ECHOCARDIOGRAPHY IN CYSTIC FIBROSIS. Howard Miller, Eshagh Eshaghpour, Douglas Holsclaw, Michael Gewitz and Nobuyoshi Kawai. (Intro. by Robert Kaye). Hahnemann Med. Coll., Dept. of Ped. and Med., Philadelphia

Because of the desirability of developing better non-invasive techniques in the detection of early right ventricular changes in children with cystic fibrosis (CF), echocardiography, using continuous strip recording, was performed in 15 patients and compared to other parameters of assessment of cor pulmonale. Right ventricular anterior wall thickness per meter square ($RVAW/m^2$) and right ventricular internal diameter in diastole per meter square ($RVID_d/m^2$) were compared to clinical score of severity of disease, thoracic index, pulmonary involvement by x-ray, pulmonary function, electro- and vectorcardiography.

Using $RVAW/m^2$, positive correlation was obtained with clinical score, chest x-ray, FVC and FEV_1, while $RVID_d/m^2$ was significantly related to chest x-ray only. The internal relation of $RVID_d/m^2$ to $RVAW/m^2$ was quite good ($P=<.001$). $RVAW/m^2$ appears to be more sensitive than $RVID_d/m^2$ in the group studied, and becomes more sensitive with increasing age and severity of disease. This technique was easily performed in the younger age-group child (6-8 years), in those with markedly increased A-P chest diameters, and was not effort-dependent.

It appears that echocardiography may be a valuable adjunct to other non-invasive techniques in the evaluation of these patients.

HEMODYNAMIC STUDIES IN DOWN'S SYNDROME PATIENTS WITH CONGENITAL HEART DISEASE. Jacqueline A. Noonan and Linda R. Walters, Dept. of Pediatrics, University of Kentucky, Lexington, Kentucky.

The early development of pulmonary vascular disease in cardiac patients with Down's syndrome has been suspected but few hemodynamic studies have been reported. We have studied by cardiac catheterization 47 patients aged seven weeks to 22 years with a variety of cardiac defects including 22 with some form of an A-V cushion defect (AVC), 17 with a ventricular septal defect (VSD), four with Tetralogy of Fallot, three with isolated patent ductus arteriosus (PDA) and one with total anomalous pulmonary venous return to the coronary sinus. Pulmonary artery hypertension (PAH) was present in 41 of the 47 patients. The pulmonary vascular resistance (PVR) was at least moderately elevated in 23 patients and normal or slightly raised in 18.

Of interest was the demonstration of a PDA in 11 patients. Systemic PAH was found in one of the three with an isolated PDA while all eight patients, including six under age six years with a VSD (4 of 17) or AVC (4 of 21), had severe PAH with a mean PA pressure ranging from 60-84 mm Hg. and an elevated pulmonary vascular resistance. This unexpected high incidence of associated PDA in patients with Down's syndrome may be one explanation for the frequent and early development of pulmonary vascular disease. Significant pulmonary disease was a problem in seven patients but was not clearly related to a high PVR.

ELECTROCARDIOGRAPHIC CHANGES IN INFANTS OF DIABETIC MOTHERS. Jim B. Norton, Jr.; Betty C. Evans. (Intr. by M. J. Sweeney) Dept. of Ped., Univ. of Texas Health Science Ctr.,San Antonio.

To investigate the electrocardiographic abnormalities that might occur in infants of diabetic mothers (IODM), we compared electrocardiograms obtained from 35 such infants with those from 23 normal newborns of average weight and 18 normal newborns whose weight $\geq$ 9 pounds. Tracings were obtained on the first and second days, and in the IODM, additional records were obtained at 4-6 weeks. Of the 35 IODM, 2 were found to have congenital heart disease (CHD) and were excluded from this study. Fifteen others had radiographic evidence of cardiomegaly. Electrocardiograms from both normal groups were comparable in all respects. Significantly increased posterior voltage was noted in the IODM [normals, SV_1 = 5.6 mm ± 4.4; IODM, SV_1 = 12.4 mm ± 11.8 (P = < .01)]. This was most striking in infants with cardiomegaly (SV_1 = 16.1 mm ± 13.4). The mean QRS axis was shifted less to the right in the IODM compared to the normals, and no abnormality of the QT interval was found. Evidence of right atrial enlargement (P II > 3 mm) was found significantly more often in IODM. T wave changes were more likely in the IODM but were non-specific in character. The initial abnormalities noted in IODM disappeared by 6 weeks of age if CHD was not present. We have recognized what seems to be a common electrocardiographic pattern in IODM that may be the consequence of transient myocardial changes related to the metabolic abnormalities of this entity.

