ABSENT PULMONARY VALVE WITH VENTRICULAR SEPTAL DEFECT (APV WITH VSD). Jane L. Todd, Ellis L. Jones, Dorothy E. Brinsfield and William H. Plauth (Intr. by Richard W. Blumberg); Emory Univ. Sch. of Med., Grady Mem. Hosp., Dept. of Pediatrics, Atlanta.

A characteristic clinical picture, recognizable in the first hours of life, consisting of extreme respiratory distress, cyanosis, a to-and-fro murmur at the upper left sternal border, and bilateral pneumothorax is described in two infants. This syndrome, the result of aneurysmal dilitation of the main and branch pulmonary arteries producing compression of the bronchi, is unusual but well-described in slightly older infants. Chest tube drainage and respirator support were required by 5 hours of age in both our patients. Immediate and lasting relief from respiratory obstruction was obtained by surgical reimplantation of the right pulmonary artery to main pulmonary artery, anterior to the ascending aorta, as described by Litwin, et al., in the second infant. Severe pulmonary valve insufficiency with intractable right heart failure and inadequate net pulmonary flow persisted, leading us to believe that this syndrome, if present at birth, is characteristic of the severest type of pulmonary valve insufficiency. Prompt recognition, relief of respiratory obstruction and, probably, total correction of the cardiac lesion with a valved pulmonary conduit will be necessary for management of these patients.

INFLUENCE OF HEART RATE, AFTERLOAD AND BETA BLOCKING AGENTS ON STARLING VENTRICULAR FUNCTION CURVES IN PUPPIES. Nestor J. Truccone, Henry M. Spotnitz and Welton M. Gersony. Col. of Physicians and Surgeons. Columbia Univ., Dept. of Ped., N.Y.

In order to more precisely characterize left ventricular function in puppies, Starling ventricular function (SF) curves were compared in 12 4-8 weeks old animals. The puppies were anesthetized with morphine and chloralose, and administered practolol (PRAC) [6] or propranolol (PROP) [6]. Three SF curves were obtained: a) control, b) after beta blockade, c) after administration of 7 x blocking dose. In 3 subjects, left ventricular end diastolic pressure (EDP) was varied without controlling heart rate (HR) or afterload (AL); in 4, HR and AL were regulated; and in 5 HR, alone was controlled. AL was regulated with an intraaortic balloon to maintain mean aortic pressure within 2-5mmHg range at matched points of EDP in each curve, but pressure was allowed to increase at successive points in any given curve.

Control of HR after beta blockade did not qualitatively alter the shape of the SF curves. Regulation of AL did not affect the SF curves during the control period, but a descending limb at 8-15mmHg EDP, previously noted during 7 x blocking doses of propranolol with no AL control, was not observed. The SF curves are consistent with an increase in contractility for large doses of PRAC compared to blocking doses, and a decrease in contractility for large doses of PROP. The contractility parameter, (dp/dt)/P appeared to be preload dependent, decreasing with increasing EDP.

LEFT VENTRICULAR DYSFUNCTION IN PATIENTS WITH CYSTIC FIBROSIS. Charles R. Tucker, David H. Johnson, Amnon Rosenthal, Roberta G. Williams, Kon Taik Khaw and Harry Shwachman, Children's Hosp. Med. Ctr., Depts. of Ped. and Card., Boston, Ma.

To evaluate the possibility of left ventricular(LV) dysfunction in patients(pts) with cystic fibrosis(CF), we studied 80 ambulatory and hospitalized CF pts with a mean Shwachman-Kulczyki clinical score of 55(range 15-100) using standard pulmonary function tests, echocardiography (ECHO) and systolic time intervals(STI). ECHO measurements were made of LV internal dimensions at end-diastole(ED) and end-systole, LV posterior wall thickness(PWT) and septal thickness(ST). All values were indexed by body surface area. LV-ED volume (LVEDV) and LV ejection fraction(EF) were calculated. We observed an increase in PWT which was directly related to a decrease in clinical score($p<.01$) and mid-maximal expiratory flow rate ($p<.05$) and increased functional residual capacity (FRC)($p<.05$). EF decreased significantly with increased FRC($p<.05$). LV-EDV decreased with increased residual volume (RV)($p<.05$) and total lung capacity($p<.05$). When compared with age-corrected normal values, the ratio of the STI's, pre-ejection period and LV ejection time(PEP/LVET) increased with decreased clinical score($p<.05$) and increased RV($p<.01$) and FRC($p<.05$). PEP/LVET also increased with increased PWT($p<.001$), ST($p<.01$) and indexed right ventricular dimension by ECHO($p<.001$) and with decreased LVEDV($p<.05$). These data showing increased PWT and PEP/LVET, and decreased EF suggest LV dysfunction with increasing severity of pulmonary involvement in pts with CF.

