

109

SERIAL ECHOCARDIOGRAPHIC (ECHO) PROFILES IN INFANTS WITH COARCTATION OF THE AORTA (COARCT) William A. Findlay, Jennifer P. Wing, David J. Sahn, Stanley J. Goldberg, Gail McDonald, Hugh D. Allen, Dept. of Peds., Univ. of Az. Health Sciences Center, Tucson, Az.

Angiographic studies suggest that infants with coarct have enlarged right ventricles (RV) and small, poorly functioning left ventricles (LV). We have performed 83 M-mode echos on 13 medically managed infants with documented coarct (age 1 day-3½ yrs, BSA 0.16-0.78 m²). Associated lesions included VSD (2), ASD (2) and aortic stenosis (6). Group A included 1 echo from infants 1 day-6 wks (N=9); group B, 6 wks-4 mos (N=9); group C, 4 mos-1 yr (N=7); and group D 1-4 yrs (N=4). LV diastolic dimension (LVD), 1.6 ± 1.2 (SE) cm, in A was significantly decreased, (p<0.05) 64±5% (SE) of nl for BSA, Z=-2.5 (SD's). LVD increased in B to 82±4%, Z=-1.3, in C to 94±8% and was 97±7% of nl in D. RVD, 1.3±.15 cm, was increased (p<0.05) in A to 159±19% of nl, Z=1.8. RVD for B was 141±8%, Z=1.3, and decreased to 131±10%, Z=-.9 for D. RV wall in A was thickened, (p<0.001) 0.4±.04 cm, 247±17%, Z=4.4, and remained so, D=201±19%, Z=3.4. In A infants, septum was 96±6% of nl, Z=-.21 and LVPW 93±8%, Z=-.46. These thickened to 123±8%, Z=1.37, and 117±8%, Z=1.1, respectively, in D. Mean Vcf, corrected for heart rate, was depressed in A, 81±6% of nl (p<0.05), despite digoxin, but was normal thereafter. RV PEP/ET ratio was elevated, (p<0.05) 0.36±.03 and 0.40±.05 in A & B respectively (0.24±.005 = nl), suggesting increased pulmonary vascular resistance. This study documents LV growth and hypertrophy, improvement in LV function and regression of RV enlargement and hypertrophy in unoperated infants with coarct.

110

INFLUENCE OF MANNITOL ON SALVAGING MYOCARDIUM DURING VENTRICULOTOMY. David E. Fixler, L. Maximilian Buja, James M. Wheeler, James T. Willerson. Dept. of Pediatrics, Univ. of Texas Health Science Center, Dallas, Texas

The purpose of this study was to measure changes in coronary flow and the extent of myocardial necrosis occurring after ventriculotomy in 5 control dogs and 7 dogs receiving mannitol (M). M was infused 1 hour before and 4 hours after vertical incision of the right ventricle. Coronary flow was measured with 9 micron microspheres (during anesthesia) before and 15 minutes after incision and (during wakefulness) at 4 and 24 hours after incision. At 48 hours the dogs were killed and the areas of necrosis measured by planimetry of photomicrographs. M increased serum osmolality from 282 ± 4 (SEM) to 331 ± 3 mOsm. In controls coronary flow (ml/min·100g) to myocardium within 1 cm of the incision fell from 61 ± 8 pre-cut to 40 ± 6 at 15 minutes, 43 ± 10 at 4 hours, and 45 ± 11 at 24 hours. Coronary flow to more distal myocardium did not fall significantly after incision. In M animals coronary flow to the incisional region fell from 71 ± 15 to 41 ± 5 at 15 minutes, 20 ± 3 at 4 hours and 28 ± 6 at 24 hours. Heart rates, cardiac outputs, right ventricular, and aortic pressures did not differ between M and control animals. Control dogs had necrosis involving 21.1 ± 3.7% of the right ventricular cross sectional area compared to 74.6 ± 2.8% in M dogs. Necrosis did not extend beyond 1 cm from the incision in control animals. These data indicate that infusion of M after ventriculotomy did not increase coronary flow and failed to reduce the extent of myocardial necrosis.

111

VALUE OF ECHOCARDIOGRAPHY IN THE ASSESSMENT OF PULMONARY RESISTANCE (PR) IN D-TRANSPOSITION OF THE GREAT ARTERIES (DTGA). Fouron, J.C., Batlle-Diaz, J., Bozio A., Payot, M., Davignon, A., Spon. by Ducharme, J.R., University of Montreal, School of Medicine Ste-Justine Hospital.

The reliability of echocardiography (EC) in predicting PR was investigated in 20 DTGA (mean age: 20 months). Ten were simple DTGA, among them two had Eisenmenger reaction (ER). Three had VSD with PS, the rest either VSD, PDA or PS alone. All had an atrial septal defect. Three groups of parameters were studied: 1) EC patterns of the pulmonary valve (PV). 2) Dimensions of cardiac structures: left ventricle (LV), pulmonary artery (PA) and aortic (Ao) diameters and posterior wall thickness of LV. 3) LV and right ventricular (RV) systolic and diastolic time intervals (SDTI). These parameters were correlated with the systolic, mean (MPA) and diastolic (DPA) pressure in the PA.

