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CHRONIC AFTERLOAD REDUCTION IN INFANTS AND CHILDREN WITH PRIMARY MYOCARDIAL DISEASE

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The purpose of this study was to determine the long-term effects of chronic afterload reduction with oral hydralazine (H) in patients with primary myocardial disease (PMD). Twelve children aged 3 to 36 months presented with signs of severe heart failure. The PMD was confirmed by angiography. Each patient was treated with digoxin and diuretics. After obtaining control echocardiogram (ECHO), oral H was started on a small dose which was increased to 4.0 mg/kg/day. ECHOs were first repeated at 1- to 3-month intervals and later every 6 months. The follow-up period was from 3 to 30 months with a mean of 18 months. The pre-H ECHOs were compared with those at the last follow-up visit. The shortening fraction rose from 15 ± 3 to 22 ± 7 ($p < 0.05$) while the PEP/LVET ratio decreased from 0.5 ± 0.05 to 0.3 ± 0.08 ($p < 0.001$). The left ventricular size decreased in 8 patients (from $44.3 \text{ mm} \pm 5.5$ to 37 ± 3.7 , $p < 0.02$) while it remained unchanged in the other four. However, left ventricular size normalized to body surface area decreased in all patients. One infant was lost to follow-up and 11 were clinically well while receiving H. Two patients developed antinuclear antibodies and the H was stopped. Based on these data, it is concluded that H therapy is a useful adjunct in the management of PMD in infancy and childhood.

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TRANSCATHETER PERCUTANEOUS BALLOON PULMONARY VALVULOPLASTY. P. Syamasundar Rao, Mohamed K. Mardini, King Faisal Specialist Hospital and Research Centre, Department of Pediatrics, Riyadh, Saudi Arabia

The purpose of this presentation is to review our experience with percutaneous balloon pulmonary valvuloplasty (PBPV). Seven children between the ages of 15 months and 17 years (median age 5 years) underwent PBPV. No. 9F Meditech catheters with 15 or 18 mm balloons were used depending upon the size of the angiographically measured pulmonary valve annulus. The peak inflation pressure used in the balloon varied between 2 to 5 atmospheres of pressure, and the duration of inflation was ten seconds. Following PBPV the right ventricle systolic pressure fell from 92 ± 16 to 57 ± 15 ($p < 0.001$) and the peak systolic pressure gradient from 75 ± 21 to 33 ± 16 ($p < 0.01$). The cardiac index did not change. Sudden disappearance of "waisting" of the balloon, increase in the width of the jet of contrast material as it passed through the pulmonary valve, decrease in Doppler estimate of pulmonary valvar gradient, and decrease in duration and intensity of the systolic murmur also occurred. No significant complications were encountered. PBPV is a safe and effective alternative to surgical pulmonary valvotomy and may become the procedure of choice for treatment of valvar pulmonary stenosis once favorable long-term results are documented.

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SYNDROME OF ABSENT PULMONARY VALVE

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This presentation reviews our experience with the syndrome of absent pulmonary valve (APV). Ten infants and children with APV seen during 1979-83 were analyzed. Their mean age was 11 months with a range of 1 day to 3 years. Five group I patients ($< 6 \text{ mo.}$) presented with severe tracheobronchial obstruction, cyanosis, and heart failure; five group II patients ($> 6 \text{ mo.}$) presented with history of recurrent respiratory tract infection and a murmur. Clinical, x-ray, echocardiographic (ECHO) findings were classic for this anomaly. Two children were symptom-free on medical management, and two died following surgery. Plication of PAs was performed as the sole procedure in a 5-day-old respirator-dependent infant. Seven patients underwent closure of VSD and relief of PV ring stenosis; two of these had additional PA plication and two additional PV replacement. Follow-up of these patients ranged between 1.5 and 3.5 years and all underwent ECHO and catheterization studies. Symptomatic relief and normalization of cardiac structure and function were greatest in the patients who had plication of PAs as an integral part of total repair as compared with those who had other types of surgical correction or no surgical repair. Based on this experience, we recommend total surgical correction with PA plication but without PV replacement as the procedure of choice in symptomatic patients with APV.