ETIOLOGY OF RIGHT BUNDLE BRANCH BLOCK (RBBB) FOLLOWING CLOSURE OF VENTRICULAR SEPTAL DEFECT (VSD). Edwin O. Okoroma, Barbara Culler, James D. Maloney, William H. Weidman, and Ralph E. Smith. Mayo Clinic and Mayo Foundation, Rochester, MN.

Recent reports on the etiology of RBBB after closure of VSD have implied that RBBB is caused solely by the ventriculotomy and that it is absent when VSDs are repaired via the tricuspid valve; this suggests that postoperative RBBB in VSD is primarily due to interruption of the right ventricular subendocardial Purkinje network. To answer this question, we reviewed the electrocardiograms, available vectorcardiograms, and operative notes of two groups: group 1 (38 patients with ventricular septal defects closed via the tricuspid valve) and group 2 (26 patients with isolated muscular VSD closed via ventriculotomy). Fifteen patients (37%) of group 1 and 14 (54%) of group 2 developed RBBB characterized by a delay in terminal forces in the immediate postoperative period. Initial activation was altered in the horizontal plane in 12 of the 15 patients (80%) of group 1 with RBBB and in 6 of the 14 patients (43%) in group 2 with RBBB. This difference between the groups was statistically significant ($P < 0.05$). We assume that this alteration in initial forces results from delay in transmission through the right bundle system. These findings suggest that damage to the right bundle near the VSD (1) alters the initial phase of ventricular depolarization and (2) may also produce a delay in terminal forces--a delay that is known to occur after interruption of the subendocardial Purkinje network resulting from ventriculotomy.

MITRAL VALVE ABNORMALITIES ASSOCIATED WITH COARCTATION OF THE AORTA: A SPECTRUM THAT INCLUDES PARACHUTE DEFORMITY OF THE MITRAL VALVE. Glenn C. Rosenquist. Johns Hopkins University School of Medicine, Department of Pediatrics, Baltimore, Md.

Fifty-three specimens in which the primary diagnosis in life had been coarctation of the aorta were examined; only nine had a mitral valve of normal size and configuration. Ten specimens had a normally formed valve mechanism which was small in comparison to both tricuspid valve and left ventricle; 3 other specimens had a normally formed but hypoplastic valve which exhibited a short free margin of the anterior leaflet. The remaining 31 specimens demonstrated a spectrum of anomalies that included some forms of parachute mitral valve and may be divided into three basic types. Some showed fused or closely apposed chordae tendineae, due to nondevelopment of the intervening space; in others the space between the papillary muscles and the ventricular wall was underdeveloped, which prevented the papillary muscles from moving independently of the ventricular wall. In one case of parachute mitral valve the chordae tendineae were attached solely to the anterolateral papillary muscle; the posteromedial papillary muscle was inserted directly to the valve, near the annulus. Although the spectrum of congenital anomalies reported here does not include all forms of mitral stenosis, it does indicate that mitral valve disease probably occurs more frequently in coarctation of the aorta than previously recognized.

THE INFLUENCE OF INTRACELLULAR RED BLOOD CELL (RBC) pH ON HEMOGLOBIN OXYGEN EQUILIBRIUM AFTER ANGIOGRAPHY. Amnon Rosenthal and Alexander Mesrobian, Harvard Med. Sch. and Children's Hosp. Med. Ctr., Dept. of Card., Boston, Mass.

Our previous observation that a leftward shift in the hemoglobin oxygen dissociation curve (HODC) occurs within minutes after injection of contrast material (Inves. Rad. 8:191,1973) prompted us to investigate the in vivo effect of contrast media on RBC pH. Serial determinations of whole blood (WB), plasma (P) and RBC pH were obtained during cardiac catheterization, prior to, and after a bolus injection (1-1.8 ml/kg) of sodium and meglumine diatrizoate (Renovist-II) in 28 patients (pts) with congenital heart disease. In 17 additional pts concomitant determinations of HODC were obtained. RBC pH was measured by freeze thaw method utilizing liquid nitrogen and a glass electrode. Mean RBC pH increased from 7.21±.01(1SE) to 7.24±.01 ($p<.001$; paired t test) 1-2 minutes after injection and remained elevated at 7.23±.01 for as long as 20 minutes ($p=.02$-$.05$). There were no significant changes in WB and P pH, although P pH decreased from 7.46±.01 to 7.43±.02($p=.06$)1-2 minutes after the injection. In the 15/17 pts with HODC determinations, an increase in RBC pH was associated with a simultaneous decrease in P50(pO_2 at 50% oxyhemoglobin saturation, pH 7.4 37°C)($p<.01$). The average increase in RBC pH of 0.03(range .01-.12)was accompanied by a decrease in P50 of 1.2 mmHg(0.4-2.8). These data suggest that a fall in RBC hydrogen ion concentration (Bohr effect) may be responsible for the leftward shift in HODC (decrease P50) after angiography.