Results show that: 1) EC patterns described in the PV with pulmonary hypertension (PH) are not seen in DTGA. 2) LV volume and thickness are poorly related to the PR. 3) Best correlations are found with the SDTI. The RV pre-ejection period (RVPEP) was always greater than LVPEP when PR and systemic resistance were within normal limits. Changes in the LVPEP over LV ejection time ratio were slightly more closely related to the DPA (r = 0.70) than the MPA (r = 0.65). More significant was the correlation between the LV isometric relaxation time (LVIRT) and the DPA (r = 0.90). All nine patients with an LVIRT lower than 20 msec had DPA lower than 20 mmHg. The two with ER had an LVIRT of 40 msec.

112

MYOCARDIAL BIOPSY IN INFANCY AND CHILDHOOD: MODIFIED SMOOTH MUSCLE CELLS IN ENDOCARDIAL ELASTOMYO FIBROSIS. Masahiro Fujita, Harry B. Neustein and Paul R. Lurie. University of Southern California School of Medicine and Childrens Hospital of Los Angeles, Departments of Pathology and Pediatrics, Los Angeles.

Ten patients, aged 4.5 months to 7 years with cardiomyopathies (9 congestive) had transvascular myocardial biopsies by a miniaturized technique. 4 had endocardial thickening defined as >20% of left ventricle (LV). LV endocardium was not thickened in the one patient with LV myocardial interstitial fibrosis. LV endocardial thickening resulted from the increase of two layers adjacent to the myocardium, the smooth muscle (SM) layer (normally only a few SM cells) and the subendocardial layer (normally connective tissue, capillaries, unmyelinated nerves and a few cells). Thickening was due to increase in cells, elastic fibers and collagen fibers, without inflammatory cells or increased capillaries. Cell morphology changed from (a) an innermost (toward the lumen) layer of dark SM cells with surface vesicles and myofilaments to (b) light SM cells with fewer vesicles and myofilaments to (c) "leiomyoid" cells resembling both SM cells and fibroblasts, partly lacking basement membrane but containing fusiform densities, myofilaments, rough endoplasmic reticulum, ribosomes and Golgi to (d) typical fibroblasts. Light SM cells may be able to divide. Elastin is found adjacent to leiomyoid cells and may be formed by these cells. These findings may shed light on the genesis of "endocardial fibroelastosis."

113

DEVELOPMENT OF PULMONARY ARTERY HYPERTENSION IN RATS UNDER HYPOXIC CONDITIONS, UTILIZING CHRONIC PULMONARY AND AORTIC CATHETERS. Walter J. Gamble, Marlene Rabinovitch, Pamela Allard, Mark Aronovitz, and Alexander S. Nadas; Harvard Medical School, Children's Hospital Medical Center, Dept. of Cardiology, Boston.

Under pentobarbital anesthesia, pulmonary and aortic indwelling catheters were successfully placed in 11 rats (Charles River CD; mean weight 256gm). Daily measurements were made of pulmonary artery pressure (Ppul), aortic pressure (Paos), heart rate (HR), and respiratory rate (RR) for an average of 2.5 days to assure a stable preparation. The rats were then placed in a hypobaric chamber (air at 380 mmHg absolute) for a median period of 7 days (range 4-12). At the end of the hypoxic exposure, repeat measurements were made breathing room air and after 5 min. breathing 10% O₂. The animals were then sacrificed and the right & left ventricles (RV, LV) were separated and weighed.

The Ppul rose 5 mm after 4 days and 17 mm average after 12 days exposure. No significant difference was found in the other parameters after hypoxia. Repeat measurements in 10% O₂ demonstrated no significant change from those made in air. Right ventricular hypertrophy was found in 10/11 rats exposed to hypoxia LV/RV = 2.6 ± .3, this was significantly decreased from controls (LV/RV = 3.8 ± .3 (p<.0025). We conclude, that during 12 days of hypoxic exposure pulmonary artery pressure rises continually and right ventricular hypertrophy occurs secondarily. This new technique of pulmonary artery catheterization via the jugular vein and aortic cannulation may well be applicable to the study of a broad range of chronic physiological conditions.

114

EFFECT OF ALTERATIONS IN EXTRACELLULAR pH ON ELECTROPHYSIOLOGIC PROPERTIES OF PUPPY MYOCARDIUM. Henry Gelband, Kristina Nilsson, Diane Rodman, Robert J. Myerburg, Arthur L. Bassett, Univ. of Miami School of Med., Fla.

The basis for cardiac arrhythmias observed in the pediatric patient with arterial acidosis remains undetermined. To elucidate possible arrhythmogenic mechanisms we used standard microelectrode techniques to monitor electrophysiologic characteristics of isolated preparations of puppy (3-6 days old) and adult dog Purkinje fibers (PF) and ventricular muscle (VM) superfused in Tyrodes solution at 37°C and stimulated at a cycle length of 630-800 msec. Preparations were exposed to sequential changes in extracellular pH by altering external NaHCO₃ concentration while maintaining isoosmolarity. Measurements were made on sustained impalements of PF and VM fibers at normal pH (7.35-7.45), after 1 hr. equilibration in altered pH, and again in normal pH. At pH 6.75-7.0, PF resting membrane potential (RMP) and action potential amplitude (APA) increased slightly while dV/dt was unchanged; action potential duration (APD) at 70% and 100% repolarization prolonged significantly. These changes were reversible in normal pH. In contrast, puppy VM showed much less change in APD. At pH 7.6-7.7 there were no significant electrical changes. In spontaneously beating PF automaticity decreased in low pH, and increased slightly in high pH. APD lengthening (70% and 100%) also occurred in adult dog PF but was not as prominent. Our data suggest that acidosis alone does not initiate cardiac arrhythmias observed clinically at a low pH.