151

CIRCULATORY RESPONSE TO FEEDING IN LOW BIRTH WEIGHT INFANTS. Khaja Raziuddin, Alice C. Yao, and Jing J. Yoon, S.U.N.Y., Downstate Medical Center, Department of Pediatrics, Brooklyn, New York

Previous studies in term neonates have documented a decrease in calf blood flow (CBF) immediately postprandial, followed by hyperdynamic circulatory state 2 to 3 hours later (Pediat 47:378, 1971). The circulatory response to feeding of 24 low birth weight (LBW) infants was investigated at ages 4 to 24 days. Twenty of 24 infants were preterm, mean birth weight (BW), 1323 (SD ± 125G) and gestational age (GA) 33.6 ± 0.65 wks. The remaining 4 were term SGA infants, GA, 38.5 ± 0.5 wks, and BW 1747.5 ± 125G. Feeding consisted of standard formula, with mean amounts of 20 ml/kg in preterm and 30 ml/kg in term infants. The CBF was measured by venous occlusion plethysmographic method 15 to 30 minutes before feeding and ½ hourly for 2 to 3 hrs. postprandial. The calf skin temperature was kept constant throughout the study (mean 35°C). Blood pressure (BP), and pulse rate (PR) were monitored. Preterm infants showed no significant change in their ½ hour postprandial CBF from a control value of 7.04 ml/min/100 ml, while term SGA infants showed a decrease of 33% from 8.88 ml/min/100 ml. The 2½ hr. postprandial CBF in both groups increased by 16 to 30%. There were no significant changes in the PR or BP. It is suggested that unlike the term infant, preterm infants showed no immediate peripheral circulatory response to feeding. This difference may be related to gestational age or the amount of feeding. Although, the studies in term infants showed no correlation to amount of feed.

152

CARDIOVASCULAR EFFECTS OF HYPOTHERMIA AND REWARMING IN NEWBORN DOGS. John H. Reuter, Leonard I. Kleinman, Irwin J. Light and James M. Sutherland. University of Cincinnati College of Medicine, Dept. of Pediatrics, Cincinnati.

The cardiovascular effects of acute hypothermia and rapid rewarming were studied in 11 newborn dogs, 2-5 days of age, anesthetized lightly with penthothal, 20 mg/kg. Cardiac output and organ blood flows were measured by the radioactive microsphere reference organ technique. Animals were made hypothermic by placing them in an incubator without heat, resulting in rectal and skin temperatures of 33.0±.25° C and 33.7±.28° C, respectively (mean±SE). Animals were rapidly rewarmed by setting the incubator heater to maximum, resulting in a rise of rectal and skin temperatures to 35.1±.49° C and 35.5±.40° C, respectively, within 60 minutes. Rapid rewarming resulted in a slight rise in BP from 45.0 to 49.4 mmHg ($p<.025$), a slight rise in renal blood flow from 2.17 to 2.81 ml/min/g ($p<.05$) and a large (65%) increase in cerebral blood flow (CBF) from .31 to .51 ml/min/g ($p<.05$). There were no significant changes in cardiac output or blood flow to the GI tract. Measurements of the effects of cooling were made in 4 animals and revealed a marked drop in CBF from .51 to .20 ($p<.025$). During the rewarming procedure, 4 animals became apneic and bradycardic, responding to tactile stimulation. These studies demonstrate that acute changes in body temperature result in altered function of the cardiovascular system, particularly flow to the brain, and such changes may contribute to the clinical disorders found in infants under similar thermal conditions.

153

PROSTAGLANDIN E_1 INFUSION FOR DUCTUS DEPENDENT CARDIAC LESIONS. J-Michel A. Roland, Jean S. Kan, Langford Kidd. Johns Hopkins Hospital, Baltimore, Md.

Prostaglandin E_1 (PGE) infusion (0.1 µg/kg/min) was used in 15 neonates with ductus dependence of the pulmonary (Group I) or systemic (Group II) blood flow. Group I consisted of 9 patients, 8 with severe right ventricular outflow obstruction and 1 with transposition of the great arteries with severe pulmonic stenosis. The age at which infusion of PGE was started ranged from 12 hours to 34 days. Duration of infusion was from 10 minutes to 3 days. Improvement in pO_2 was seen in 7/9 pts. No increase was seen in 1 infant with hypoplastic pulmonary arteries and 1 infant age 34 days. Two pts developed fever and 1 bradycardia during infusion. In the 5 pts of Group II, 3 had extreme coarctation of the aorta (CA) and 2 interpted aortic arch. The age of infusion ranged from 24 hours to 8 days. Duration of infusion was from 70 minutes to 6 days. Increased pulse pressure in the descending aorta was noted in all 5 pts. Spontaneous correction of metabolic acidosis was preceded by a transient decrease in arterial pH in 3 pts. One infant developed seizures during infusion. One additional infant with pulmonary hypertension secondary to severe mitral insufficiency received PGE because of clinical impression of CA. There was marked improvement in cardiac output. Following surgical intervention PGE was used in 3 Group I and 1 Group II pts because of poor response to the initial surgery. Shunt revision was performed in 3. PGE is effective in ductus dependent right and left heart lesions. Prolonged infusion and effectiveness in the post-operative period is demonstrated.