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EFFECTS OF RIGHT AND LEFT HEART LOADING ON LV FILLING. James L. Robotham, R. Scott Stuart, A. Michael Borkon, Kevin C. Doherty, William A. Baumgartner (Spon. by Mark C. Rogers). Johns Hopkins Medical Institutions, Johns Hopkins Hospital, Depts. of Anes./Crit. Care Med. and Surgery, Baltimore, MD 21205

Consideration of changes in LV performance during respiration generally invokes changes in either preload or afterload. We evaluated the effects of acute changes in right and left heart loading on LV filling in 5 open chest dogs acutely instrumented with mitral and aortic flow probes (QM and QA). With respiration halted, we acutely obstructed the inferior vena cava, pulmonary artery, and the ascending and descending thoracic aorta. Both IVC and PA occlusion cause QM to fall within one beat with QA then falling. Thus the lung acts as a conduit between the right and left hearts, with less than 500 msec required for volume transfer. The rapid infusion of blood into the RA with the pericardium in place results in QM falling for 2 beats and then sharply increasing. Changes in QA follow QM. In each dog the diastolic changes (QM) precede systolic (QA) consistent with diastolic interdependence being more important than systolic interdependence. Acute cross clamping of the descending aorta causes an immediate fall in QA with QM following. In conclusion: 1) Right sided events have an immediate effect on LV filling and must be accounted for in explaining any changes in the LV during respiration. A rapid increase in systemic venous return may paradoxically initially decrease LV filling. 2) Changes in LV afterload immediately influence QA with QM following.

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PHASIC MITRAL FLOW DURING SPONTANEOUS RESPIRATION James L. Robotham, R. Scott Stuart, A. Michael Borkon, Kevin C. Doherty, William A. Baumgartner (Spon. by Mark C. Rogers) Johns Hopkins Medical Institutions, Johns Hopkins Hospital, Depts. of Anes./Crit. Care Med. and Surgery, Baltimore, MD 21205

We have previously demonstrated that changes in preload will influence mitral flow (QM) prior to changes in aortic flow (QA), while a change in afterload will influence QA first. During spontaneous unobstructed inspiration and Mueller maneuvers, QA is minimum during inspiration and reaches a maximum in early expiration. We tested whether primary changes in QM or QA were dominantly responsible for the observed respiratory variation in QA. In 7 acutely instrumented dogs, we placed both mitral and ascending aortic flow probes, closed the pericardium and chest and allowed spontaneous respiration to resume. We evaluated preload (QM) or afterload (QA) dominance by measuring which integrated flow reached its inspiratory minimum or expiratory maximum first during a single respiratory cycle. Multiple cycles were analyzed in each animal:

Spontaneous Cycles		Mueller Maneuvers	
Minimum First	Maximum First	Minimum First	Maximum First
QM 53 (73%)	57 (86%)	89 (87%)	67 (87%)
QA 20 (27%)	9 (14%)	13 (13%)	10 (13%)

These data suggest that while an afterload influence does exist, preload changes dominate over afterload changes in determining changes in LV stroke volume during respiration under our experimental conditions with small changes in pleural pressure with or without a change in lung volume.

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EFFECT OF CHEST SHIELDING DURING PHOTOTHERAPY ON THE INCIDENCE OF PATENT DUCTUS ARTERIOSUS (PDA) IN PREMATURE INFANTS. Warren Rosenfeld, Ramesh Jhaveri, Verlaine Brunot, Shashi Sahdev and Hugh E. Evans. Dept. of Pediatrics, Interfaith Medical Center, SUNY/Downstate Medical Center, Brooklyn, New York.

PDA is common among premature neonates, especially those $< 1500 \text{ gms.}$ In vitro, room light inhibits contraction of immature piglet's ductal rings. Since phototherapy is used frequently on the first days of life, we compared the occurrence of PDA among prematures exposed to this intense light source with those whose chests were shielded. Sixty babies with IRDS were randomly assigned to either a treatment group (chest shielded with aluminum foil while on phototherapy-30 babies) or control group (no shield-30 babies). All were on radiant warmers, received mechanical ventilation for respiratory distress syndrome, and phototherapy (Air Shields Model #PTU 78-1) from day 1 of life. Irradiance was maintained at $> 4.0 \text{ W/cm}^2/\text{nn}$ in all cases.

		BWT (Gms)	GA (Wks)	PHOTO (Days)	PDA NO	PDA
SHIELDED	Mean	1020	30	7.7	12	18
	Range	620-1430	26-33	2-15		
NO SHIELD	Mean	1050	30	8.9	20*	10
	Range	600-1570	26-32	1-13		

*p = 0.035 (Fisher's exact test)

The significant reduction of PDA with shielding ($p < 0.035$) suggests that phototherapy is a factor that may play a role in the occurrence of PDA in prematures. Shielding may be a practical method to decrease this common complication should this initial observation be confirmed.