERS. Luis A. Caba, John Reed, Frank Miller, Joan E. HODGMAN. Univ. of So. Calif. Sch. of Med. LAC/USC Med. Center. Dept. of Peds. Los Angeles, California

Although Woodbury et al, (Am.J. Physiol.122:472, 1938) implicated maternal toxemia as a cause of high blood pressure in neonates, there have been no further studies confirming this association. We retrospectively analyzed data from 500 high-risk pregnancies studied in 1972*. Eighteen term infants born to preeclamptic mothers (group 1) were matched for birth weight, GA, Apgar scores and hematocrit with infants born to non-preeclamptic and otherwise asymptomatic mothers (group 2). We retrieved the heart rate (HR) intra-arterial blood pressure (ABP), and arterial blood gases. The data were obtained from one min. windows at a mean age of 4 mins. and at 10 min. intervals for the first 60 min. Heart rate levels were lower in group 1 than group 2 (p<.01) throughout the first 50 min. of life. Initial values for mean ABP were 49±5.0 in group 1 and 43±5.1 in group 2. Mean ABP remained higher in group 1 up to 30 min. (p<.001). No significant differences were found in PaO₂ or PaCO₂. These results show that term infants of preeclamptic mothers have significantly elevated ABP associated with lower HR during the immediate postnatal period, confirming the observations of Woodbury et al: The association of high blood pressure and pre-eclampsia has also been reported in older children. Transient elevation of blood pressure in the infant born to mothers with pre-eclampsia may be a precursor of later hypertension in childhood.