COMPARISON BETWEEN PUPPIES AND ADULT DOGS FOLLOWING INFUSION OF DIGOXIN

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The purpose of this experiment was to determine if 3-5 week old puppies, compared to adult dogs, require proportionately larger doses of digitalis. Digoxin 5 μg/kg body weight/minute (min) was infused until a strain gauge arch on the left ventricle reached a 30% increase in the force of contraction. At that time serum and myocardial specimens were taken to measure digoxin levels by radioimmunoassay. Puppies reached 30% increase in force at 23.4(SEM 1.3) min compared to 23.0(SEM 2.7) min in the adult dogs. Correspondingly the total amount of digoxin infused was not different between puppies and adult dogs, 117.0(SEM 6.5) versus 115.9(SEM 13.3) μg of digoxin/kg body weight. There was no significant difference in the serum and myocardial digoxin values between puppies and adult dogs.

In other dogs digoxin was infused until digitalis toxicity was produced. Puppies required significantly greater amounts of digoxin before toxicity ensued. Puppies averaged 53.5(SEM 1.3) min infusion of digoxin before reaching toxicity; whereas adult dogs became toxic at 29.0(SEM 2.3) min. These experiments confirm that puppies tolerate proportionately larger doses of digitalis without becoming toxic; however the relatively larger dose of digitalis usually given to the young animal may not be necessary for an equivalent inotropic response.

PLATELET STUDIES IN CONGENITAL HEART DISEASE WITH CYANOSIS

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The etiology of thrombocytopenia in children with congenital heart disease with cyanosis (CHDWC) is unknown. Disseminated intravascular coagulation (DIC) as a possible cause of thrombocytopenia is controversial. We studied the hemostatic mechanism in 20 children with CHDWC (arterial O_2 saturation, 50-90%). Tests included platelet count, Cr^{51} platelet survival times, prothrombin and partial thromboplastin times, and levels of fibrinogen, fibrin degradation products, factors V and VIII. Defects of the soluble coagulation system were found but no coherent pattern was seen. No evidence for DIC was found. Abnormal platelet half-lives(<80 hrs)were found in 12 of 20 patients (32-75 hrs); 9 of these 12 patients had normal platelet counts. Our patients may be divided into three groups: a) 8 patients with normal platelet count and normal survival, b) 9 with normal count and shortened survival, and c) 3 with thrombocytopenia and shortened survival. Our data suggest two new findings: 1) thrombocytopenia in CHDWC can be due to rapid platelet destruction without evidence of DIC, 2) some patients have a thrombocytolytic state where increased platelet production compensates for increased destruction resulting in a normal peripheral platelet count. We postulate that in CHDWC there is a spectrum of platelet survival from normal to a "compensated" thrombocytolytic phase, sometimes progressing to a "decompensated" stage where thrombocytopenia results from destruction too rapid to be met by increased production.

ECHOCARDIOGRAPHIC FEATURES OF LEFT VENTRICULAR OUTFLOW OBSTRUCTION IN TRANSPOSITION OF THE GREAT ARTERIES. Roberta G. Williams, (Intr. by Amnon Rosenthal), Children's Hosp. Med. Ctr., Dept. of Card., Boston, Mass.

The detection and characterization of subpulmonary obstruction by echocardiography would be useful in the preoperative evaluation of patients with transposition of the great arteries (TGA). Twenty-two unselected patients with cath proven TGA were studied by standard echocardiographic techniques utilizing a Hoffrel 101B ultrasonoscope interfaced with a Cambridge fiberoptic recorder. At catheterization, 7/22 patients had evidence of significant left ventricular outflow tract (LVOT) obstruction as evidenced by the presence of a systolic pressure gradient greater than 30 mmHg or angiographic evidence for severe narrowing of the LVOT. Of these 7 patients, 6 demonstrated anterior displacement of the mitral valve ring (MVR). The remaining patient with LVOT obstruction showed a discrete posterior displacement of the anterior border of the LVOT which was responsible for narrowing of the subpulmonary area. Of the 15/22 patients with a gradient of less than 30 mmHg across the LVOT, only one had anterior displacement of the MVR which was greater than 1 cm. That patient also showed subpulmonary narrowing by angiography. These data suggest that anterior displacement of the MVR or posterior deviation of the anterior border of the LVOT indicates the presence of subpulmonary obstruction in TGA.