154

HYPERTENSION IN COARCTATION OF THE AORTA: THE ROLE OF THE RENIN-ANGIOTENSIN SYSTEM. J-Michel A. Roland, Paul Whelton, R. Patterson Russell, W. Gordon Walker, Donald Harrington, Langford Kidd. Johns Hopkins Hospital, Baltimore, Maryland.

The role of renal ischemia in coarctation of the aorta (CA) was assessed by comparing levels of plasma renin activity (PRA) in 6 patients with CA and 13 pts with essential hypertension(EH). The patients with CA ranged from 2 months to 17 years of age and were not receiving medications at the time of study. All had clinical and arteriographic evidence of CA and demonstrated resting gradients from 20 to 110 mmHg across the CA. Samples for PRA were obtained from the right renal vein (R-PRA), the left renal vein (L-PRA) and the lower inferior vena cava (I-PRA) before (Pre-F) and ten minutes after (Post-F) the administration of intravenous furosemide (1 mg/Kg).

	Pre-F		Post-F	
	CA	EH	CA	EH
Pts	6	13	4	13
R-PRA	4.7±1.3	.5±.1	6.1±.6	.8±.2
L-PRA	4.3± .9	.5±.1	6.9±.9	.9±.2
I-PRA	3.1± .6	.3±.1	4.2±.3	.6±.1

PRA (ng/ml/hr)was significantly higher in those with CA ($P<.001$ for all values). Furosemide increased PRA in both groups but the percentage increase was greater in those with EH (CA 35%; EH 100% for I-PRA). These results suggest an important role for the reninangiotensin system in the patients with CA.

155

THE PATHOLOGY OF CONGENITAL MITRAL STENOSIS. Roger N. Ruckman, Richard Van Praagh. Children's Hospital Medical Center, Departments of Pediatrics and Pathology, Boston, Mass. (Spon. by Glenn C. Rosenquist)

This study was undertaken in an effort to clarify the principal pathologic findings of congenital mitral stenosis and to propose an anatomic classification. In a series of 64 autopsied cases, four anatomic types were found: (1) typical congenital mitral stenosis with short chordae tendineae, obliteration of interchordal spaces, and reduction of interpapillary distance (32 cases, 50 percent); (2) hypoplastic congenital mitral stenosis, associated almost always with a hypoplastic left heart syndrome (26 cases, 41 percent); (3) supramitral ring (5 cases, 8 percent); and (4) parachute mitral valve (4 cases, 6 percent). The median ages at death were: parachute mitral valve, 9 11/12 years: supramitral ring, 5 7/12 years; typical congenital mitral stenosis, 7 years; and hypoplastic congenital mitral stenosis, 7 days. Thus, parachute mitral valve had the best natural history and the hypoplastic type had the worst, while that of the other two types was intermediate. Associated malformations were present in 94 percent of cases, those with the greatest frequency being endocardial sclerosis or florid endocardial fibroelastosis in 41 percent, valvular aortic stenosis in 33 percent, hypoplasia of the aortic isthmus in 31 percent, aortic atresia in 23 percent, and coarctation of the aorta in 20 percent of the cases.

156

MITRAL VALVE PROLAPSE (MVP) SYNDROME IN DUCHENNE'S PROGRESSIVE MUSCULAR DYSTROPHY. Shyamal K. Sanyal, Robert Leung, Ralph C. Tierney, Warren W. Johnson, Samuel E. Pitner. St. Jude Children's Research Hospital, Cardiopulmonary Disease Service, Memphis, Tennessee.

Cardiac status was *prospectively* evaluated non-invasively in 20 male children, each with clinical, biochemical, electromyographic and muscle-biopsy evidence of Duchenne's progressive muscular dystrophy. A nonejection systolic click, confirmed by phonocardiography, was identified in 7 patients (3 with midsystolic murmur as well) and suggested the presence of MVP syndrome. Echocardiography supported this diagnosis in all 7 patients and in 4 others without systolic click. An abrupt midsystolic posterior motion (> 3mm) was noted in 5 patients, while a smooth, pansystolic, anteriorly concave ("hammock-like") posterior motion of the leaflet > 3mm from the CD line was recorded for all 11 patients. Additional findings were: multiple sequence lines in 6 patients and posterior coaptation of the mitral leaflet near the left atrial wall in 6.

Pathogenesis of these abnormal findings was studied by detailed macroscopic, histologic and ultrastructural examination of the heart in a patient with a nonejection systolic click and echo evidence of the syndrome. The entire heart was perfused and fixed within 2 hours of death with 2.5% glutaraldehyde at 4°C for 4 hours. The most characteristic ultrastructural findings were multifocal areas with total loss of thick as well as thin myofilaments but preservation of the transverse "T" system. These degenerative changes affected predominantly the left ventricular posterobasal area and posterior papillary muscle. *Mitral valve leaflets were normal.*

We conclude i) a high prevalence of MVP syndrome exists in Duchenne's dystrophy ii) the syndrome in these patients is an expression of cardiomyopathy rather than an *isolated dystrophic* involvement of the mitral valve leaflets.